

Update in **Anaesthesia**

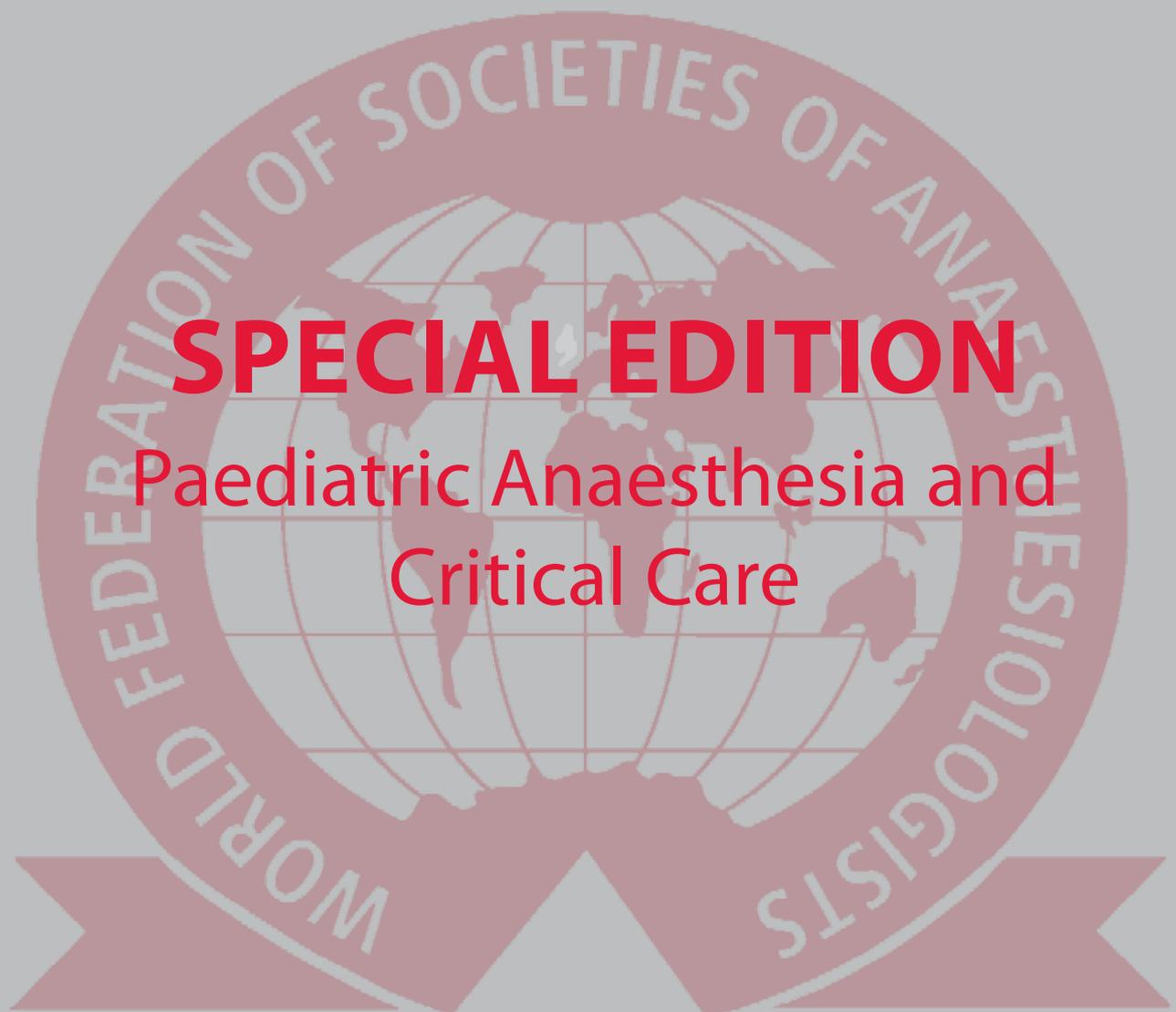
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Guest Editorial

In my opinion, paediatric anaesthesia is one of the most interesting, rewarding and fulfilling specialities – but I'm aware this may not be an opinion shared by all! There can be nothing more frightening than to be faced with an acutely ill or injured child when you don't have any colleagues to help you or to discuss the case with, and there is no time to transfer to a specialist centre, or there is no specialist centre. I imagine this must be the case particularly for those who only anaesthetise children occasionally. This edition of Update in Anaesthesia includes a wealth of information on different areas of paediatric anaesthetic practice, and will be enormously useful to all those who care for children.

Core lifesaving skills relating to airway management and fluid resuscitation are fundamental our practice, no matter what the age of the patient, and maintaining these basic skills, and basic anaesthesia skills, should form the basis of our on-going professional development. For some, this may involve spending time with a colleague during an elective operating list, so that when you need to look after a child in an emergency you feel more confident. For others, it may mean updating local guidance, for instance relating to pain management and fluid management, and making sure that the appropriate equipment is available when you need it. Even the normal infant airway can be difficult for those who are inexperienced, and it helps to have thought about your plan in advance. Neonatal anaesthesia presents very particular challenges of its own.

Preparation of a child for surgery is vital to ensure smooth and safe anaesthesia, especially in the presence of comorbidities. Asthma is increasingly common. Environmental pollution particularly affects our younger patients, and makes them more prone to respiratory infections. Whether to proceed or to cancel the child with a common cold is often a difficult dilemma, even for the experienced paediatric anaesthetist. In any setting, even those with the best of resources, anaesthetists have had to learn to trust their instincts and their senses (their eyes, ears and touch).

Modern anaesthetic teaching often emphasises the latest developments, but we should not forget the importance of the vigilant anaesthetist and the use of equipment such as the pre-cordial stethoscope, now seldom used in high resource countries.

Effective pain management in children undergoing surgery should always be a high priority, and the authors of the excellent section on regional anaesthesia highlight the importance of local blocks in children. Much is possible with simple equipment using landmark techniques, and local blocks such as the caudal provide excellent analgesia for common surgical interventions. The newer ultrasound-guided techniques described help us to perform a wider range of blocks with great accuracy and safety, and using smaller doses of drug.

The sections on resuscitation and critical care highlight some important differences between adults and children. For example, in adults, cardiac arrest is usually due to a primary cardiac cause, whilst in children, cardiac disease is rare, and the most common cause of cardiac arrest is hypoxia or hypovolaemia, or in parts of the world where halothane is still used routinely, due to deep halothane anaesthesia. This is reflected in the resuscitation guidelines for children that emphasise identification and prevention of cardiac arrest as much as treatment itself. Early recognition of a seriously ill or injured child, whether due to a common or rare disease condition, is essential to achieve a good outcome.

Paediatric anaesthesia is an important sub-speciality of anaesthesia, but sadly the facilities to deliver safe anaesthesia care are not always available everywhere. The mission of the WFSA is to 'improve patient care and access to safe anaesthesia by uniting anaesthesiologists around the world'. I believe that this edition of Update in Anaesthesia, written by experts in paediatric anaesthesia from around the globe, offers an important contribution to this mission.



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Update in Anaesthesia

Editors notes

Dear readers

Welcome to this special edition of *Update in Anaesthesia*, which focuses on paediatric anaesthesia and critical care. Anaesthetists play an important role in the care of children in hospital, providing anaesthesia, pain relief, resuscitation and critical care services for some of our most vulnerable patients.

The speciality of paediatric anaesthesia has developed over the last 30–40 years as the particular requirements for safe care of the newborn, infant, young child and adolescent have been recognised. In many countries, there are now sub-speciality paediatric anaesthesia societies, and anaesthetists can specialise in paediatrics as a sole area of practice. Whilst the approach ‘the child is not a miniature adult’ is important, it is also essential to recognise that the fundamental principles of safe anaesthetic practice can be applied to all our patients, and that there is a need for the ‘generalist’ anaesthetist to maintain their skills in caring for children. In many parts of the world, more than 50% of the population is under 14 years, and it has been estimated that more than 85% of children will require some form of surgery before their 15th birthday – whether this is for minor trauma, hernia repair or tonsillectomy, to treat common congenital abnormalities such as cleft lip and palate, or as the result of trauma from a road traffic accident. In all these areas, essential anaesthetic skills play a key role.

This edition of *Update* represents the contributions of paediatric anaesthetists from around the globe; we are grateful to them for their hard work and for sharing their wisdom. We have aimed to provide both theoretical background and practical advice that will be useful in every day practice. The section on basic science includes a description of physiological and pharmacological differences between young children and adults, and advice about the selection of equipment for children. There is a section to describe the anaesthetic implications of both common and rarer co-morbidities in children. The section on principles of basic clinical anaesthesia describes the essentials of preoperative preparation, intravenous fluid management, analgesia and sedation, that are applicable in any setting. The articles describing speciality areas of practice are written by experts in the field, and we are grateful to them for making their contributions so relevant to the practice of anaesthetists worldwide. Some of the articles have been published previously in *Update in Anaesthesia* and *Anaesthesia Tutorial of the Week*, and we have indicated this in the article where relevant. We are particularly pleased to include the

contribution from Dr Sam Richmond, who sadly died in April 2013 – Sam was a consultant neonatologist at the City Hospitals Sunderland, UK, and was dedicated to furthering education in newborn resuscitation in low- and middle-income countries.

Providing high quality anaesthesia and critical care requires a trained workforce, but it can be difficult to access refresher training in some parts of the world. The WFSA and AAGBI have pioneered the ‘Safer Anaesthesia From Education’ (SAFE) short courses for physician and non-physician anaesthetists to address these training needs. This edition of *Update in Anaesthesia* has been designed to support the SAFE Paediatric Anaesthesia course, and we hope that the SAFE course participants find it useful. We also hope that the regular readers of *Update* find it a useful addition to their anaesthesia libraries; this edition will be available along with all the other WFSA education resources at www.wfsahq.org.

All previous *Update in Anaesthesia* articles and *Tutorials of the Week* are available to download for free from <http://www.wfsahq.org/resources/virtual-library>

Finally, I would like to offer particular thanks to my fellow editors, especially to Rachel Homer for all her hard work and dedication that has seen this project through to completion.

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Update in Anaesthesia

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Basic science relevant to practical paediatric anaesthesia

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SUMMARY

This article considers the basic science and calculations commonly used in paediatric anaesthesia, including the anatomical, physiological and other differences relative to adults that impact on anaesthetic practice.

It is often said that paediatric patients are ‘not simply small adults’. The truth is that from the premature neonate to the near-adult adolescent, children are very diverse (see Table 1 for age definitions used in this article). This article will consider the basic science and calculations commonly used in paediatric anaesthesia; our challenge is to consider the anatomical, physiological and other differences that impact on anaesthetic practice.

ESTIMATION OF WEIGHT

It is essential that every child is weighed prior to anaesthesia. This allows correct calculation of drug doses and selection of anaesthetic equipment. In emergencies, weight can also be estimated from the age of the child from standard growth charts (use the weight at the 50th centile), from the length of the child using a Broselow tape, or using the formulae shown in Table 2.

AIRWAY AND RESPIRATORY TRACT

Anatomical differences of the paediatric airway influence airway management and the selection of appropriate equipment.

The major anatomical differences affecting airway management in neonates and infants are as follows:

- Relatively large head and prominent occiput
- Small mandible
- Relatively large tongue
- Short neck
- Soft tracheal cartilages, easily compressed.

These differences predispose to airway obstruction, particularly if the child’s head is placed on a pillow, or the soft tissues on the floor of the mouth are compressed, or the head is hyperextended. Ideally, maintain the child’s head in a neutral position, or slightly extended, possibly with a small pad under the shoulders, and open the airway using a chin lift or jaw thrust, avoiding compression of the soft tissues of the floor of the mouth (see Figure 1).

Anatomical differences affecting the larynx include:

Table 1. Age definitions

Neonate	Up to 44 weeks post conception (includes premature neonates)
Infant	From 44 weeks post conception – 1 year
Child	1 – 12 years of age
Adolescent	13 – 16 years of age
Adult	Greater than 16 years of age

Table 2. Formulae to estimate the weight of children at different ages

Age of child	Formula to estimate weight in kg
0-12 months	(0.5 x age in months) +4
1-5 years	(2x age in years) +8
6-12 years	(3x age in years) +7

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Chin lift



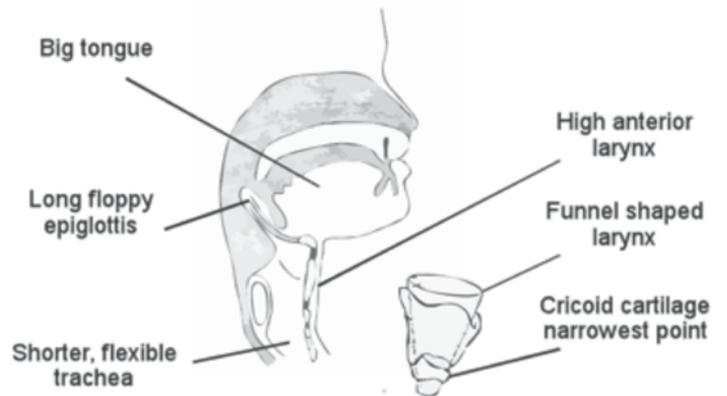
Jaw thrust



Figure 1. Chin lift and jaw thrust in a child, avoiding compression of the soft tissues

- A high, anterior position of the larynx (level of C3-4 in infants compared to C5-6 in adults)
- A long, U-shaped epiglottis projecting at 45 degrees posteriorly
- A 'funnel shaped' larynx. The narrowest part of the airway is at the cricoid cartilage (below the vocal cords). The narrowest part of the airway in adults is at the vocal cords.
- A thin loose lining to the airway which is easily damaged

Paediatric Airway



Adult Airway

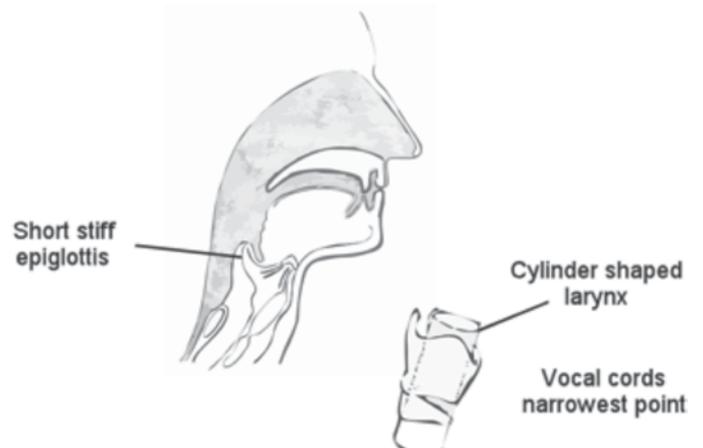


Figure 2. The paediatric airway compared to the adult airway (illustration by Mrs P. Klebe, used with permission)

The intubation technique in infants needs to take account of these anatomical differences:

- Prepare all your equipment, find an assistant, monitor the child and preoxygenate; give yourself enough time
- Handle the tissues gently and choose the appropriate sized tube. Multiple attempts at intubation may result in post-extubation stridor
- Place the head in a neutral position or slightly extended, and stabilise the head with your right hand; use your right index finger to open the mouth
- Hold the laryngoscope close to the hinge of the blade, using the thumb and index finger of your left hand.
- If necessary, use the little finger of your left hand to press on the larynx to bring the laryngeal structures into view.

- Place the tip of the laryngoscope blade into the vallecula, the space above the epiglottis (curved blade), or beneath the epiglottis (straight blade) to lift the epiglottis to expose the larynx and vocal cord.
- Pass the tube carefully between the vocal cords, under direct vision. Do not insert the tube too far; the tracheal length is approximately 4.5cm in most infants.

Adenotonsillar hypertrophy is common in children 3 – 8 years of age. Airway obstruction may develop when pharyngeal tone is lost after induction of anaesthesia; an oropharyngeal may help to maintain a patent airway. Take care when passing nasopharyngeal, nasotracheal and nasogastric tubes in these children.

Children aged 5-13 years may have loose teeth; take note of loose teeth at your preassessment visit.

RESPIRATORY CONSIDERATIONS

Up to 6 months of age, infants are almost exclusively breast fed, and need to breathe through their nose rather than their mouth (obligate nasal breathers). Respiratory difficulties may result if the nose is blocked, for instance due to secretions from upper respiratory tract infections, or if a nasogastric tube is present.

Neonates have very limited respiratory reserve, and become hypoxic very easily. They have a high metabolic rate and twice the oxygen consumption compared to older children and adults (6-7ml.kg⁻¹.min⁻¹ in neonates compared to 3-4 ml.kg⁻¹.min⁻¹ in adults).

The respiratory exchange surface is immature, with only 1/10 the number of alveoli compared to adults; in premature neonates this is compounded by a lack of respiratory surfactant that helps to reduce surface tension and to stabilise the alveolar air spaces. The lack of surfactant in premature infants predisposes them to airway collapse, poor gas exchange and increased work of breathing. Ventilation with high airway

pressures and high-inspired oxygen concentration predisposes to bronchopulmonary dysplasia and chronic lung disease.

The airways are very small in neonates, and easily obstructed. The flow in the airway can be described by the Hagen Pouiselle formula, assuming laminar flow:

$$Q = (\Delta P \pi r^4) / (8 \mu L)$$

where

Q = volumetric flow rate

ΔP = pressure drop

π = a constant

r = radius

μ - dynamic viscosity

L – airway length

The flow is therefore proportional to the radius⁴; halving the radius of the airway decreases the flow rate by a factor of 16. A small amount of airway oedema from a difficult intubation, or infection, or respiratory secretions, will significantly reduce airflow and increase the work of breathing for a neonate.

Respiratory mechanics in the neonates are not very efficient. The rib cage is soft and compliant, and the ribs move in the horizontal plane only (rather than in the horizontal and anterior-posterior direction in adults, like a bucket handle). This means the tidal volume is relatively fixed (5-7 ml.kg⁻¹), and the infant can only increase minute ventilation by increasing respiratory rate (see Table 3 for normal values). Dead-space volumes should be kept to a minimum for neonates and infants to reduce the work of breathing and to reduce re-breathing.

The diaphragm is the main muscle of respiration in neonates and infants, but is prone to fatigue due to a lack of type 1 (oxidative, fatigue resistant) muscle fibres. The diaphragm may be splinted by gastric distension due to swallowed air

Table 3. Cardiorespiratory physiology – normal values

Age (years)	Respiratory rate (breaths per minute) Rate reduces with increasing age	Heart rate (beats per minute) Rate reduces with increasing age	Blood pressure (mmHg) Pressure increases with increasing age
<1	40-30	160-110	80-90
>1-2	35-25	150-100	85-95
>2-5	30-25	140-95	85-100
>5-12	25-20	120-80	90-110
>12	20-15	100-60	100-120

(very common with crying or facemask ventilation), or due to bowel obstruction. It is important to consider placement of a nasogastric tube to decompress the stomach in infants with abdominal distension or respiratory distress.

The soft rib cage means that the chest wall in babies is highly compliant, and there is less 'outward spring' exerted by the rib cage, and less negative intrapleural pressure to keep the lungs expanded. This results in a relatively low functional residual capacity (FRC), and airway closure may occur during normal breathing. Intercostal and sternal recession is common in babies if there is any airway obstruction, or reduction in lung compliance (for example due to infection) (see figure 3). Tracheal tug also occurs. An infant with respiratory distress may 'grunt' to maintain airway volumes – there is partial closure (adduction) of the vocal cords during expiration, which effectively provides physiological continuous positive airway pressure (CPAP), and helps to keep the airways open. Conversely, the presence of recession, tracheal tug and grunting in an older child with a more rigid rib cage is an ominous sign, and suggests very severe respiratory distress. Increased respiratory rate is an important sign of respiratory distress at any age.

The control of respiration is immature in neonates. Responses to hypercarbia and hypoxia are blunted and poorly sustained, and neonates often respond to hypoxia by stopping breathing

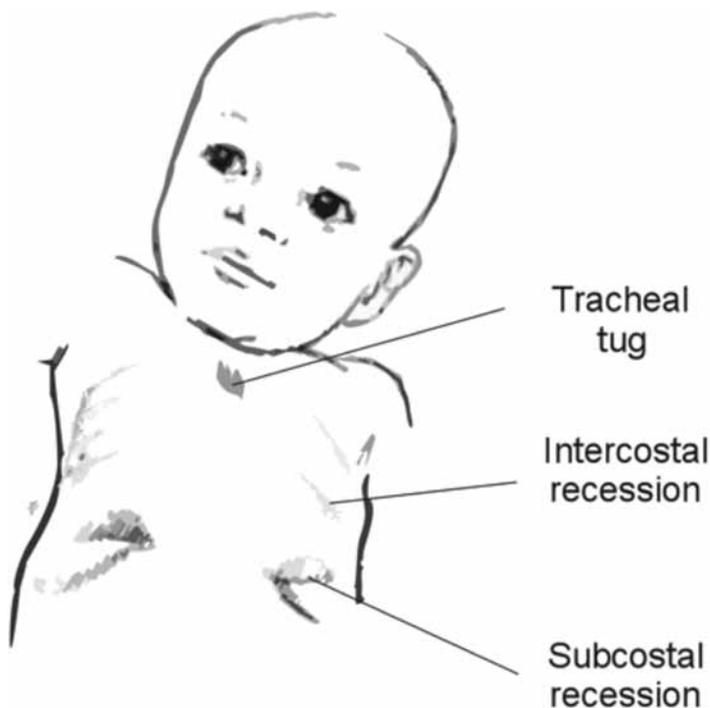


Figure 3. Tracheal tug, subcostal and intercostal recession is common in babies with respiratory distress (illustration by Mrs P. Klebe, used with permission)

(apnoea). Apnoeas are particularly common in premature and ex-premature infants, and are significant if they last longer than 15 seconds or are associated with desaturation or bradycardia. Volatile anaesthetics and opioids reduce the respiratory drive further, and should be used cautiously in neonates, especially premature neonates. Consider opioid sparing techniques (paracetamol, local anaesthetic blocks) whenever possible. Caffeine given orally prior to surgery has been used to reduce the risk of apnoea. All ex-premature neonates <60 weeks post conception should be monitored carefully after an anaesthetic, ideally with an apnoea monitor if available. Term neonates are also susceptible to apnoeas after routine anaesthesia for minor procedures, likely until they are at least a month old.

Premature infants (before 35 weeks gestational age) are at risk of retinopathy of prematurity due to abnormal vessel proliferation in the vitreous of the eye, which may result in haemorrhage, scarring and retinal detachment, and is a common cause of blindness in this population. High PaO₂ may worsen retinopathy, which is also seen in term infants given unmonitored oxygen therapy. If oxygen therapy is required on the ward, saturations of 87-94% are acceptable in neonates, particularly premature neonates. Preoxygenation is still indicated prior to intubation, but if possible, avoid 100% FiO₂ during the maintenance phase of anaesthesia.

Summary - practical implications for the anaesthetist:

- Consider the normal values for respiratory rate by age
- If a mechanical ventilator is available, select the appropriate tidal volume and respiratory rate for age – pressure control ventilation is preferred
- Recognise the signs of respiratory distress; increased respiratory rate, grunting and recessions in an older child are extremely ominous
- Neonates and infants have a high oxygen requirement and limited reserve; they become hypoxic rapidly if they are apnoeic or if the airway is obstructed. This is particularly important during induction of anaesthesia
- Small children may not tolerate preoxygenation well, but you should always try
- Gastric distension splints the diaphragm, the main muscle of respiration in neonates and infants; consider a nasogastric tube (NGT) in cases of gastric distension
- CPAP/PEEP (positive end-expiratory pressure) are useful and may help avoid airway collapse and maintain oxygen saturations in neonates and infants
- Select anaesthetic equipment with low deadspace volume to reduce the work of breathing

- Monitor all babies for apnoeas after surgery; ex-premature babies are at increased risk until they are 60 weeks post conception
- If oxygen therapy is required, SpO₂ 87-94% is recommended in premature neonates.

CARDIOVASCULAR CONSIDERATIONS

Transitional circulation

With a newborn's first breath, there is a transition from the fetal circulation (gas transfer at the placenta) to the newborn circulation (gas transfer at the lungs). Pulmonary vascular resistance decreases with the first breath by up to 80% (mainly due to the rise in PaO₂ and in part due to the rise in pH and the fall in PaCO₂ at birth). Systemic vascular resistance increases with clamping of the umbilical cord hence exclusion of the low resistance placental bed (see Figure 4).

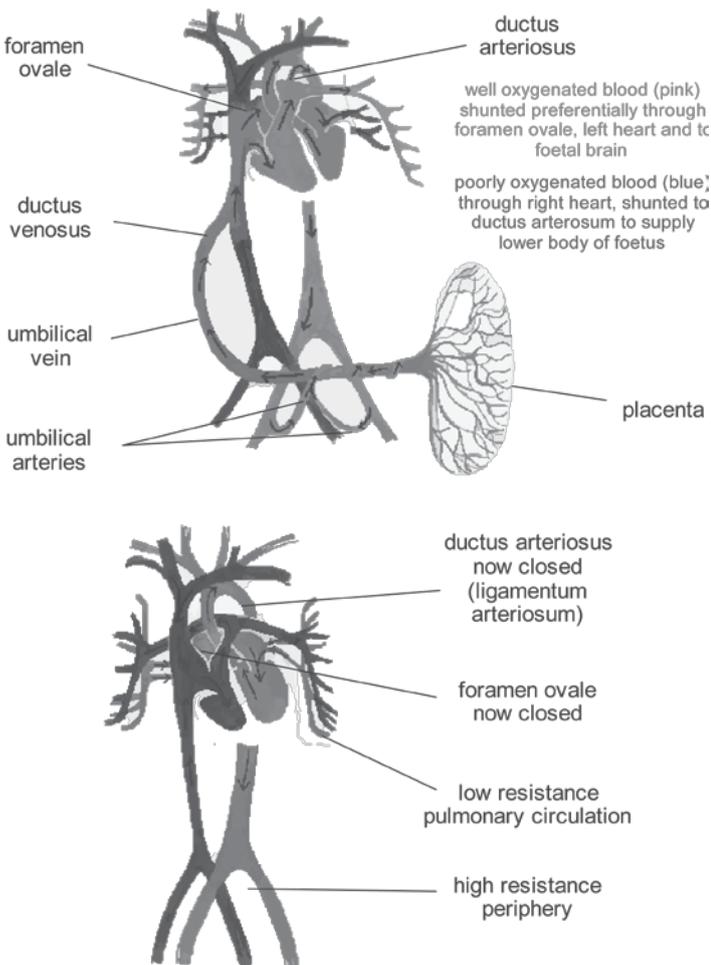


Figure 4. Fetal and neonatal circulation (illustration by Mrs P. Klebe, used with permission)

Venous return from the lungs to the left atrium increases and the pressure gradient reverses across the foramen ovale, which begins to close. The pressure in the pulmonary artery falls, and blood flow through the ductus arteriosus is reversed so that blood flows from the aorta to the pulmonary artery. The ductus arteriosus begins to constrict due to increasing PaO₂ and decreasing levels of prostaglandin E₂ (PGE₂). This is a transitional period. The ductus does not undergo full fibrosis for one month, and the foramen ovale may reopen in the first 5 years of life. Large decreases in systemic vascular resistance or increases in pulmonary vascular resistance due to hypoxia, hypercarbia, sepsis or acidosis in the first few weeks after birth may cause the pulmonary vascular resistance to rise, and the fetal shunts to reopen with right to left shunting; the baby will become very cyanosed as deoxygenated blood flows from the pulmonary artery to the aorta and pulmonary blood flow falls. Worsening hypoxia leads to increased pulmonary vascular resistance, which further amplifies the right to left shunt. This is called persistent pulmonary hypertension of the newborn (PPHN).

Neonatal considerations

At birth the right ventricle is a similar size to the left ventricle, due to the high PVR in fetal life. There is therefore right-sided dominance on the newborn ECG. By two months of age the left ventricle is twice the size of the right, and a left dominant ECG is seen from 4 – 6 months of age. As the heart grows, the PR interval, QRS duration and the QRS size all increase.

The newborn period is a time of rapid growth and development. High tissue oxygen delivery is required for the developing brain and other organs. The cardiac output is therefore relatively high compared to adults (see Table 4).

The ventricles are immature, and less compliant, with a relatively fixed stroke volume (1.5mls.kg⁻¹ at birth), so increase in cardiac output is achieved through an increase in heart rate, rather than an increase in stroke volume as in adults (see table 3). This limits the ability to increase the cardiac output with a fluid challenge in a neonate, and it is easy to push the neonate into pulmonary oedema if too much fluid is given. Bradycardia (most commonly due to hypoxia) will reduce both cardiac output and blood pressure significantly.

In the newborn, vagal tone predominates. Hypoxia, airway manipulation, surgical stimuli and deep halothane anaesthesia are all likely to provoke bradycardia. Hypoxia should always be corrected and a dose of atropine (20mcg.kg⁻¹) should always be drawn up when anaesthetising children. Start CPR if the HR drops below 60 bpm; small doses of adrenaline up to 10 mcg.kg⁻¹ may be required if the heart rate is unresponsive to atropine.

Table 4. Changes in cardiac output with age

Age of child	Cardiac output (ml.kg ⁻¹ .min ⁻¹)
Neonate	300
Adolescent	100
Adult	70-80

Normal blood pressure varies with age, and can be estimated using the formula below (see table 3 for normal values):

$$\text{Systolic BP (50th centile)} = 85 + (\text{age in years} \times 2)$$

$$\text{Systolic BP (5th centile)} = 65 + (\text{age in years} \times 2)$$

As the child grows, the stroke volume rises, the heart rate falls and the systemic vascular resistance rises. The response to volume loading is more predictable from 2 years of age.

Summary – practical implications for the anaesthetist

- Normal values for HR and BP vary with age
- The response to a fluid challenge may be blunted in the neonate or small infant
- It is important to avoid bradycardia. This should be treated rapidly should it occur; the most common cause is hypoxia
- There is a ‘transitional’ circulation at birth, and fetal shunts may reopen in the critically ill neonate

FLUID BALANCE

At birth total body water may be as high as 80%, which gradually decreases to 65% in the adult. Extracellular fluid accounts for 40% of this volume (higher in prematurity), more than in adult patients. Children are particularly prone to dehydration as they have a higher metabolic rate and extracellular fluid turnover, they do not concentrate the urine well to conserve water, and infants also have a higher surface area to weight ratio with relatively higher insensible losses. There is poor compensation for extracellular fluid dehydration as the intracellular compartment is relatively small.

The stress response to surgery may result in hyponatraemia due to the release of antidiuretic hormone (ADH) from the pituitary, over-riding the effect of plasma osmolarity (see chapter on fluids, page 81). The use of hypotonic fluids (such as 0.18% saline with 4% glucose) may exacerbate this and should be avoided. Normal saline or Ringers lactate would normally be chosen for fluid maintenance. Due to the risk of hypoglycaemia, glucose containing solutions should be given to neonates or children below the 3rd centile of weight (for example 0.9% saline with 5% dextrose). Use close monitoring of sodium and glucose levels when giving IV fluids in these patients.

It is important to monitor and replace intraoperative losses meticulously. It is estimated that 10mls.kg⁻¹.hr⁻¹ evaporative losses occur during paediatric laparotomy. Swabs should be weighed and suction fluid losses measured carefully.

Summary – practical implications for the anaesthetist:

- Children are prone to dehydration, and should not be fasted excessively. Allow free clear fluids (water) up to 2 hours before surgery
- Fluid balance should be monitored carefully during surgery
- Children are prone to hyponatraemia in the perioperative period; use only isotonic fluids.

HAEMATOLOGY

At birth 70% of haemoglobin is HbF; this has alpha and gamma haemoglobin chains, and is ideally suited to efficient delivery of oxygen to the tissues in the hypoxic environment in the fetus. However, tissue oxygen delivery by HbF at the higher values of PaO₂ found in the newborn is less efficient. Haemoglobin concentrations, blood volume and cardiac output are relatively high in the newborn to facilitate oxygen delivery to the tissues, and to meet the high metabolic demand for oxygen. Premature neonates may have a low haemoglobin as iron stores are laid down in the final three months of gestation. HbF declines to negligible levels by 6 months of age. Production of haemoglobin HbA₂ increases from birth, and reaches adult levels by 6 months of age; ‘physiological anaemia of infancy’ is seen at 3-6 months as HbF levels decline and HbA₂ levels increase (see Table 5 for normal values).

The relative blood volume in neonates is high, but the absolute volumes are very small. If the absolute volume is 300mls, blood loss of just 60ml is 20% of the circulating volume. Transfuse blood (packed red blood cells) if 20% of blood volume is lost or if the haematocrit falls to less than 25%. Consider clotting factors, platelets and fibrinogen (or use whole blood) if blood losses are more significant. (See the article on ‘major haemorrhage’ for more information, page 195).

THERMOREGULATION

Heat stores in children are small due to their low body mass. Heat is lost rapidly due to high evaporative losses, and poor insulation:

Table 5. Haemoglobin levels and blood volume – normal values

Age group	Blood volume (mls.kg ⁻¹)	Haemoglobin concentration (g.l ⁻¹)
Neonate	90	180-200
Infant	85	90-120
Child	80	110
Adult	70	110

- Large surface area to body weight ratio
- High minute ventilation
- Thin skin
- Poor subcutaneous fat stores.

Neonates are unable to maintain body heat as the thermoregulatory centres are immature, and they cannot shiver or sweat efficiently. They can generate heat through the metabolism of brown fat (non-shivering thermogenesis). Brown fat accounts for about 5% of a neonate’s body weight and is distributed around the scapulae, kidneys, adrenals and mediastinum. Metabolism is neurally mediated (β_3 adrenoreceptors) and generates heat. This is inefficient and increases oxygen consumption. Older children (>3 months) have ineffective shivering due to limited muscle mass. Vasconstriction in children is inefficient.

Children are therefore prone to hypothermia. The consequences of hypothermia include:

- Increased oxygen consumption
- Respiratory depression
- Cardiac arrhythmias
- Coagulopathy
- Acidosis
- Hyperglycaemia
- Immunosuppression
- Prolonged drug metabolism.

Keeping children warm improves outcomes. (See Equipment article for strategies, page 13).

RENAL CONSIDERATIONS

Renal vascular resistance is higher in the newborn and there is a relatively low renal blood flow and glomerular filtration rate. The ability to concentrate urine is poorly developed as

the renal cortical tubules are immature; babies therefore produce large volumes of dilute urine and do not tolerate dehydration well. Sodium is required for normal growth, and sodium is actively conserved. Dietary sodium requirements may be higher in premature neonates due to increased renal sodium losses. Expansion of the plasma fluid volume should be avoided in the first few days of life before the post-natal diuresis has occurred; babies do not require added sodium if they receive IV maintenance fluids in the first few days of life; add sodium should be added to maintenance fluids after the first few days of life. Isotonic fluid should always be used during surgery. Urine output in the newborn is approximately 1-2mls.kg⁻¹.hr⁻¹.

HEPATOBIILIARY CONSIDERATIONS

Neonates have poor liver glycogen stores and are at risk of hypoglycaemia. A 10% dextrose infusion may be required to prevent this. There is an increase in red cell breakdown and a limited ability to handle unconjugated bilirubin, so physiological jaundice is common in the first two weeks of life. Vitamin K dependent clotting factors (II, VII, IX and X) are deficient in the newborn and ideally, vitamin K should be given at birth to prevent haemorrhagic disease of the newborn, particularly in a neonate who requires surgery. Platelet function is reduced and there is a risk of bleeding, particularly in the septic neonate (the platelet count falls in sepsis).

Hepatic metabolism of drugs is reduced in the neonatal period, and may take 3 months to reach full activity. This is particularly relevant to opioids - the half-life of morphine is 6-8 hours in the term neonate compared to 2 hours in infants and older children, so increased dosing intervals should be used and opioids strictly titrated to effect in neonates.

IMMUNE SYSTEM

The neonate is relatively immunosuppressed. Maternal IgG antibodies, which can cross the placenta, fall during the first 6 months. Breast-feeding helps protect against gastrointestinal and respiratory tract infections.

NERVOUS SYSTEM

The central nervous system is immature at birth and cerebral myelination continues for up to 3 years. The process of

Table 6. Central nervous system – normal values

Patient group	Cerebral blood flow (ml.100g ⁻¹ .min ⁻¹)	Cerebral metabolic rate for oxygen (CMRO ₂) (ml.100g ⁻¹ .min ⁻¹)	CSF volume (ml.kg ⁻¹)	CSF volume (ml.kg ⁻¹)
Newborn	50		4	<2
Child	100	5.8		
Adult	50	3.5	2	7-15

Table 7. Comparison of volatile agents in children

Halothane	Sevoflurane	Isoflurane
MAC (%)	MAC (%)	MAC (%)
Infant: 0.9	Infant: 3.3	Infant: 1.6
Child: 1.2	Child: 2.5	Child: 1.9
Adult: 0.75	Adult: 1.7	Adult: 1.2
Sweet smelling, well-tolerated	Non-irritant	Pungent, irritant, difficult to use for inhalational induction
High lipid solubility, slow onset/offset anaesthesia; induction rate increased if used with nitrous oxide.	Low lipid solubility, rapid onset/offset of anaesthesia	Low lipid solubility, rapid onset/offset of anaesthesia
Arrhythmias common, myocardial depression common	Good haemodynamic stability; blood pressure may fall due to fall in systemic vascular resistance	Good haemodynamic stability; blood pressure may fall due to fall in systemic vascular resistance
Respiratory depression seen	Respiratory despression common	Respiratory depression seen
Inexpensive	Expensive, although costs coming down	Expensive, although costs coming down
Hepatic metabolism; halothane hepatitis seen rarely (repeat anaesthetics)	Not highly metabolised	Low hepatic metabolism
Compatible with draw-over systems; not all modern anaesthesia systems include a halothane vaporiser	Compatible with modern anaesthesia systems; not all draw-over systems include a sevoflurane vaporiser	Compatible with modern anaesthesia systems; can be used in a halothane vaporiser

central nervous system maturation continues during fetal and neonatal development, but the ability to feel pain is well developed even before birth. Pain in neonates is associated with behavioural changes such as tachycardia, hypertension, grimacing, crying and restlessness; pain scores in children take these behavioural features into account. There is concern about the effect of anaesthetics on the developing brain, and unnecessary anaesthetics should be avoided in infancy.

Cerebral blood flow is high in childhood, and intracranial pressure (ICP) is lower in the newborn (see table 6 for normal values). Rises in intracranial pressure are well-compensated in infancy, in part by expansion of sutures and bulging of fontanelles, and intracranial pressure can be estimated by palpation of the fontanelles. Ossification of the fontanelles is complete by about 18 months.

PHARMACOLOGICAL CONSIDERATIONS

Volatile anaesthetics

Neonates are more sensitive to volatile agents than older children, and the minimum alveolar concentration (MAC) values are decreased in neonates but increased by up to 30% in infants and children compared to adults. Halothane and sevoflurane remain the agents most popular for inhalational induction of anaesthesia (see table 7 for comparison between volatile agents).

Sedatives and hypnotics

Children are particularly sensitive to sedative and hypnotic drugs such as barbiturates and benzodiazepines due to the immature blood brain barrier and reduced drug metabolism/excretion; these drugs should be used with caution, in weight

appropriate doses, titrated according to effect. Propofol infusions may be used in children older than 3 years. See article 'Paediatric procedural sedation' on page 77).

Muscle relaxants

The volume of distribution of water soluble drugs is higher at birth due to the increased extracellular fluid volume. (This is true for all water soluble drugs, not only muscle relaxants). This explains the need for a higher dose of suxamethonium in infants relative to adults ($2\text{mg}\cdot\text{kg}^{-1}$ vs $1\text{mg}\cdot\text{kg}^{-1}$). A second dose of suxamethonium may provoke bradycardia due to the high parasympathetic tone in infants; atropine should always be available.

Neonates and infants are more sensitive to non-depolarizing neuromuscular blocking drugs. A normal loading dose is given but subsequent doses should be reduced.

Analgesics and local anaesthetics

Drug doses need to be reduced due to immature metabolism, and in the case of local anaesthetics, lower levels of plasma binding proteins (serum albumin and alpha-1 acid glycoprotein). Take extra care not to exceed maximum doses or dose intervals of analgesics such as paracetamol, ibuprofen,

opioids or local anaesthetics. (This topic is considered in more detail on page 72).

ACKNOWLEDGEMENTS

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FURTHER READING

1. Berg S. Chapter 33: Neonatal/infant physiology. In: Allman KG, Wilson IH, eds. Oxford handbook of Anaesthesia (3rd edition) Oxford: Oxford University Press, 2011: 800-809.
2. Jewitt H. Chapter 2.8: Principles of Neonatal Physiology. In: Allman KG, Wilson IH, eds. Dr Podcast Scripts for the final FRCA (1st edition) Cambridge: Cambridge University Press, 2011: 325-331.
3. Macfarlane F. Paediatric anatomy and physiology and the basics of paediatric anaesthesia. *World Federation of Societies of Anaesthesiologists* 2005; Tutorial of the week number 7.
4. Brown K. The application of basic science to practical paediatric anaesthesia. *Update in Anaesthesia* 2000; 11: 73-77. Available from: http://www.wfsahq.org/components/com_virtual_library/media/f28df37342cf2cb1f6390ccf0e49453d-7a87e9e9c2b627962e0aeb1ae752e45-The-Application-of-Basic-Science-to-Practical-Paediatric-Ana.pdf

Equipment in paediatric anaesthesia

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AIRWAY ADJUNCTS

Oropharyngeal ("Guedel") airways

These are universally popular in children as they are easy to use and fast to insert. They can be used to supplement airway manoeuvres such as jaw-thrust and chin lift during resuscitation, induction, and emergence from anaesthesia. The airway should only be inserted when the child is deeply anaesthetised. If the child is too light the airway will not be tolerated, potentially leading to coughing, gagging, expulsion of the airway, or laryngospasm.

Oropharyngeal airways in children are best inserted correctly oriented. You should take care to avoid damage to the delicate soft palate in young children, and to the posterior pharyngeal wall if a relatively large airway is used. They are less likely to cause trauma on insertion than a nasopharyngeal airway.

Nasopharyngeal airways

These are particularly useful in children with a tendency towards upper airway obstruction or a reduced level of consciousness. They are better tolerated than an oropharyngeal airway. To insert:

- Lubricate with gel to minimise trauma to the turbinates
- Avoid local anaesthetic preparations because these can alter pharyngeal muscle tone and impair airway patency
- Insert gently along the floor of the nose
- Take care not to exert too much pressure at the post nasal space where the adenoids may be damaged.

Nasopharyngeal airways are usually used during recovery rather than during anaesthesia. A common indication is for the child at risk of upper airway obstruction after cleft palate repair. To prevent damage to the suture line during insertion, railroad the airway over a smaller nasogastric tube (confirm that this is in the stomach first), or allow the surgeon to insert the nasopharyngeal airway under direct vision.

Facemasks

A plastic facemask with an inflatable cuff/rim gives a better seal on the face, and is less frightening to the child than the traditional black rubber facemask. The correct size of facemask is one that reaches from the cleft of the chin

SUMMARY

Differences in anatomy and physiology between children and adults mean that much of the equipment needed to anaesthetise children needs to be specially adapted.



Figure 1. Oropharyngeal airways come in a range of sizes suitable for premature neonates up to adults. To find the correct size, hold with the flange in line with the middle of the incisors. The tip should just reach the angle of the child's jaw

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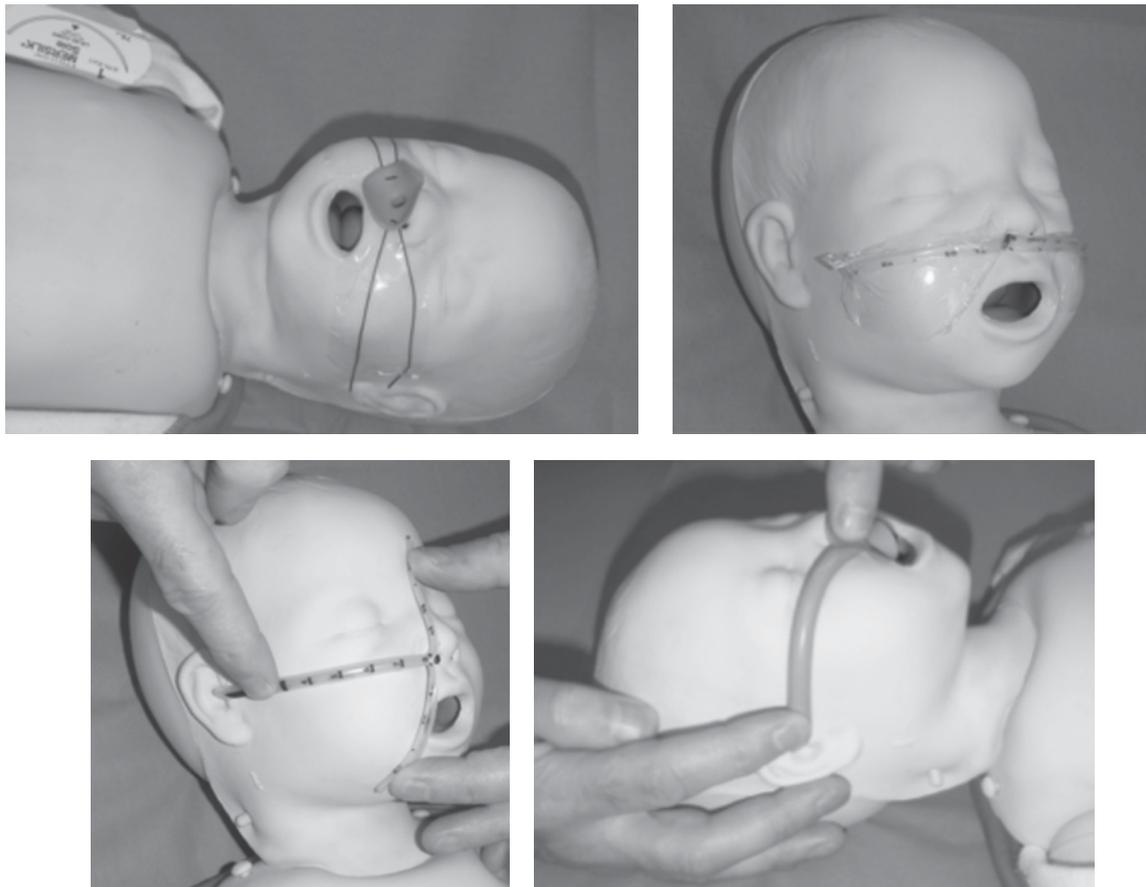


Figure 2. The correct diameter of the nasopharyngeal airway is the same as the size of the endotracheal tube for that child (see Table 2). Estimate the length by the distance from the nostril to the tragus of the ear as show. An uncuffed endotracheal tube makes a satisfactory nasopharyngeal airway. Either attach a standard connector (to stop the tube from advancing and becoming 'lost' in the nose); or split the tube to facilitate secure taping as shown. Sutures may be tied around the tube to secure the airway as shown.

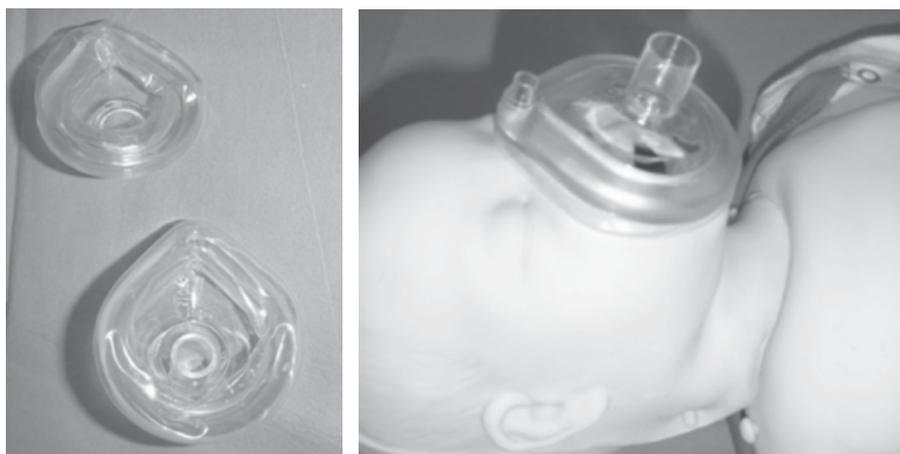


Figure 3. Clear plastic facemasks

to the bridge of the nose, but does not cover the eyes. For small babies, round masks are available, or turn a shaped mask 'upside down' for an acceptable seal.

Laryngeal mask airways (LMAs)

The LMA is the most widely available example of a supraglottic airway device. They are suitable to use during maintenance

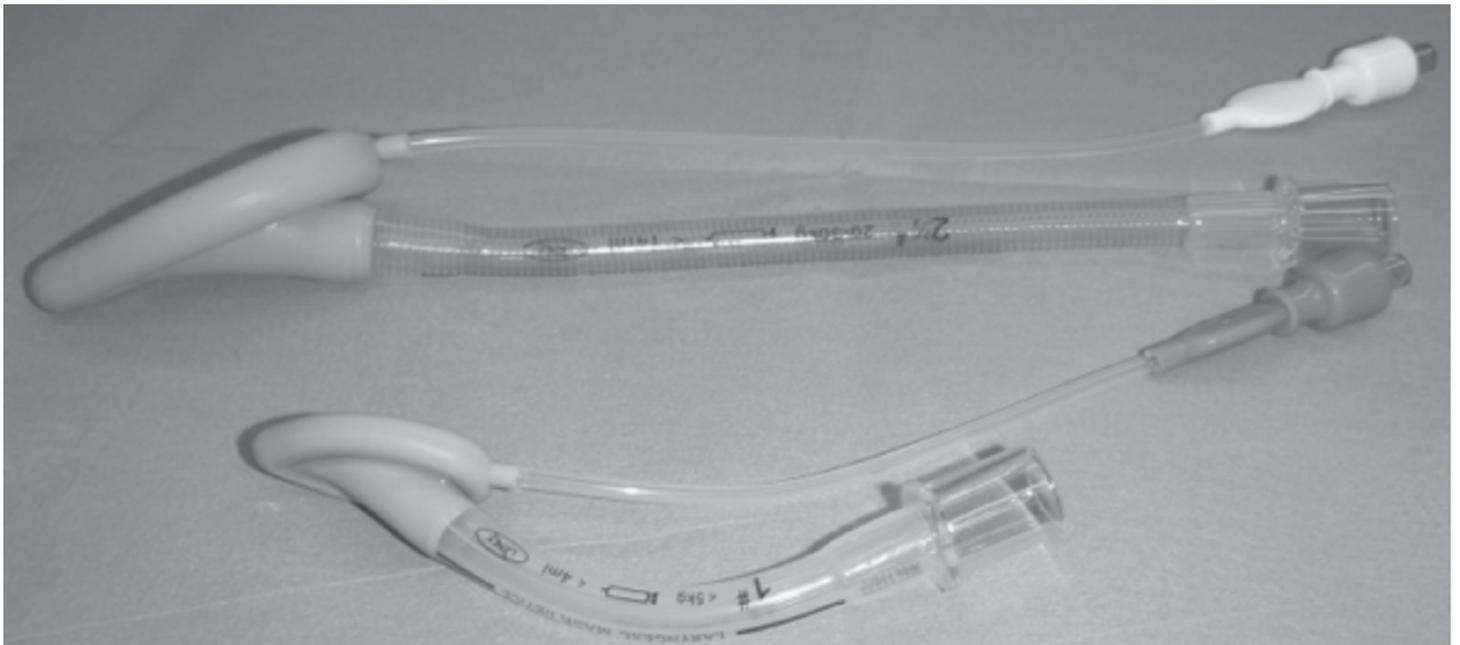


Figure 4. LMAs are available in a range of sizes appropriate for babies up to adults. They may be either single-use or designed for repeat resterilisation. Rigid versus flexible stems can be easier to insert but more frequently displaced by heavy drapes or when close to the surgical field

of anaesthesia in cases which you would otherwise manage by holding a facemask (low aspiration risk), to free the anaesthetist's hands for other tasks.

The size 1 LMA is less useful for routine anaesthesia because:

- It is designed for children <5kg in whom intubation and ventilation are generally preferred
- It tends to dislodge, in which case it occludes rather than maintains the airway
- Coughing and laryngospasm on insertion are more common with smaller LMAs.

It may still be useful to rescue a difficult airway, even in a small baby (see Difficult airway article, page 116).

Table 1. Manufacturers' guide to LMA sizes

Weight (kg)	LMA size	Maximum cuff volume (mls)
<5	1	4
5-10	1 ½	7
10-20	2	10
20-30	2 ½	14
30-50	3	20

In adults it is common to mechanically ventilate the lungs through the LMA. In children there is a higher risk of gastric insufflation, so this is not widely practiced.

Serious airway damage from the LMA cuff has not been reported, but be aware that the cuff pressure exceeds that of a tracheal tube. Capillary perfusion pressure in children is lower than in adults, thus the potential for injury is probably greater, even if the cuff is not inflated above the recommended maximum volume (see Table 1). Be even more cautious in longer procedures, as lingual oedema has been reported after LMAs have been in for a long time.

Avoid overinflation of the LMA cuff:

- Lower cuff pressures tend to give a better seal
- There is less risk of pressure injury to tissues.

Opinions vary as to when to remove the LMA in children. The choice is either deeply anaesthetized ("asleep") or fully awake; between these extremes, you risk precipitating laryngospasm. Some suggest that the best time to remove the LMA is when the pharyngeal reflexes return and the child spits the airway out.

Use in difficult intubation – see article page 116

The intubating LMA is not made below a size 3, therefore is not suitable for young children.

Laryngoscopes

The epiglottis is long and U-shaped in infants and small children. This is advantageous when breast-feeding but requires a different approach for intubation. A curved blade placed in the vallecula will not provide a reliable view of the glottis. Straight blades designed to lift the epiglottis along with the soft tissues at the base of the tongue give an easier view.

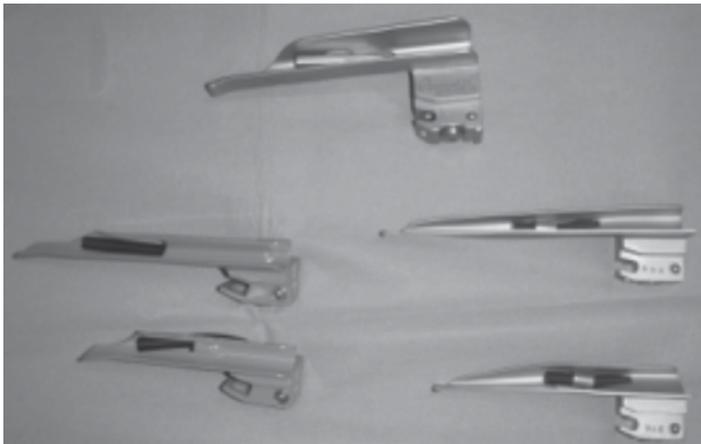


Figure 5. The most popular straight blades are [left to right] the Miller, the Robertshaw and the Oxford [top in (a)]. Many paediatric anaesthetists use a MacIntosh (size 3) curved blade in all children over 1 year old. Others prefer a straight blade until 5 years old. No one design has been shown to be truly superior.

The international standards organization has a size mark for laryngoscopes from 5 (largest) to 000 (smallest – too small even for tiny premature babies). A size 1 blade is the correct size for a normal term baby, and a size 0 for premature neonates who weigh less than 3kg.

It is possible to intubate with a laryngoscope that is slightly too big, and rarely possible with one too small. MacIntosh considered smaller sizes of his original (size 3) blade unnecessary, as the distal portion of the size 3 blade is straight and therefore suitable for smaller children. He was correct!

Many disposable laryngoscope blades exist. Some are of high quality but this cannot be assumed, for example some have

broken during clinically realistic force tests. Manufacturers label these blades Miller, Robertshaw etc., but they are often not exact copies of the blades whose name they bear. Both laryngoscopic view and illumination may vary.

Endotracheal Tubes (ETT)

The black marks used for guidance during laryngoscopy vary widely between manufacturers and between tube types. Some are meant to indicate the appropriate position at the cords. Others simply make the tip easier to see. The only reliable advice that can be given on ETT markings is to ignore everything except the actual distance markings in centimetres.

A 3.5mm uncuffed ETT is suitable for the normal term neonate. Use the formulae below to guide ETT selection in older patients (Table 2). There should be a small leak around the ETT to avoid excessive pressure on delicate mucosa. If ventilation is compromised, the leak is too large. The nose will accommodate the same size ETT as the larynx prior to puberty.

A neonate's trachea may be less than 4cm in length. The ETT should sit at least 1cm above the carina, which is anteriorly inclined and more symmetrical than in adults. Small flexion/extension movements may cause accidental endobronchial intubation or extubation respectively. You must be meticulous in placing and securing the ETT.

Some children receiving intensive care benefit from a cuffed tracheal tube. Most paediatric anaesthetists prefer uncuffed tubes for routine anaesthesia in children up to 8 – 10 years old.

Reasons to choose an uncuffed tube:

- Larger internal diameter tube can be used
- Confusing tube markings and long cuff shoulders have led to frequent endobronchial intubation with traditional cuffed tubes. The microcuff tube® avoids these problems
- Overinflation of endotracheal tube cuffs risks the potential but rare complication of tracheal stenosis

Reasons to choose a cuffed tube:

- Fewer intubation attempts with different sized tubes
- Reduced leak – more important in ICU, also in anaesthesia as better paediatric circle systems are developed and use of low flow anaesthesia in children increases

There is no good evidence that either cuffed or non-cuffed tracheal tubes are better.

BREATHING SYSTEMS

Semi-open (see article on drawover anaesthesia, p23)

Semi-closed

The T-piece is the most popular system for anaesthetising small children in UK with many advantages:

- Lightweight
- Compact
- The bag is easily visible whilst observing the patient
- Simple to use for spontaneous or controlled ventilation
- No valves
- Low internal resistance
- Low compression volume (i.e. the operator has a good 'feel' for the lung compliance).

Jackson-Reece modified the system by adding an open-ended bag. This bag has several advantages:

- Respiratory monitor during spontaneous ventilation
- Easy to convert to manual controlled ventilation
- Easy to apply PEEP.

In skilled hands the T-piece is the best system for resuscitation. The application of CPAP / PEEP splints the upper airway open and so improves the efficacy and ease of ventilation.



Figure 6. T piece attached to oxygen cylinder. The same connection will fit the outlet of an oxygen concentrator. Alternatively, the T piece's green tubing connects directly to an anaesthetic machine's common gas outlet. A T piece requires gas flow.

However, in less skilled hands, the T-piece may result in

- Inadequate ventilation (requires practice to use)
- Gastric insufflation
- High peak inflation pressures
- Inadvertent barotrauma.

It is customary to use adult breathing systems in children > 20 – 30kg. To prevent hypercarbia during controlled ventilation, the fresh gas flow in ml can be predicted by the formula $(1000 + (200 \times \text{kg}))$ (minimum of at least 3 l/minute), which is approximately 1.5x the minute volume. During spontaneous breathing in young children, there is no end expiratory pause and the system becomes less efficient; twice the fresh gas flow required for controlled ventilation should be used (i.e. about 3 x the minute volume).



Figure 7. Humphrey ADE. Newer versions are, in fact, AE systems; there is no option for a Mapleson D. The E system (lever down) is a T-piece; it has no bag and no valve and is used for controlled ventilation (fresh-gas flow as for the T-piece, above). The A system (lever up) has the configuration of a parallel Lack circuit. This is ideal for children breathing spontaneously because of the low resistance of both the smooth inner bore of the tubing and the expiratory valve. A flow rate of 3 l/minute is sufficient for most children before adolescence.

Closed

Early circle systems imposed a high work of breathing due to the resistance of one-way valves. This is much less of a problem with modern circles but other issues remain:

- A leak around the tracheal tube means that low flows cannot be used efficiently (solve by using cuffed tracheal tubes)
- The volume of the breathing system is large compared with minute volume, giving longer equilibration times than in adults
- The tubing has a significant compression volume, which may alter the characteristics of ventilation during positive pressure ventilation. Avoid by using stiffer tubes and smaller bellows
- Paediatric circle systems do not maintain the temperature or humidity of inspired gases adequately.

IV ACCESS

Administering fluids

Standard blood and fluid administration sets are suitable for children. A burette should be used for small children (<10kg) so that the volume of fluid can be measured accurately and to avoid giving excessive volumes of fluid.

In-line fluid warmers are of great benefit, but they add considerable deadspace into the system, and require electricity, and costly single-use disposables. Water baths are not commonly used to warm IV fluids because contamination with *Pseudomonas* species can cause serious infection. Fluids may be stored in a warming cabinet or other warm place prior to administration. Some anaesthetists place the sealed bag of IV fluid on top of the monitor as this becomes warm during use. This is only partially effective, but makes use of a readily available source of heat!

Peripheral cannulae and flushing

Young children more commonly have a patent foramen ovale. This means that bubbles administered IV may embolise to the systemic circulation rather than the lung. Remove all air bubbles from the fluid administration system. It is difficult to give guidelines about the amount of air that poses a danger because speed of injection is as important as volume. Deaths have occurred after as little as 0.5ml.kg⁻¹ injected rapidly.

The 'dead-space' of injection ports and cannulae can be significant. For example, neonates have been paralysed by the quantity of suxamethonium present when the hub of the cannula was flushed post-operatively. Anaesthetists must flush the cannula carefully with saline after injecting drugs,

particularly if an IV extension has been used. Avoid flushing with heparinised saline in neonates. Heparinised saline contains 10 iu.ml⁻¹ of heparin; 10ml given to a 3kg neonate is 33 iu.kg⁻¹ and this could easily affect clotting.

Central venous catheters

These are smaller versions of adult catheters, with up to 4 lumens.

Sites for central venous access

Femoral venous access is more popular in children than adults. The tip of the catheter is the most likely place for thrombus formation, so choose the length of the catheter so that the tip does not lie at the junction of the renal veins with the inferior vena cava (IVC). Either short lines (catheter tip in the iliac vein or lower IVC) or long lines (tip just below the diaphragm) are appropriate. Central venous pressure monitored from the latter position is as accurate as from the superior vena cava (SVC). Confirm the position of longer femoral lines on X-ray if available, because the line may pass into the contra-lateral femoral vein, the renal vein or the epidural venous plexus. In children <2 years old, the J-portion of J wires may exceed the dimensions of the central vein which will make the wire difficult to thread.

The umbilical vessels are easily accessible during the first 24 hours of life (and less easily so for a further 3 days). Placing the tip of an umbilical venous catheter in the correct position is easier if you adapt the catheter tip to be a unipolar electrode and monitor the ECG signal until you obtain a characteristic atrial ECG. Umbilical arterial lines have been linked with the development of necrotising enterocolitis, although this is not proven. Umbilical venous catheters are particularly prone to thrombosis, which may lead to portal hypertension. They should be replaced as soon as alternative access is practical.

Peripherally inserted central catheters (PICC lines)

These are available as small as 27g and provide access for drugs or parenteral nutrition. Smaller lines cannot be used for pressure monitoring or sampling. Hypertonic drugs (e.g. thiopentone) or drug precipitates (e.g. thiopentone/ suxamethonium) are likely to block these lines. Flush all medicines in immediately after administration.

Intra-osseous needles

There are discussed in a separate article (see page 242)

DEFIBRILLATORS

There are no specific defibrillators designed for children. Many standard defibrillators have paediatric paddles similar to those shown in Figure 8: the adult paddle slides off the smaller paediatric paddle. Paddles must be positioned as far apart as possible to reduce the chances of arcing. Use gel pads (not

shown) to reduce impedance between paddle and patient as for adults. Self-adhesive pads may be available (see Figure 8).

The appropriate size of paddle is the largest pad that will fit the child's chest, as this will reduce both transthoracic impedance and the potential for chest burns. These pads are often labelled with a recommended weight range for standard hospital controlled dose defibrillators, or with an age range for automatic external defibrillators (AEDs).

The appropriate energy levels for children are different from those for adults (see Paediatric Resuscitation p264). 'Advisory' defibrillators and AEDs prompt the user down adult Advanced Life Support (ALS) algorithms and so are not recommended in infants < 1 year old, although they have been used successfully in young children when there has been no alternative. Paediatric pads, if used on an AED, will automatically deliver a lower energy, which varies between defibrillator models. Adult

pads on an AED will deliver up to 360J.

There is evidence that biphasic waveforms are more effective for the treatment of ventricular fibrillation. Advantages seen in adult practice include

- Less time needed to charge
- Improved first shock success rate (90% compared with 60%)
- Less myocardial injury
- Decreased incidence of skin burns
- Increased battery life of the defibrillator.

Current paediatric recommendations are to select the same energy ($4J.kg^{-1}$ in cardiac arrest) for a monophasic or biphasic defibrillator (see resuscitation articles p267 and 270.)

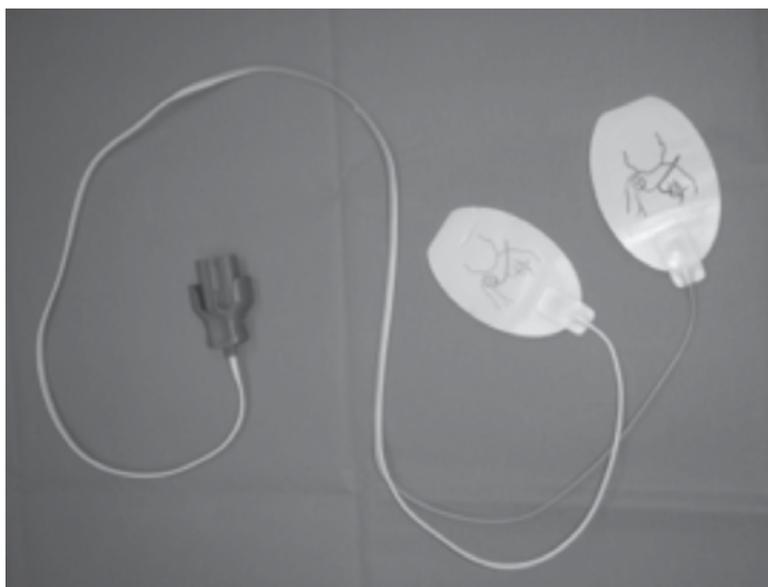


Figure 8. Paediatric defibrillator pads and paddle positions.

TEMPERATURE CONTROL

Good temperature control is vital in paediatric anaesthesia (see basic sciences article p4). You must monitor temperature during active warming.

Warming methods include:

- **Forced air heating blankets** e.g. the Bair Hugger. These devices deliver warm air up to 43°C for active warming. They can also blow room temperature air to cool patients who are pyrexial
- **Simple warming blankets** reduce conductive losses to the table and are cheap and effective although not as efficient
- **Overhead radiant heaters** are also cheap and effective. Pay strict attention to the safe minimum height of radiant heater above the patient. Patient signs of systemic overheating will be late. Local overheating presents as burns. Radiant heaters increase insensible water loss in neonates
- Prevent heat loss by **insulation** when active heating is impractical or unavailable. Almost any insulating material may be used e.g. clean blanket or clothing, cotton wool, tinfoil or bubble wrap
- **Raised operating theatre temperature** for babies or small children. You will need to compromise between the thermo neutral temperature for the anaesthetised patient (34°C for premature neonates, 33°C for term neonates reducing through childhood to 30°C at adolescence) and a comfortable working temperature for staff (up to 25°C). One solution is to have the temperature relatively high during induction of anaesthesia, and then reduce it once the patient is draped and use a forced air warmer if available to maintain a 'microclimate' during surgery
- In theory **circle systems** warm and humidify inspired gases. In practice these effects are small. Some circle systems have additional heaters to aid gas warming. HME (heat moisture exchange) filters help
- **Heated humidifiers** may be added to any breathing circuit, and will help warm the patient by warming airway gases. Monitor gas temperature at the patient end to avoid airway burns
- **Warm fluids** – as above.

MONITORING

Standard monitors for a child are the same as for an adult. It is acceptable to complete the attachment of patient monitors after induction of anaesthesia to prevent a child becoming distressed as long as there is no compromise to safety.

Oesophageal or precordial stethoscope

This low-technology device offers continuous, immediate information on breath and heart sounds without the lag times associated with oximetry and capnography.

Pulse oximeter

Oximetry probes need to be matched to the size of the patient. Too large a probe will give a poor reading due to light scatter and easily falls off. When you choose the site carefully, you can use adult sized probes for younger children e.g. an adult ear probe can be attached to a digit. These ear probes have spring closures, which may produce sufficient pressure to cause ischaemia in a child's finger. Wrap-around probes are available and may be used on digits, or in small babies wrapped around a foot or hand. They have no risk of pressure necrosis but do produce heat. Burns have occurred in children with low peripheral perfusion e.g. during cardiopulmonary bypass.

The absorption spectra of foetal and adult haemoglobin are similar, and at 660nm and 910nm in a basic pulse oximeter the device is accurate in detecting desaturation of foetal haemoglobin.

Problems and pitfalls

- Pulse oximeters are poor indicators of hyperoxia due to the shape of the oxyhaemoglobin dissociation curve. Whilst the pathogenesis of retinopathy of prematurity is multifactorial, it is important to avoid hyperoxia in at risk babies (particularly premature neonates)
- Pulse oximeters may be inaccurate in the presence of certain pigments: methylene blue gives a falsely low reading. Bilirubin does not affect pulse oximetry
- Potential inaccuracies in children with congenital heart disease: the oximeter derives arterial saturation by subtracting the 'fixed absorbance' part of the signal. In tricuspid regurgitation, the monitor may detect a pulsatile venous component of the signal, and display the venous saturation
- In infants with a physiological shunt through a patent ductus arteriosus, the pulse saturation will vary between the right arm (pre-ductal) and other limbs (post-ductal). The pre-ductal blood supplies the cerebral circulation, so measure saturation on the right arm as a minimum.

Phone app based pulse oximeters are appearing on the market. All necessary hardware is incorporated into the finger probe with a connection to a mobile phone where the phone app provides the software and display. Some of the probes now have FDA approval, but experience of their use is limited at present.

Apnoea monitors

These are frequently requested by paediatric anaesthetists to monitor babies <6 months old after anaesthesia. They are designed to alarm after 15 or 20 seconds of apnoea. They detect breathing in one of two ways:

- By changes in impedance measured through ECG electrodes on the chest
- By a change in pressure detected by a sealed mattress or a small sensor on the abdomen as shown in Figure 8 (abdominal excursions move a fine foil strip in the monitor) (More common).

Apnoea monitors are cheap and non-invasive but have some disadvantages:

- Prone to false positive alarms, and to interference from mobile phones
- Will not detect airway obstruction, because movement of the chest and abdomen still occurs
- Cardiac pulsation may trigger the monitor, even if the child is apnoeic.

These monitors should be used as an adjunct to high dependency nursing care, not a substitute for it.

Electrocardiography (ECG)

As children do not usually have ischaemic heart disease, the ECG is used mainly as a heart rate and rhythm monitor and rarely to detect ischaemia. At birth, the right ventricle is as thick as the left. This manifests as right axis deviation and right ventricular hypertrophy on the ECG (dominant R wave in V1, deep S wave V5 & V6, inverted T wave V1-4). With growth, the left ventricle becomes larger than the right. By age



Figure 10. An apnoea monitor for a baby

2-5 years, the axis is within the adult range. T wave inversion reverses by 10 years old when the ECG becomes similar to that of an adult.

Lead II is usually the best choice in all ages as the prominent P wave aids rhythm recognition. A common error is that large T waves may be read as QRS complexes by the monitor causing double counting of the rate.

Blood pressure measurement

Non-invasive blood pressure measurement

All non-invasive blood pressure machines that measure from a cuff are automated oscillotonometers. The most important considerations are the width and length of the bladder used in the cuff (see Table 3).

Table 3. Determining the appropriate size of a blood pressure cuff. If the cuff is too small, the reading will be artificially high; the reverse is true if the cuff is too big. Common Error: generally, oscillotonometers over-read lower pressures, i.e. they over-read values within the normal neonatal range.

BP cuff bladder width	Should cover approximately 2/3 of the upper arm	Neonate 3cm Baby 5cm Small child 6cm Large child 9cm
BP cuff bladder length	Minimum 40% of the circumference of the upper limb; $\geq 80\%$ is ideal	

The machine settings for adult, paediatric or neonatal modes usually control only the maximum inflation pressure. This is to avoid excessive pressure and the risk of bruising or trauma in babies or small children if exposed to the values in a hypertensive adult.

Direct arterial blood pressure measurement has the same principles as in adults. If using an umbilical arterial catheter in neonates, which is relatively long and thin, inspect the trace for signs of overdamping.

Safety point: take care with manual flushing. Meticulously avoid bubbles, and note that a brisk 1ml flush in a neonate is sufficient to push bubbles or thrombus from the radial artery back as far as the vertebral artery.

Peripheral nerve stimulators

Peripheral nerve stimulators may be used in children for monitoring neuromuscular blockade, or localization of peripheral nerve. The response of the paediatric neuromuscular junction to neuromuscular blockade is similar to that of

adults. In neonates, the immature junction is sensitive to non-depolarising muscle relaxants, with reduced prejunctional reserves of acetylcholine.

CO₂ monitoring/end-tidal agent monitoring/oxygen monitor

End-tidal gas measurements are unreliable in babies. The tidal volume is small compared with equipment dead space volume, fresh gas flow, and leak around the endotracheal tube. Small tidal volume and rapid respiratory rate confound the monitor's real-time analysis.

You should still obtain some CO₂ tracing if you are careful to minimise equipment deadspace, but with a large and variable A-a gradient.

CONCLUSION

Consideration of the anatomical and physiological differences between paediatric and adult patients makes it easier to select the correct equipment to anaesthetise all patients safely. Availability of the appropriate equipment may be the more difficult problem.

Paediatric drawover anaesthesia

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INTRODUCTION

The drawover system provides anaesthesia without requiring compressed gases and is the most commonly used method for delivering anaesthesia wherever there is not a reliable supply of compressed gases.

The essential components of the drawover system are

- A vaporiser
- A self-inflating bag or Oxford inflating bellows
- Two valves.

BASIC PRINCIPLES OF USE

Air is the carrier gas, which can be enriched with oxygen if available. This is drawn by the patient's inspiratory effort through the vaporiser into the patient's lungs via a non-rebreathing valve. Drawover is a low pressure system and without the one way valves gas flow could occur in any direction. The valves ensure flow of air and oxygen through the vaporiser to the patient.

One of the earliest systems consisted of:

- EMO (Epstein-Mackintosh-Oxford vaporiser

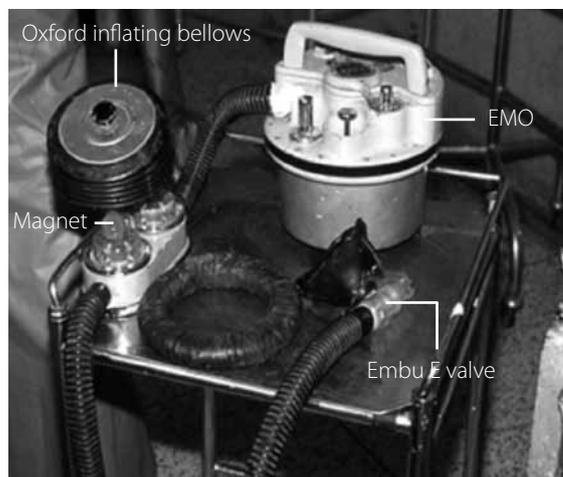


Figure 1. A basic drawover system incorporating an EMO vaporiser

designed specifically for ether,

- Oxford inflating bellows with two one way valves
- Heidbrink valve at the patient end.

Over the years the Ambu E valve was introduced to replace the Heidbrink valve. This enables both spontaneous and assisted ventilation but makes the second valve on the Oxford inflating bellows (OIB) redundant. There is a magnet which sits over the second valve to disable it when an Ambu E valve is being used.

The drawover system is very robust, portable, compact and easy to maintain and as it does not rely on a supply of compressed gases it is not only popular in developing countries but also with the military for battlefield anaesthesia.

The vaporisers used in drawover anaesthesia have a very low resistance. During anaesthesia the gas flow is intermittent (during inspiration with spontaneous ventilation and during expiration with assisted ventilation). The volume of air passing through the vaporiser is determined by the patient's tidal volume and respiratory rate. There may be huge variations depending on the respiratory effort and size of the patient. The dialled concentrations remain very accurate despite these massive variations.

The first drawover vaporisers were designed for ether. Characteristics of ether include:

- Low boiling point - 34°C
- High saturated vapour pressure - 56.6kPa
- Blood gas solubility coefficient (12) which is much greater than most commonly used volatiles like sevoflurane (0.9) and isoflurane (1.2)
- Both induction and wake up are slow
- Atropine is required in all patients due to the excessive salivation produced

Summary

The drawover system provides anaesthesia without requiring compressed gas. It is very robust, portable, compact and easy to maintain.

Even though it is challenging anaesthetising small babies with minimal equipment, the drawover system can provide a safe, robust, portable and cost effective system for anaesthetising babies and children.

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- Respiratory stimulant
- Sympathomimetic
- Causes bronchodilatation
- Some analgesic effect at low doses
- Potentiates the effect of muscle relaxants
- Tends to increase blood glucose concentration, and to relax uterine muscle
- Gas induction is very slow as it is also a respiratory irritant. Increase the concentration by 1% every 6-8 breaths up to 10% and then increase it by 2.5% every 6-8 breaths up to 20%
- Maintenance concentration is 6-8% with spontaneous breathing and 2-4% if the patient is being ventilated.

Although ether was removed from the WHO essential drugs list in 2012, it is a useful and safe volatile anaesthetic especially in areas where oxygen is scarce or where there is no trained anaesthetic provider. There is a risk of explosion which is minimised when using only air as the carrier gas and in remote locations diathermy is rarely available. If diathermy is being used a safe distance of 20cm between the expired port and the diathermy should be maintained. Ether is heavier than air and a scavenging tube can be fixed to the expiratory port and dropped to the ground to minimize the amount ether around the surgical site.

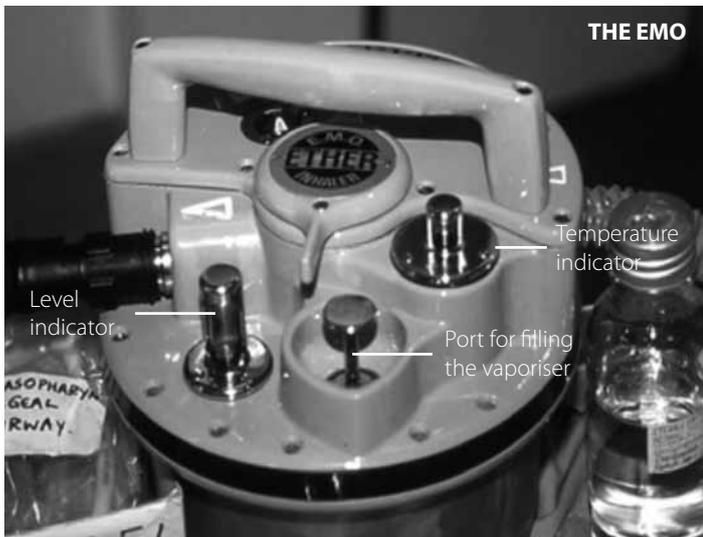


Figure 2. The EMO vaporiser

As such high concentrations are required to maintain anaesthesia, the EMO and “Afy” - the two drawover vaporisers designed for ether - are very bulky. The EMO is thermally buffered with a 1500ml water jacket and compensated with a small ether bellows and requires no keyed filling. The resistance through the EMO is only 0.5cm of water (= 0.049kPa) unlike

a TEC 3 or TEC 4 vaporiser which has a resistance of 21-29cm of water (= 2.0–2.84kPa). It has cloth wicks which still contain 200mls of ether when the level indicator is empty. If the EMO is overfilled the wicks will be submerged and the output will decrease. It has a temperature indicator which is red when above 30°C, aluminium at less than 10°C and black at the optimal working temperature 10°C to 30°C. It produces very accurate concentrations with variable flow rates but loses precision with continuous flow and with extremely low flow rates.

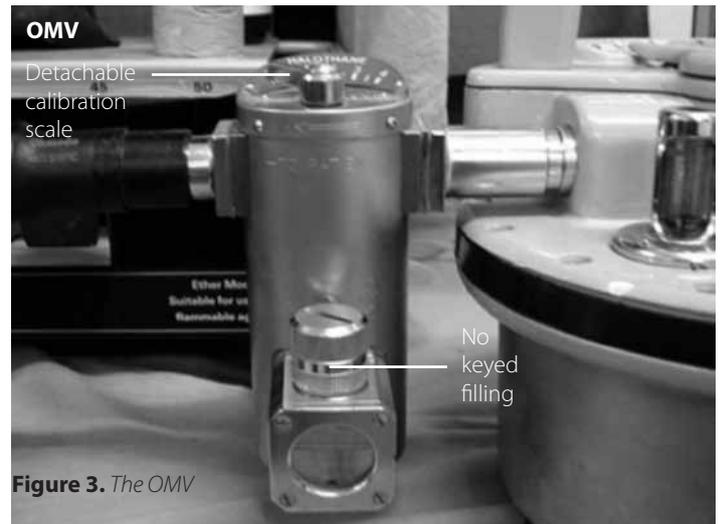


Figure 3. The OMV

A version of the Oxford miniature vaporiser (OMV) is used as part of the Triservice apparatus. Although the carrier gas flows from right to left in most drawover apparatus, in the triservice version of the OMV carrier gas flows from left to right. It is very compact with a capacity for 50ml of anaesthetic agent. It has a metal wick and is not thermally compensated but only thermally buffered with an ethylene glycol jacket. In the original design the downstream vaporiser was filled with trichloroethylene. The calibration scale can be detached allowing use of different inhalational agents, most commonly halothane. It is accurate with variable flow rates but this accuracy drops off in the continuous flow mode. As it is only thermally buffered, with prolonged and high gas flows the concentration decreases as the temperature decreases and you will see condensation appear on the outer surface.

The PAC vaporiser is another drawover vaporiser originally made with different models for ether, methoxyflurane, trichloroethylene and halothane. It is now only used for halothane. Some models have keyed filling. It is thermally compensated with a bimetallic strip. Its accuracy decreases rapidly when used with continuous flow and very low flows.

TECHNIQUE

In the standard drawover system the inspired oxygen concentration is determined by:

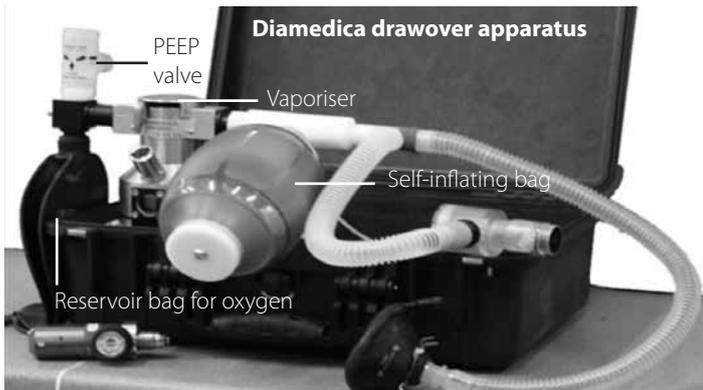


Figure 2. The Diamedica drawover apparatus. In recent years this system has been incorporated into a more typical anaesthesia machine, the Glostavent

- Total ventilation
- Oxygen flow rate
- Size of the reservoir tubing upstream of the vaporiser.

The respiratory rate and inspiration/expiration ratio have much less influence on the inspired oxygen concentration. If the reservoir tubing is short, with a volume of 104ml, then a high inspired oxygen concentration is impossible whatever flow rate is used. With a reservoir tubing of at least one metre in length and 415ml volume an inspired oxygen concentration (FiO_2) of 30% can be achieved with a flow rate $1L.min^{-1}$ and 60% with a flow rate of $4L.min^{-1}$. As air is the main carrier gas for the drawover system it is impossible to achieve 100% inspired oxygen concentration for pre-oxygenation unless you fill a large bag with 100% oxygen and attach it tightly to the reservoir tubing.

The portable Diamedica drawover system has overcome the limitation on inspired oxygen fraction by adding a reservoir bag which is constantly being filled with oxygen throughout the respiratory cycle.

Standard drawover systems have to be adapted for use in small children to overcome the deadspace and turbulence in the apparatus and the resistance in the valve. These all increase the work of breathing for small children and potentially cause alveolar collapse.

Mask induction, providing CPAP and pre-oxygenation are all more problematic with drawover anaesthesia in small children.

The other issue in paediatrics is the performance of the vaporiser. Generally the drawover vaporisers continue to be efficient at small tidal volumes as their output is affected more by a drop in temperature than tidal volume. With all the vaporisers except the recently designed ones, there is a noticeable loss of output with continuous flow.

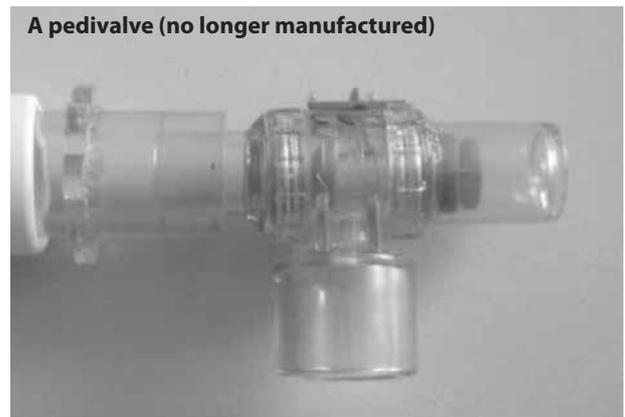


Figure 5. Paediatric Drawover using a paedivalve, a small self-inflating bag and two OMV vaporisers as in the Triservice apparatus.

In the earliest system everything was downsized except the EMO. There was a paediatric Oxford inflating bellows and a paedivalve instead of the standard Ambu E valve. A small self-inflating bag (SIB) can also be used to replace the Oxford inflating bellows.

Another way to use drawover in children is to convert it to a “manual continuous flow”. Attach an Ayre’s T-piece to the outlet of the Oxford inflating bellows (OIB) or self-inflating bag (SIB) which is attached to the outlet of the vaporiser (EMO, OMV, PAC). With the Oxford inflating bellows both valves are in use to ensure unidirectional flow as this is still a low pressure system. Compress the bellows about 6-8 times per minute with a rapid upward jerk and a slow downward movement. This sucks a flow of air and oxygen across the vaporiser and fills the bag on the T-piece which is used to ventilate the baby. The compression of the bellows has to be constant and continue in both spontaneous and assisted ventilation. A similar effect can be achieved with a self-inflating bag but as by its nature a self-inflating bag reinflates automatically with air 60 small squeezes per minute will achieve a tidal volume of

approximately 100ml. It is important not to over compress the self-inflating bag as this will be transmitted to the baby's lungs and may cause overinflation. This "manual continuous flow" mimics adult drawover but the output of the vaporisers is not as expected. You will usually need to dial up a higher inspired concentration and is therefore less economical. It can be used when there is no oxygen or electricity but it is very labour intensive as one hand is pumping the bellows and the other ventilating with the Ayre's T-piece. This can make it difficult at induction especially if there is no assistance to the anaesthetic provider.

Another way of using the drawover apparatus in small infants is to convert it to simple continuous flow. This can be done using a Farman's entrainer:

- Fits into the distal end of any drawover vaporiser.
- Acts as a venturi entraining air as oxygen flows through at $2L \cdot min^{-1}$. This produces total flow approximately $10L \cdot min^{-1}$ and FiO_2 35%.
- Oxford inflating bellows have to be closed but remain in circuit for their valves: this is still a low pressure system and flow could become bidirectional without the valves.
- Produces a continuous flow of air and oxygen so that the vaporisers markedly lose their accuracy: drawover vaporisers were designed for intermittent gas flow.
- Attach the side arm of the vaporiser to a mercury sphygmomanometer. Adjust the flow of oxygen until the manometer reads 100mmHg which is supposed to ensure a total flow of $10-12L \cdot min^{-1}$.

You can also produce continuous flow by simply attaching the oxygen supply directly onto the vaporiser and putting an Ayre's T-piece on the outlet of the vaporiser. This needs a high flow of oxygen and a one way valve between the vaporiser and the T-piece to prevent backflow and ensure filling of the bag on the T-piece. As with the Farman's entrainer the vaporiser will deliver significantly lower concentrations than expected.

In the newer designed systems with a reservoir bag attached to the inlet of the vaporiser, you can connect a T-piece with a one way valve directly to the vaporiser outlet or substitute a paediatric self-inflating bag for the adult one.

In situations where there is a lack of paediatric equipment it is safe to use the adult drawover equipment. You must ventilate all babies under 5kg body weight however short the procedure. You may allow 5-10kg infants to breathe spontaneously for short cases but use assisted ventilation for the longer cases. Allow spontaneous respiration for children over 10kg. If only the Oxford inflating bellows is in use then you must remember

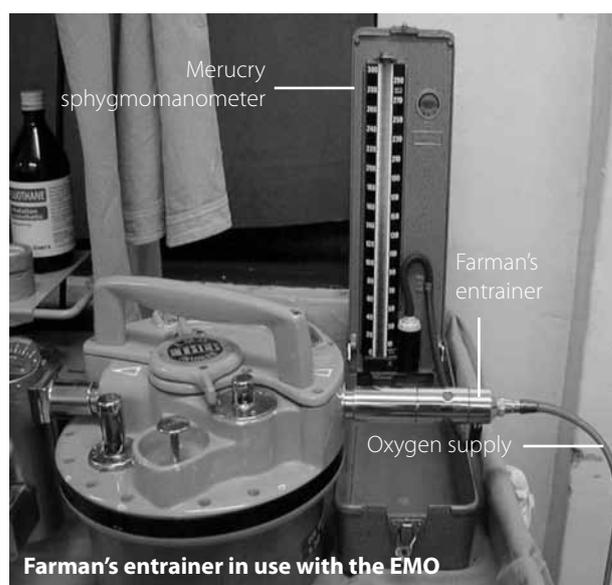


Figure 5. Farman's entrainer in use with the EMO.

that its total volume is 2 litres: only slight movement is needed to ventilate babies. You may prefer to insert a paediatric self-inflating bag instead. As ether has now been removed from the WHO essential drugs list halothane will be the main volatile in use in remote locations. With halothane it is much safer if oxygen can be added to air using reservoir tubing at least 1 metre in length.

Even though it is challenging anaesthetising small babies with minimal equipment, the drawover system can provide a safe, robust, portable and cost effective system for anaesthetising babies and children.

Key points:

- <5kg assist ventilation for all cases
- 5-10kg spontaneous ventilation for short cases, assisted ventilation if longer than 20 minutes
- >10kg spontaneous ventilation unless muscle relaxation required.

REFERENCES

1. Primary Anaesthesia Edited by Maurice King.
2. Anaesthetic vapour concentrations in the EMO system. H.G. Schaefer & J.V. Farman. *Anaesthesia* 1984; **39**: 171-80.
3. Minimum oxygen requirements during anaesthesia with the Triservice apparatus. A study of drawover anaesthesia in the young adult. S.Q.M. Tighe, et al. *Anaesthesia* 1991; **46**: 52-6.
4. The Oxford miniature vaporizer in paediatric anaesthesia. An experimental study. I.H. Wilson et al. *Anaesthesia* 1988; **43**: 700-2.

Figure 1 and 6 courtesy of Dr Iain Wilson.

Anaesthesia and congenital abnormalities

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There are many congenital abnormalities that affect children and where children may require surgery in childhood. Many conditions affect multiple systems, so it is important for the anaesthetist to have a thorough understanding of the problems that may be encountered. This article will consider Down syndrome and other congenital syndromes of relevance to the paediatric anaesthetist.

DOWN SYNDROME (TRISOMY 21)

Down syndrome (DS) is associated with major congenital abnormalities including cardiac lesions, craniofacial abnormalities and intellectual impairment. It is the commonest chromosomal abnormality with an incidence of 1 in 740 live births in the UK.¹ First described by John Langdon Down in 1866,² the association between DS and trisomy of chromosome 21 was identified in 1959.³ 95% of people with DS have trisomy 21, 3-4% have an unbalanced translocation for all or part of chromosome 21 or mosaic trisomy 21.⁴ There is higher incidence with increasing maternal age (1 in 1600 at 25 years old, 1 in 40 at 43 years old).⁵ The diagnosis of DS is made by chromosome analysis, either antenatally when risk factors are identified or postnatally in infants with the characteristic features of the condition.

Fifty years ago, 30% children with DS died in their first month, 53% in their first year, and 70% in their first 10 years.⁶ Today, 1-year survival is 90% or more, with 45% surviving to 60 years old in some countries.⁵ This increase in 1-year survival is partly due to improving surgical techniques and more aggressive treatment of life-threatening abnormalities.⁵ However, outcomes differ throughout the world. Children with DS present for surgery and general anaesthesia throughout childhood and many of the features of DS have significance for the conduct of anaesthesia.

Clinical features and anaesthetic implications of DS

The characteristic phenotypic features of patients with DS include:

- Flat occiput, short neck, small low set ears,

a flat nasal bridge, midface hypoplasia and a protruding tongue.

- Prominent epicanthic folds, upwardly slanting palpebral fissures.
- Generalised hypotonia and joint laxity.
- Single transverse palmar crease and wide 1st and 2nd toe gap.
- Brushfield spots (light coloured spots near the periphery of the iris).

Children are often low birthweight and small in infancy; however there is a tendency to be overweight by 3-4 years and to obesity in adolescence. This may in part be due to their reduced resting metabolic rate.³ Venous access may therefore be difficult. Small stature, hypotonia and increased ligament laxity require vigilance during positioning under anaesthesia. Hearing impairment, ophthalmological and dermatological problems, dental disease and orthodontic problems contribute to the many surgical interventions required, and also carry implications for development of children with DS.

Cardiovascular

Up to half of children with DS have congenital cardiac lesions.³ DS is the condition most frequently associated with congenital heart disease, therefore all children with DS who require anaesthesia should be assessed for signs and symptoms of cardiac disease. The commonest lesions are:

- Atrioventricular septal defect (AVSD) (45%)
- Ventricular septal defect (VSD) (35%)
- Atrial septal defect (ASD) (8%)
- Patent ductus arteriosus (PDA) (7%)
- Tetralogy of Fallot (TOF) (4%)
- Pulmonary vascular disease (PVD).

Children with DS develop pulmonary vascular disease and pulmonary hypertension in the presence of cardiac defects with a left to right shunt at an earlier stage than children without DS and the same defects.⁷ Pulmonary vascular disease (PVD) may also occur in

Summary

This article reviews some of the congenital conditions a paediatric anaesthetist can expect to encounter, with an emphasis on Down syndrome.

Many conditions affect multiple systems, so it is important to have a thorough understanding of the potential problems. Children presenting for surgery should be assessed carefully, with particular attention to the cardiorespiratory system and the airway.

It is not possible to cover all syndromes in one article, but a Google search on the internet provides an invaluable resource when faced with a child with a rare congenital syndrome.

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the absence of cardiac lesions as a result of hypoxaemia due to chronic pulmonary infections, hypoventilation due to muscle hypotonia, and obstructive sleep apnoea. It is recommended that all newborns with DS be assessed with an echocardiogram by a paediatric cardiologist by 6 weeks old, and a plan of action be put in place early for those with a cardiac defect.⁵ This is not always practical in low and middle income (LMIC) settings; careful auscultation and pulse oximetry, especially if both of these are repeated, will inform a discussion with a specialist centre about which patients should be seen early. Morbidity and mortality for uncorrected lesions is high. Surgical correction of AVSD should take place by 6 months of age, before the development of PVD, with some authorities recommending correction by 4 months for optimum outcome.⁵ 46% of adolescents and young adults with DS develop mitral valve prolapse with or without tricuspid valve prolapse and 17% will develop aortic regurgitation;^{2,3} repeat echocardiographic screening should be undertaken for those requiring anaesthesia as adolescents, young adults and in later life. Conduction defects can occur following surgical repair of defects.⁴

Respiratory/airway

Airway management during anaesthesia can be challenging and there is a higher incidence of perioperative respiratory complications in patients with DS:

- The incidence of obstructive sleep apnoea (OSA) is 50-75%.⁵ This is related to the craniofacial abnormalities and pharyngeal hypotonia seen in DS and will be worsened in the presence of hypertrophy, inflammation and infection of the tonsils and adenoids (children with DS will frequently present for adenotonsillectomy). Patients with DS require close observation for obstruction post anaesthesia,
- Recurrent respiratory tract infections,
- Macroglossia, small mouth, midface hypoplasia, mild mandibular hypoplasia, oropharyngeal hypotonia,
- Adenotonsillar hypertrophy,
- Subglottic or tracheal stenosis is seen in 2-6% of patients.

Musculoskeletal

- Craniocervical instability,
- Generalised poor muscle tone.

Craniocervical instability is a feature of DS and may be due to laxity of the transverse ligament and abnormalities of C1 (atlas) or C2 (axis) vertebrae. Asymptomatic atlantoaxial dislocation occurs in 12 to 20%, however spinal cord compression is rare.² Any history of neck pain, limited neck mobility or head tilt, change in gait pattern, clumsiness, abnormal neurological reflexes, abnormal sensation or the presence of bladder and bowel dysfunction suggest the possibility of cervical spine pathology. Diagnosis of cervical spine instability is difficult. Lateral flexion, extension and odontoid cervical spine radiographs are poorly predictive of cord compression. Atlantoaxial instability (AAI) is defined by an anterior atlantodental interval (AADI) greater than 4-5mm (the distance between the posterior border of the anterior arch of the atlas and the anterior

border of the odontoid peg). Where history or examination reveals any signs or symptoms suspicious of cervical cord compression, elective surgery should be postponed and specialist referral made.⁸ For urgent surgery in this setting, standard precautions should be taken to immobilise the cervical spine.⁸ There have been a number of case reports of cervical spinal cord injury related to anaesthesia and surgery.⁸ If the patient has no clinical evidence of cervical cord compression, but requires prolonged positioning in a non-neutral position or difficult laryngoscopy is anticipated, it is recommended that radiological evidence of cervical spine stability be obtained.⁸ Some centres conduct routine screening with upright lateral cervical spine Xrays, but this is not universally recommended.⁷ In the absence of abnormal neurological signs and symptoms on history and examination, avoidance of forced neck flexion, extension or rotation under anaesthesia should be adequate.⁷

Central Nervous System

- Developmental delay and moderate to severe learning disability,
- Microcephaly,
- Epilepsy (10%).

A degree of intellectual impairment and developmental delay is universal. There is a higher incidence of behavioural disorders and autism.

Gastrointestinal

Gastrointestinal abnormalities are a frequent association with DS and a common reason for surgery in early life.⁷

- Duodenal atresia is the most common association,
- Gastro-oesophageal reflux,
- Hirschsprung's disease.

Tracheoesophageal fistula, pyloric stenosis and imperforate anus may also occur.

Endocrine

Hypothyroidism occurs in up to half of affected individuals.⁵

Screening for thyroid disease should be undertaken at birth, 6 months and yearly thereafter. 1% children and adolescents with DS develop diabetes mellitus.³ Relatively low catecholamine levels and diminished sympathetic nervous system activity have been reported.⁷

Haematological/immunological

- Acute lymphoblastic leukaemia, acute myeloid leukaemia,
- Polycythaemia in neonates,
- Impaired cellular immunity with increased susceptibility to infections.

Preoperative evaluation of a child with Down syndrome

Thorough pre-operative evaluation includes a detailed history taken from the parent or carer and examination of the child, performed with particular emphasis on the cardiovascular and respiratory

systems, airway and cervical spine. The degree of cooperation from the child will vary with age and severity of learning disability.

Cardiovascular evaluation includes assessment for symptoms and signs of congenital heart disease, an ECG, and an echocardiogram and cardiology opinion where appropriate. Symptoms suggestive of congenital heart disease (CHD) include failure to thrive, dyspnoea, poor exercise tolerance, recurrent respiratory infections, cyanotic spells, syncope and chest pain. Signs include tachycardia, tachypnoea, cyanosis, finger clubbing, heart murmurs, precordial thrills, cardiomegaly, hepatomegaly, and decreased or absent femoral pulses.

You must assess the airway and evaluate for features of craniocervical instability. History and examination of the respiratory system should search for features of OSA, subglottic or tracheal stenosis and respiratory tract infection.

Children with DS are generally gentle, cheerful and outgoing, but may have a tendency to hyperactivity and will have limited understanding compared to a normal child of the same age. A clear explanation of the anaesthetic plan should be given to the parent or carer, and explain to the child in appropriate language. As with other children with developmental delay and associated behavioural disorders, careful preoperative preparation is essential, enlisting the help of parents, in order to try to gain the child's trust and cooperation. Premedication may be necessary more frequently due to limited understanding, developmental delay and a higher incidence of behavioural disorders. Premedicate with caution in the presence of severe OSA. Preoperatively the child and parent should be given the choice of either inhalational or intravenous induction as appropriate and prepared accordingly. A supportive parent or carer can often be of great benefit to the child at induction. Intravenous access can be more challenging in children with DS, so inspect potential cannulation sites carefully and apply topical local anaesthetic cream at an appropriate preoperative interval. Antacid premedication may be beneficial in those with gastro-oesophageal reflux disease. Antimuscarinics (atropine or glycopyrrolate) may be indicated for their antisialogogue effect and to attenuate a bradycardic response on induction of anaesthesia.

No abnormal responses to anaesthetics or other agents have been documented;⁶ nor has a reported excessive heart rate response to atropine,^{7,9} Vagal block with atropine may in fact be beneficial for several reasons including the higher reported incidence of bradycardia on induction of anaesthesia,⁴ the chronotropic effects in the presence of low plasma catecholamine concentrations and for its antisialogogue effects.⁷

Intraoperative considerations in Down syndrome

Routine monitoring should be used as for any other case. If inhalational anaesthesia is chosen skilled assistance should be present at induction to maintain the airway while intravenous access is obtained - this allows for the challenging nature of the airway, higher incidence of airway obstruction on induction and the occurrence of severe bradycardia on induction. Children with DS are prone to infection and all invasive lines should be inserted under full aseptic precautions.

In a retrospective review of anaesthesia for patients with DS for noncardiac surgery, severe bradycardia occurred in 3.66% cases during induction of anaesthesia.⁴ This may be associated with significant haemodynamic instability in children, who are more reliant on heart rate to preserve cardiac output. Intravenous atropine should be readily available on induction, and pre-treatment may be indicated, for instance if halothane is used.

The same study found a 1.83% incidence of airway obstruction on induction of anaesthesia and 0.54% incidence of difficult intubation.⁴ Be prepared for potentially difficult bag-mask ventilation, laryngoscopy and/or intubation. Appropriately-sized oro/naso-pharyngeal airways and laryngeal mask airway must be immediately available to rescue a potentially difficult airway. A smaller endotracheal tube (ETT) than predicted by age/weight is frequently required in the presence of subglottic or tracheal stenosis. There is also a higher incidence of airway obstruction on emergence and an increased incidence of post-extubation stridor (1.83%).⁴ Intubation should be carried out with minimal manipulation of the craniocervical spine by an experienced anaesthetist if there is a suspicion of atlantoaxial instability.

During positioning avoid excessive neck movement. During procedures such as laryngoscopy, rigid bronchoscopy and oesophagoscopy, attempt to maintain the neck in a neutral position avoiding excessive extension or flexion. Table tilt may be preferable to neck rotation, although a study investigating the safety of neck rotation for ear surgery concluded that patients with DS who have no neurological symptoms and normal cervical spine radiographs do not appear to be at high risk with neck rotation up to 60 degrees.¹¹

Routine antibiotic prophylaxis against infective endocarditis is no longer recommended.¹²

Adequate analgesia is important as postoperative pain assessment and management can be challenging due to intellectual impairment. The use of regional anaesthetic techniques may be challenging in obese patients with behavioural difficulties but the benefits include avoiding the respiratory depressant effects of opioids and improved compliance with chest physiotherapy.

Postoperative considerations in Down syndrome

Patients should be observed closely in the recovery room until full recovery from anaesthesia, as there is a higher incidence of airway obstruction. Hypotonia may affect the ability to maintain the airway, which may require simple airway manoeuvres (head tilt, chin lift or jaw thrust) or positioning the child in the lateral position to maintain airway patency. Where craniocervical instability is suspected or present, only the jaw thrust manoeuvre should be used. Use of airway adjuncts (oropharyngeal or nasopharyngeal airway) may be helpful depending on the child's level of consciousness. There is a higher incidence of post-extubation stridor in children with DS. This is managed with humidified oxygen, nebulised epinephrine (adrenaline) and intravenous dexamethasone. Postoperative agitation can be problematic, occasionally requiring sedation to prevent harm. This may be limited by the presence of parents or carers in combination with effective analgesia. Postoperative chest infections are more frequent in patients with DS.

Summary – Down syndrome

DS is the commonest congenital abnormality and is associated with defects concerning for safe anaesthetic management including congenital cardiac disease, craniofacial abnormalities, cranio-cervical instability and intellectual impairment. There is a higher incidence of perioperative respiratory complications including airway obstruction and postextubation stridor. Rarely neurological problems may occur due to atlantoaxial subluxation. Children with DS presenting for surgery require careful preoperative evaluation and perioperative care to minimise these risks

CONGENITAL CONDITIONS AFFECTING THE AIRWAY

Many congenital syndromes present challenges with airway management due to difficulty with one or more of maintaining a mask airway, mask ventilation and intubation.

There are distinct anatomical features associated with difficult airway management in children, each producing its own challenges:¹⁰

- Macroglossia (large tongue)
- Micro/retrognathia (small or receding mandible)
- Limited mouth opening
- Midface hypoplasia
- Cervical spine instability/limited motion
- Abnormal soft tissue airway structures and other facial abnormalities impairing mask fit.

It is important to perform comprehensive preoperative evaluation of the child's airway in order to plan management. Features possibly indicating airway compromise are poor feeding, apnoea, airway obstruction (OSA, stridor), respiratory failure, subcostal or intercostal muscle recession, tracheal tug, use of accessory muscles and pulmonary hypertension.

Examination should seek the features above and make an assessment of jaw protrusion, Mallampati score, mouth opening, dentition, nasal patency, neck abnormalities and restriction of movement. Formal airway assessment is often difficult in uncooperative children. Review previous anaesthetic records for details of mask ventilation, Cormack and Lehane laryngoscopy grade, special techniques used during airway management and any difficulties experienced. All of the conditions described here may be associated with awkward direct laryngoscopy, and alternate means of glottic visualization (e.g. fibre-optic or video-laryngoscopy) plus airway 'rescue' devices (e.g. supraglottic airway) should be immediately available. It is essential that the anaesthetist has an assistant when managing a child with a difficult airway; it is particularly important to address this in settings where the anaesthetist usually operates alone without a specifically trained anaesthetic assistant.

Cleft lip and/or palate

This is a relatively common abnormality and may be an isolated finding or associated with a number of different syndromes. It may contribute to difficult laryngoscopy particularly if associated with micrognathia or other craniofacial abnormalities. A more 'lateral' approach for direct laryngoscopy may be helpful. Following palate

repair, the tongue may cause airway obstruction, so these children should be extubated when they are fully awake and observed closely for impending airway obstruction.

Pierre Robin sequence

Features of this syndrome are micrognathia, posterior displacement of the tongue, cleft palate and an association with congenital heart disease (commonly VSD, PDA, ASD). With severe micrognathia there can be life threatening airway obstruction soon after birth requiring prone positioning, a nasopharyngeal airway and in very severe cases, intubation or tracheostomy. Upper airway obstruction often occurs after induction of anaesthesia, necessitating the use of airway adjuncts or a LMA to maintain the airway. Direct laryngoscopy can be very difficult. With age and growth of the mandible these children usually become easier to manage. It often occurs in otherwise normal children but can occur as a feature of a multiple malformation syndrome.

Goldenhar syndrome

Features of this syndrome are asymmetrical hypoplasia of malar, maxillary and mandible (hemifacial microsomia), associated with epibulbar dermoids, congenital heart disease (VSD, TOF) and cervical vertebral defects. Face mask ventilation is challenging due to poor mask seal and intubation may be difficult due to mandibular hypoplasia and limited mouth opening especially in the setting of right sided hemifacial microsomia and a short immobile neck. The facial asymmetry may worsen as the child grows, increasing the difficulties associated with airway management.

Treacher Collins syndrome

This condition is characterised by bilateral malar, maxillary and mandibular hypoplasia, small mouth, temporo-mandibular joint abnormalities, downslanting palpebral fissures, defect of the lower eyelid and malformation of the external ear.² Direct laryngoscopy and intubation may be very difficult and become more so with increasing age. There is an association with cleft palate and cardiac anomalies.

Syndromic craniosynostosis

Apert, Crouzon, Pfeiffer and Saethre-Chotzen are the commonest syndromes. They are characterized by prematurely fused skull-bone sutures and involvement of the facial skeleton, which can result in upper airway obstruction and raised intracranial pressure. Almost 50% have OSA and there is risk of airway obstruction on induction of anaesthesia, difficult mask ventilation and perioperative respiratory complications. Difficult laryngoscopy is unusual.

Mucopolysaccharidoses

These are classified into the following sub-types:

- I H: Hurler syndrome
- I S: Scheie syndrome; previously classified Type V
- I HS: Hurler-Scheie compound
- II: Hunter syndrome
- III: Sanfilippo syndrome

IV:	Morquio syndrome	
V:	Formerly Scheie syndrome	
VI:	Maroteaux-Lamy syndrome	
VII:	α -Glucuronidase deficiency	
Type I H, I HS, II, VII		Affects bones and intellect
Type III		Affects intellect only
Type I S, IV, VI		Affects bones only

The mucopolysaccharidoses (MPS) are lysosomal storage disorders caused by enzyme defects that result in accumulation of glycosaminoglycans in cellular lysosomes throughout the body. This results in coarse facial features, airway abnormalities, organomegaly, cardiac dysfunction, joint and bone deformities, visual, auditory and intellectual impairment. The consequences of deposition in the soft tissues of the mouth and pharynx are macroglossia, thickened oropharyngeal and nasal mucosae, adenoidal and tonsillar hypertrophy, copious secretions and a narrow trachea. Deposition in joints and bones can result in a short, immobile and potentially unstable neck and reduced temporo-mandibular joint movement. All of the above can combine to make airway management and intubation extremely difficult. The most challenging are those with Hurler (MPS I) and Hunter (MPS II) syndromes. These conditions are progressive and lethal, but the natural history of airway disease may be altered by successful bone marrow transplantation. Spontaneous ventilation should be maintained until the airway has been secured; however airway obstruction can occur early during inhalational induction. Placement of LMA airway in these patients may be difficult, require several attempts, and be only partially effective or ineffective. While managing the airway, it is important to maintain the neck in a neutral position as there may be cervical spine involvement, as in Morquio syndrome (MPS IV). Spinal cord compression may occur due to thickening of the dura and to odontoid hypoplasia. Preoperative MRI scans (without anaesthesia) of the spinal cord may be appropriate in affected individuals.

Beckwith-Wiedemann syndrome

This is a syndrome of macrosomia, visceromegaly, macroglossia, omphalocele and hypoglycaemia. Cor pulmonale may result from chronic airway obstruction. Cardiomegaly may be part of the visceromegaly which may also involve other organs. Macroglossia can cause upper airway obstruction and may complicate direct laryngoscopy; partial glossectomy is occasionally required. Serum glucose should be monitored perioperatively.

Klippel-Feil syndrome

This condition is due to a failure of cervical vertebrae segmentation during foetal development. These children have a short neck with low hairline and neck rigidity due to fusion of 2 or more of the cervical vertebrae, most commonly C2 and C3.² Direct laryngoscopy/intubation may be very difficult.

CONDITIONS ASSOCIATED WITH CONGENITAL HEART DISEASE

Cardiac disease is a frequent association of a number of congenital abnormalities. Careful evaluation of the cardiovascular system should be undertaken when other major abnormalities are present. Aside from DS, other conditions of note include:

Turner syndrome

Females with short stature, short webbed neck, ovarian dysgenesis^{2,10} and XO karyotype. 30% have a bicuspid aortic valve and 10% coarctation. Micrognathia may predispose to airway difficulty. 50% have renal abnormalities; therefore take caution with renally excreted drugs.

Noonan syndrome

These individuals have phenotypic similarities to Turner syndrome; short stature and short or webbed neck. Cardiac defects associated with this condition are pulmonary valve stenosis (62%), hypertrophic cardiomyopathy (20%), ASD, TOF, aortic coarctation, mitral valve anomalies and atrioventricular canal defects.^{2,10} Other associations are micrognathia, hydronephrosis and platelet dysfunction. Preoperative assessment should focus on possible difficulty with intubation, cardiac anomalies and renal function.

CHARGE association

This is an association of the following abnormalities:²

- **C:** ocular Coloboma (80-90%)
- **H:** Heart defect (TOF, PDA, double outlet right ventricle, VSD, ASD, right-sided aortic arch-75-80%)
- **A:** choanal Atresia (58%), growth (70%)
- **R:** developmental Retardation (100%)
- **G:** Genital anomalies (75%)
- **E:** Ear anomalies or deafness (90%)

Tailor the anaesthetic according to the individual lesions present.

VACTERR (or VACTERL) association

This is an association of the following abnormalities:²

- **V:** Vertebral anomalies (70%)
- **A:** anal Atresia (80%)
- **C:** VSD and other Cardiac defects (53%)
- **T-E:** Trachea-oEsophageal fistula (70%)
- **Renal anomalies** (53%)
- **Radial dysplasia (or L for limb)** (65%)

Again, anaesthetic management is dictated by the specific lesions present.

Di George syndrome (22q11.2 deletion syndrome)

Chromosome 22 microdeletions result in the failure of the 3rd and 4th embryological pharyngeal pouches to differentiate into thymus and parathyroid glands. Hypoplasia/aplasia of the thymus results in deficiency in T-cell-mediated immunity leading to a susceptibility to

fungal and viral infections and hypoplasia/absence of parathyroids results in severe hypocalcaemia with tetany and seizures in early infancy. Cardiac defects are present in 85% and DiGeorge syndrome is the second most common cause of congenital heart disease, most commonly VSD (62%), right aortic arch (52%), TOF (21%), aberrant left subclavian artery, truncus arteriosus, double aortic arch and interrupted aortic arch.^{2,10} Affected individuals commonly have micrognathia, cleft palate and choanal atresia. Micrognathia may make intubation difficult and a short trachea predisposes to inadvertent endobronchial intubation. Serum calcium should be checked preoperatively, blood products should be irradiated to prevent a graft versus host reaction and meticulous sterile technique is important when caring for these children.

OTHER CONGENITAL CONDITIONS OF IMPORTANCE TO THE PAEDIATRIC ANAESTHETIST

Neuromuscular disease

These conditions can be divided into the following categories:¹³

- Myasthenic syndromes (abnormalities in the release or action of acetylcholine),
- Channelopathies (abnormalities in the post-synaptic membrane or the sarcoplasmic reticulum),
- Dystrophies / myotonias (abnormalities in the myofibrils),
- Mitochondrial myopathies (abnormalities in mitochondria).

Important congenital conditions in this group of disorders include:

Duchenne muscular dystrophy

Duchenne muscular dystrophy (DMD) is seen in 1 in 3500 live male births in the UK. It is an X-linked recessive disorder. This condition is associated with rapidly progressive weakness presenting between 2 and 5 years of age with sufferers usually wheelchair bound by the age of 12. Myocardial degeneration leads to cardiac failure and respiratory muscle weakness results in ventilatory failure. Death usually occurs by third or fourth decade. There is a weak recommendation that volatile agents be avoided due to the risk of anaesthesia-induced rhabdomyolysis. Succinylcholine (suxamethonium) must NOT be used due to risk of hyperkalaemia.

Myotonic dystrophies

These are a group of hereditary diseases of skeletal muscle associated with sustained contraction after stimulation. Dystrophia myotonica is the commonest form and typically presents in late adolescence; 50% develop cardiac conduction defects. Other diseases in this group are myotonia congenita, which presents at birth or early childhood and paramyotonia congenita, which also presents in early childhood and is induced by exposure to cold. Succinylcholine can induce generalised myotonia and is contraindicated in these conditions. If muscle spasm occurs (e.g. masseter spasm), lignocaine directly injected into the muscle may prove useful.

Mitochondrial myopathies

These are a heterogeneous group of conditions with a collective

incidence of 1 in 4000.¹³ Clinical features are a consequence of defects in electron transport or oxidative phosphorylation and tissues that are most metabolically active such as the nervous system and muscles are most affected.¹⁰ The three basic categories are:

- Respiratory chain deficiencies,
- Mitochondrial DNA mutations including mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS), mitochondrial neuro-gastrointestinal encephalopathy (MNGIE), myoclonic epilepsy with ragged red fibres (MERRF),
- Mitochondrial deletions such as Kearns-Sayre syndrome.

The severest forms present in the neonatal period with profound weakness, acidosis, liver/renal failure and substantial neurological impairment. Optimal anaesthetic technique for this group of conditions remains controversial although propofol may best be avoided, particularly by infusion.

Pre-operative work-up will depend on the clinical condition of the patient but should include a full cardiorespiratory evaluation including (in severe cases) arterial blood gases, pulmonary function tests and where appropriate assessment of other organ function with measurement of electrolytes, glucose, lactate, pyruvate, creatinine kinase, liver function and renal function.¹⁰

Malignant hyperthermia

Three conditions with a definite link with malignant hyperthermia are central core disease, King-Denborough syndrome and Evans Myopathy.¹³

Inherited metabolic disorders

These are caused by genetic defects resulting in the absence/dysfunction of structural proteins or enzymes. Some conditions present significant challenges to anaesthesia. Symptoms and signs may be related to accumulation of intermediate metabolites proximal to the blocked enzyme that may be toxic or inappropriately stored within cells and/or deficiency of a metabolite downstream of the blocked enzyme. They are broadly classified into the following categories:¹⁴

- Disorders of amino acid (and branched-chain amino acid) metabolism
- Urea cycle disorders
- Organic acidaemias
- Disorders of carbohydrate metabolism e.g. glycogen storage diseases
- Lysosomal storage disorders
- Disorders of fatty acid oxidation.

Conditions of particular concern to the anaesthetist are the mucopolysaccharidoses (described above); glycogen storage disorders, which prevent the production of glucose from glycogen and cause the accumulation of glycogen within tissues such as liver and muscle;

and organic acidaemias (propionic and methylmalonic acidaemia). As a general principle, keep children with inherited metabolic disorders well hydrated, and take care to avoid hypoglycaemia during starvation. Children with organic acidaemias are treated with a protein-restricted diet, and children with glycogen storage diseases are particularly prone to hypoglycaemia.

Epidermolysis bullosa

This is an inherited group of skin disorders characterised by cleavage at the dermal-epidermal junction resulting in erosions and blisters from seemingly minor trauma to skin or mucous membranes.

There are three main forms of epidermolysis bullosa (EB), each with several subtypes:

- *Simplex*: This is usually a relatively mild form with rapid healing and little scarring,
- *Lethalis*: This is a junctional epidermolysis bullosa. This is a severe form of the condition that presents at birth, leading to extensive scarring and death usually before the age of 2,
- *Dystrophic*: This is a rare but severe form of the disease. Lesions are slow to heal with extensive scarring resulting in strictures of the mouth, pharynx, larynx and oesophagus, limb contractures and fusion of the digits. Blistering / strictures of oesophagus and oropharynx can lead to decreased oral intake and nutrition leading to growth retardation and anaemia. Infections are common and antibiotic prophylaxis may be necessary. Adrenal suppression can occur due to use of powerful topical steroids or oral steroids necessitating perioperative steroid replacement. There is an association with dilated cardiomyopathy.

Children with EB must be assessed carefully preoperatively, with particular reference to the airway. Airway management may be difficult as a result of oral lesions, limited mouth opening, adhesion of the tongue and pharyngeal strictures. Care must be taken to avoid trauma to skin or mucous membranes. Frictional shearing forces must be avoided. Allow the patient to position himself or herself if possible. Avoid intubation and instrumentation of the airway or lubricate the ETT and laryngoscope generously and select an endotracheal tube at least 0.5mm smaller than predicted by age/size. If a LMA is used ensure that it is lubricated and it may be appropriate to select a smaller size. Use a lubricated face-mask for inhalational induction and lubricate gloves or apply paraffin gauze to the mask as well as the face. Do not use adhesive tape on the skin. Secure intravenous access and ECG dots with low-adherence dressings such as Mepiform or Mepitel, which are silicone based. If these are unavailable, an unfolded gauze swab smeared with paraffin gel makes a sticky dressing that doesn't shear the skin. This may be kept in place with a crepe bandage. Padding should be applied under the non-invasive blood pressure cuff. Oropharyngeal secretions can be cleared with lubricated soft suction catheters under low pressure, avoiding contact with the mucosa.¹⁵ Anaesthesia for this group of patients can be extremely challenging but with meticulous attention to detail good outcomes are possible.

Osteogenesis imperfecta

This is an inherited disorder of connective tissue principally involving bones, making them fragile. There are several forms of osteogenesis imperfecta (OI). The two main forms are OI tarda (type I) and congenita (type II).

- *OI Tarda* presents with pathological fractures, blue sclera and deafness. Osteoporosis can result in kyphoscoliosis and restrictive lung disease. Fragility of vessels leads to subcutaneous haemorrhage. Dentine deficiency results in carious, fragile teeth. Extreme care must be taken with positioning and moving patients; fractures have been reported from minimal manipulation, including application of a blood pressure cuff.¹⁶ Careful laryngoscopy should avoid neck hyperextension and dental damage. Intravenous access can be difficult to maintain due to fragile vessels. OI is associated with abnormal platelet function which may exacerbate perioperative bleeding.
- *OI Congenita* is the more severe form and usually presents as a stillbirth or proves rapidly fatal.

CONCLUSION

In this article we have reviewed some of the congenital conditions a paediatric anaesthetist can expect to encounter occasionally. Children presenting for surgery should be assessed carefully, with particular attention to the cardiorespiratory system and the airway. It is not possible to cover all syndromes in one article, but a Google search on the Internet provides an invaluable resource for the paediatric anaesthetist faced with a child with a rare congenital syndrome.

REFERENCES

1. Morris JK, Alberman E. Trends in Down's syndrome live births and antenatal diagnoses in England and Wales from 1989 to 2008: analysis of data from the National Down Syndrome Cytogenetic Register. *Br Med J* 2009; **339**: b 3794.
2. Jones K. Smith's recognizable patterns of human malformation. 6th ed. Philadelphia: WB Saunders, 2006.
3. Roizen NJ, Patterson D. Down's syndrome. *Lancet* 2003; **361**: 1281-89.
4. Borland LM, Colligan J and Brandom BW. Frequency of anaesthesia-related complications in children with Down's syndrome under general anaesthesia for non-cardiac procedures. *Pediatric Anaesthesia* 2004; **14**: 733-8.
5. The Down's Syndrome Medical Interest Group. <http://dsmig.org.uk>
6. Kobel M, Creighton RE and Steward DJ. Anaesthetic considerations in Down's syndrome: Experience with 100 patients and a review of the literature. *Canadian Anaesthesiology Society Journal* 1982; **29**: 593-9.
7. Mitchell V, Howard R, Facer E. Clinical review: Down's syndrome. *Paediatric Anaesthesia* 1995; **5**: 379.
8. Hata T, Todd M. Cervical spine considerations when anesthetizing patients with Down syndrome. *Anesthesiology* 2005; **102**: 680-5.
9. Roizen MF. Anesthetic Implications of Concurrent Diseases. p 1116 In: *Anesthesia* Ed: Miller RD. Churchill Livingstone 2000. 5th edition.

10. Rampersad SE, Lynn AM. Congenital and inherited disease. p 425-444. In Hatch & Sumner's Textbook of Paediatric Anaesthesia. Ed Bingham R, Lloyd-Thomas A, Sury M. Hodder Arnold 2008. 3rd edition.
11. Todd NW, Holt PJ, Allen AT. Safety of neck rotation for ear surgery in children with Down syndrome. *Laryngoscope* 2000; **110**: 1442-5.
12. Prophylaxis against infective endocarditis. NICE guidelines, March 2008. <http://www.nice.org.uk/nicemedia/pdf/CG64NICEguidance.pdf>
13. Ragoonannan V, Russell W. Anaesthesia for children with neuromuscular disease. *CEACCP* 2010; **10** (5): 143-7.
14. Stuart G, Ahmad N. Perioperative care of children with inherited metabolic disorder. *CEACCP* 2011; **11** (2): 62.
15. Herod J, Denyer J, Goldman A, Howard R. Epidermolysis bullosa in children: pathophysiology, anaesthesia and pain management. *Paediatric Anaesthesia* 2002; **12**: 388-97.
16. Lauder GR. Orthopaedic surgery. p 547-548. In Hatch & Sumner's Textbook of Paediatric Anaesthesia. Ed Bingham R, Lloyd-Thomas A, Sury M. Hodder Arnold 2008. 3rd edition.

The anaesthetic management of children with sickle cell disease

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INTRODUCTION

Sickle cell disease (SCD) is a congenital haemoglobinopathy inherited in an autosomal recessive manner. It is a multisystem disease which affects approximately 4 million people worldwide. Surgery and anaesthesia carry a high risk for these patients, and meticulous perioperative care is essential to prevent complications of SCD.¹

PATHOPHYSIOLOGY

Adult red blood cells normally contain three different types of haemoglobin; Haemoglobin A (HbA) which makes up 96-98% of total haemoglobin, haemoglobin A2 (HbA2) which accounts for 1.5-3% of the total, and foetal haemoglobin (HbF) which accounts for 0.5-0.8% of the total.

Haemoglobin S (HbS) occurs as a result of a single DNA base change (adenine to thymidine) that results in the substitution of valine for glutamic acid in the β -globin chain.

Sickle cell diseases are inherited in an autosomal recessive fashion, with homozygous expression of the abnormal gene (HbSS) producing SCD. These patients have no normal adult haemoglobin (HbA) and only have HbS, HbA2 and HbF, with approximately 95% haemoglobin as HbS. Patients who are heterozygous for HbS (HbSA) are carriers but are asymptomatic and have a normal life expectancy.

Sickle haemoglobin (HbS) polymerises into insoluble microfibrils in the deoxygenated state. It is thought that these parallel microfibrils cause red cell membrane damage and result in the classical sickle cell deformity (Figure 1). The deformed red cells are more rigid and less capable of passing through the microcirculation, causing increased blood viscosity and impaired blood flow. The cells also have a shortened survival time (5-15 days in homozygous sickle cell disease) with the resulting haemolytic anaemia that is characteristic of SCD.

There is increasing evidence that the primary event in SCD is oxidative damage to the arterial endothelium (the lining of the vessel wall) due to the effects of sickle haemoglobin breakdown. The vascular damage caused by HbS, may be due to its extremely unstable

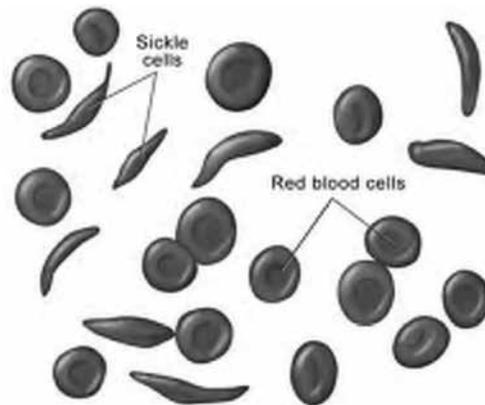


Figure 1. Normal and sickle red cell morphology

nature, rather than its insolubility. Biochemical markers of endothelial damage are present and suggest that perhaps the disease should be considered as a chronic inflammatory disorder.

The spectrum of sickling disorders is widened by the combination of HbS with other haemoglobinopathies such as thalassaemia and haemoglobin C and haemoglobin D. This is because polymerisation of HbS is affected by the presence of other haemoglobins, but in varying degrees. For example, patients with HbSD are severely affected, while patients with HbSC are less affected by sickling, and suffer more thrombotic complications. The combination of α and β thalassaemia with HbS result in disease ranging in severity depending on the nature of the thalassaemia mutation.

EPIDEMIOLOGY

SCD affects approximately 4 million people worldwide and it is most common in West African and Caribbean populations. In Equatorial Africa the sickle cell trait occurs in up to 30% of the population. This is due to a phenomenon called balanced polymorphism, which is when the heterozygote for two alleles of a gene has an advantage over either of the homozygous states. Heterozygotes for sickle cell anaemia show a marked resistance to malaria. The mechanism for this is unknown but could be explained by the fact that sickle cell trait red cells deform when infected by the

Summary

Sickle cell disease may cause serious perioperative complications. Management of these patients requires careful preparation, and close attention to those factors that could precipitate a sickle crisis. The basic principles of oxygenation, hydration, analgesia, avoidance of hypothermia and acidosis, and blood transfusion where indicated, are essential.

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parasite, and are then targeted for destruction by phagocytes. The destruction of these cells decreases the parasite burden. Because of this relative resistance, people with sickle cell trait in high malaria areas are more likely to reach reproductive age and pass on their genes to the next generation.

In North America approximately 8% of the black population has sickle cell trait, and up to 1.3% has SCD. The majority of SCD in the UK is found in African-Caribbean populations in large cities where up to 10% individuals carry the gene.

The HbS gene also occurs in some areas in Mediterranean regions such as Greece, southern Italy, Turkey, and in Saudi Arabia and central India.

DIAGNOSIS

The gold standard for the diagnosis of SCD is haemoglobin electrophoresis.

The simpler Sickledex test confirms the presence of HbS, however electrophoresis is required to distinguish between HbSA (sickle cell trait) and HbSS (sickle cell disease).

The Sickledex test uses sodium metabisulphite as a reducing agent that causes HbS to precipitate in a hyperosmolar phosphate buffer solution to produce a cloudy suspension.

The Sickledex test is not reliable in the neonatal period where low levels of HbS and high levels of HbF (with normal solubility) may result in false negative results. It becomes reliable after 6 months of age when the HbF levels have dropped.

Haemoglobin electrophoresis separates molecules on the basis of their charge at a given pH (Figure 2). Electrophoresis of umbilical cord blood can be used for diagnosis in the newborn.

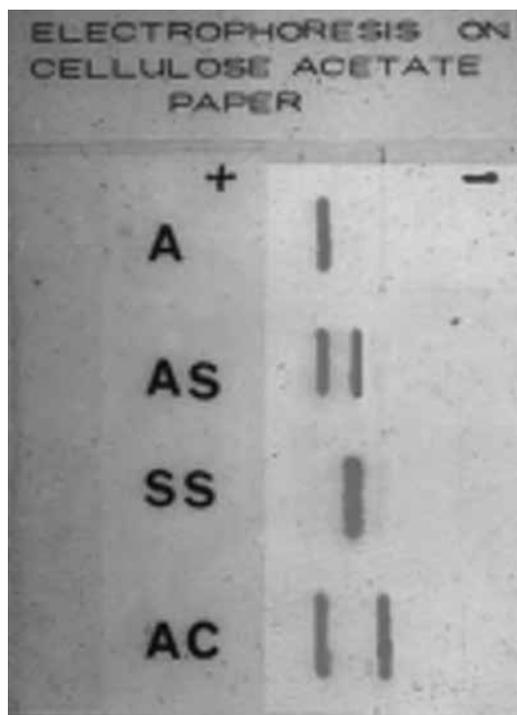


Figure 2. Haemoglobin electrophoresis results for different haemoglobin types. Antenatal diagnosis of sickle cell disease is possible by analysis of the DNA of foetal tissue from chorionic villous sampling or amniocentesis.

CLINICAL MANIFESTATIONS OF SCD IN CHILDREN

SCD has a variety of clinical presentations.

Anaemia

This is universal in patients with HbSS. Patients usually have a haemoglobin level of 6-9g.dL⁻¹. The anaemia is usually well tolerated, and adequate tissue oxygenation is maintained due to a compensatory increase in cardiac output and increased effective release of oxygen to the tissues due to the low affinity of HbS for oxygen. A systolic flow murmur (non-pathological murmur as a result of increased cardiac output) is a frequent finding, and congestive heart failure with cardiomegaly on clinical examination/chest radiograph can occur in adults. Children with SCD should receive iron and folic acid supplementation.

Painful crises

This is associated with the sudden onset of severe pain, most commonly arising in bone and joints due to ischaemia and infarction in the marrow or cortical bone. Dactylitis (painful swelling of small bones of hands and feet) occurs in up to half of children by the age of two years and is a sign of severe disease. Abdominal pain occurs in older children and can be caused by bowel dysfunction, organ infarction or referred pain from the ribs. These abdominal crises can be difficult to distinguish from other common acute surgical disorders. 1% of patients have more than six episodes of pain per year. Precipitants for acute painful crises include infection, dehydration, cold, hypoxia and stress.

Acute chest syndrome

This is defined as a fever of more than 38.5°C, respiratory distress or chest pain and the appearance of new lung lobar infiltration on chest X-ray. Hypoxia is common and ventilatory support is occasionally needed in severe sickle chest crisis. The majority of patients are managed with oxygen therapy, hydration and blood transfusion. The incidence of acute chest syndrome in the postoperative child may be as high as 10% in those with severe disease undergoing major surgery. Risk factors for sickle chest crisis are age between 2-4 years and a persistently raised white cell count. Multiple episodes of acute chest syndrome in children are likely to result in pulmonary fibrosis and chronic lung disease as the child gets older.

Cerebrovascular accidents (CVA)

The majority of CVAs in patients with SCD occur during childhood (5% of children with sickle cell disease have overt CVAs due to ischaemia). These are typically caused by vascular lesions in the cerebral vessels and may present as watershed infarctions during a sickle crisis (infarction occurring at the more vulnerable regions between major cerebral arterial zones). Transcranial Doppler ultrasonography can identify children at risk of cerebral infarction, by detecting reduced blood flow in cerebral vessels. It has been shown that treating patients at risk with regular transfusion programmes significantly reduces the incidence of stroke. Children are also at risk of intracerebral and subarachnoid haemorrhage.

Aplastic crisis

This is usually precipitated by infection, parvovirus being an important pathogen. There is suppression of erythropoiesis (red blood cell formation) in the bone marrow and a dramatic fall in haemoglobin

levels. Early diagnosis and treatment with blood transfusion is essential.

Acute splenic sequestration

This is a rare complication that is most common in children under the age of five. Large numbers of red cells are sequestered in the spleen and the haemoglobin level drops precipitously. This may present with acute collapse and shock, and may require resuscitation and blood transfusion. Children who suffer repeat episodes of splenic sequestration may require splenectomy.

More commonly, splenic infarction occurs as a result of repeated sickling episodes, which results in functional hyposplenism. Patients are at increased risk of infections, particularly with encapsulated bacteria such as *Streptococcus pneumoniae*, *Neisseria meningitidis*, and *Haemophilus influenzae B*. All children with homozygous SCD should receive prophylactic penicillin V from birth.

Osteomyelitis

Patients are at higher risk of osteomyelitis than the rest of the population, with the most common pathogens being *salmonella* and *staphylococci*.

Priapism

Attacks start as young as the age of eight and are reported by up to 30% of male sufferers of SCD. It can occur in the postoperative period. Treatment includes hydration, exchange transfusion and intracavernous injections of an alpha-adrenergic agent.

Avascular necrosis

Intravascular sickling of the red blood cells in the microcirculation of the bone results in intramedullary sludging, stasis, thrombosis, and progressive ischaemia, most often of the femoral head. These patients present with pain in the affected joint. Orthopaedic management may be conservative or surgical.

Long-term complications of SCD in adults

Recurrent sickle cell crises may cause many complications including gall stones, sickle retinopathy, leg ulcers, chronic renal failure due to renal parenchymal scarring, pulmonary hypertension, chronic lung disease, and neurological impairment. Chronic bone damage may occur, leading to avascular necrosis, impaired growth and joint damage.

ANAESTHETIC MANAGEMENT OF CHILDREN WITH SCD

Pre-operative screening

All children in a high-risk population or those with a positive family history should be screened for SCD.

Pre-operative assessment and preparation

Patients with a history of chest crisis, stroke, frequent painful crises, or those with severe obstructive sleep apnoea have a higher risk of perioperative complications. All patients with SCD require meticulous perioperative care.^{3,4}

Pre-operative assessment should involve a careful review of all systems.

- Multiple episodes of acute chest syndrome may result in reduced lung volumes, pulmonary infarction and pulmonary hypertension with low oxygen saturation. It is important to check the baseline oxygen saturation before surgery.

- Although more commonly seen in adults, cardiomegaly may be seen on chest X-ray, and echocardiography may be indicated to assess cardiac function.
- Careful neurological examination is essential and any pre-existing neurological deficit from previous CVAs should be documented.
- Renal and hepatic function should also be assessed for signs of end-organ damage. Even children with HbAS have a renal concentrating defect and may not tolerate dehydration.
- If there is any evidence of active infection, elective surgery should be postponed.

Where possible, children with SCD should be scheduled first on the operating theatre list to avoid prolonged starvation and dehydration. Patients should be encouraged to drink free clear fluids until two hours before surgery.

Blood transfusion and SCD

Preoperative blood transfusion is a controversial area,⁵ particularly now that standards of anaesthetic care have improved for patients with SCD. The pathophysiology of the disease is better understood and many of the precipitating factors for sickle crisis in the perioperative period can be avoided (see below). The NHS Blood and Transplant service in the UK recently undertook a randomized controlled trial to evaluate whether blood transfusion should be given to patients with SCD pre-operatively (so-called TAPS trial). Although the recruitment target was 400 patients, the trial was ended early (after 70 patients) as a review of patient safety identified that there were more serious complications in patients who did not receive pre-operative blood transfusion⁶ (unpublished data).

Theoretically, reducing the percentage of HbSS by prophylactic transfusion should prevent complications. However, aggressive transfusion regimens are associated with a high incidence of transfusion-associated complications.

In resource poor areas where screening for infection and highly specific blood cross matching is limited, the balance of the risks versus the benefits of blood transfusion needs to be carefully considered.

Management plans for transfusion therefore need to be individualised for each patient, taking into account the patient's medical history and type of surgery, in consultation with the anaesthetist, surgeon, paediatrician and haematologist, as well as the patient's family.

Guidelines may vary between hospitals and between regions. Transfusion may be used to increase the haemoglobin level; repeated top-up transfusion will also reduce the percentage of HbS in the blood. Below are current transfusion guidelines at Great Ormond Street Children's Hospital in London.

- Children with no special risk factors having short procedures such as insertion of myringotomy tubes or minor dental work: no transfusion, provided the haemoglobin is at the normal baseline level ($Hb > 6g.dL^{-1}$)
- Children with no special risk factors having intermediate risk surgery such as tonsillectomy or laparotomy: top-up transfusion to $Hb 9-11g.dL^{-1}$

- Children who have had a chest crisis, CVA or suffer frequent painful crises, or children undergoing major surgery such as thoracic or neurosurgery: sequential top-ups or exchange transfusion to achieve a Hb of 9-11g.dL⁻¹ and a HbS level of <30%
- As it is essential to avoid increased tissue viscosity, the Hb should not exceed 12g.dL⁻¹
- For emergency surgery, patients should ideally be treated the same, but if time does not allow, then blood should be crossmatched and ready for surgery. All cases should be discussed with a haematologist if possible

INTRAOPERATIVE MANAGEMENT

Oxygenation

The primary goal is to maintain good oxygenation during the perioperative period. Perioperative pulse oximetry monitoring is essential as patients may have impaired oxygen delivery resulting from chronic anaemia or chronic lung damage, and may have a limited ability to maintain tissue perfusion and oxygenation during hypoxic episodes. Even short periods of hypoxia must be avoided. Postoperative continuous positive airway pressure (CPAP) or a nasopharyngeal airway may be indicated in those with obstructive sleep apnoea (see below).

Dehydration

Dehydration is poorly tolerated. Dehydration may lead to increased tissue viscosity, poor perfusion, acidosis and increased sickling. Adequate hydration is essential and must be maintained before, during and after surgery. The patient should be encouraged to drink clear fluids up until 2 hours before surgery, or if this is not possible, to have intravenous fluids during the preoperative fasting period. Intravenous fluids should be used during surgery, and postoperative intravenous fluids should be prescribed until oral intake is re-established.

Acidosis

Avoid acidosis. Acidosis causes increased sickling, with subsequent increased blood viscosity and impaired tissue perfusion. This will cause the tissues to become more acidotic, causing further sickling, which may result in a sickle crisis.

Temperature management

Avoid hypothermia. Hypothermia causes vasoconstriction, hypoperfusion, increased blood viscosity, and decreased venous oxygen tension which all lead to increased sickling.

Vascular stasis

Avoid vascular stasis by maintaining a good circulating volume, careful positioning, and the use of thromboembolic deterrent (TED) stockings. Pneumatic calf compression devices can be used during prolonged surgery. These should be avoided if there is evidence of peripheral vascular occlusive disease.

Tourniquets

Studies reporting outcomes after tourniquet use are scarce in patients with sickle cell disease. A recent systematic review concluded that tourniquets may be used with relative safety in most patients, as long as other peri-operative precautions are taken. The use of tourniquets

in patients with SCD should therefore be considered on an individual basis weighing up the risks and benefits.

Tourniquet after exsanguination of the limb may be used safely in patients with sickle cell trait.

Cell saver devices

The high incidence of sickling in cell savers prevents their use in SCD.

POSTOPERATIVE MANAGEMENT

Oxygen therapy

Oxygen saturation should be monitored continuously and supplemental oxygen should be given to maintain saturations >92%.

Fluid management

Continue intravenous maintenance fluids until the child is tolerating oral fluids.

Postoperative analgesia

Management of post-operative pain is challenging. Patients may have very high perioperative analgesic requirements, and may have developed tolerance to opioids. A multimodal approach should be used with a combination of opioids where indicated, paracetamol and NSAIDs, and regional anaesthesia when possible.

Physiotherapy

Physiotherapy and early ambulation are important to avoid vascular stasis.

Nasopharyngeal airway

Obstructive sleep apnoea secondary to adenotonsillar hypertrophy is common in children with SCD. Careful attention should be paid to these patients postoperatively to avoid airway obstruction, hypoventilation or hypoxia. A nasopharyngeal airway may be used after tonsillectomy or in those with severe obstructive sleep apnoea to prevent post-operative airway obstruction and hypoxia.

POSTOPERATIVE COMPLICATIONS

Patients should be monitored carefully for early signs of complications as serious post-operative complications usually occur within 48hrs of surgery. These include:

- Painful crisis
- Cerebrovascular accident
- Acute chest syndrome.

Management of sickle complications

The anaesthetic team may be involved in managing the acute complications of sickle cell disease, both when they present post-operatively, and when the patient presents to the hospital with an acute crisis.

- The management of all sickle crises includes the same principles of establishing intravenous fluids, oxygen therapy, analgesia, and antibiotics.
- Analgesia may require high doses of opiates, as well as the use of regular paracetamol and NSAIDs such as ibuprofen or diclofenac.
- Transfusion to an Hb>10g.dL⁻¹ is important, but over transfusion

(>12g.dL⁻¹) must be avoided. Exchange transfusion to reduce HbS <20-30% may be indicated in certain situations such as acute chest syndrome or CVA. As a guide, transfusion of 4ml.kg⁻¹ of packed cells or of 8 ml.kg⁻¹ of whole blood raises the haemoglobin concentration by 1g.dL⁻¹.

- Ventilatory support (continuous positive airway pressure (CPAP), or intubation and ventilation) may be required for acute chest syndrome. Patients should be carefully monitored for signs of respiratory decompensation.
- Acute sequestration crisis is an important cause of death in children with sickle cell disease. Acute hypovolaemia can occur due to pooling of blood in the spleen. Treatment is transfusion of blood and intravenous 0.9% saline for volume replacement.

CONCLUSIONS

Anaesthetists need to be aware of the possible serious complications of SCD in the perioperative period. Management of these patients requires careful preparation, and close attention to those factors that could precipitate a sickle crisis.⁷ The basic principles of oxygenation, hydration, analgesia, avoidance of hypothermia and acidosis, and blood transfusion where indicated, are essential.

REFERENCES

- Locke C. Anaesthetic management of sickle cell disease in children. *ATOTW* (2009) **153**. Available at: http://www.wfsahq.org/components/com_virtual_library/media/e0645e0e5cce77faef81cd00f0a0efa4-134dcf42c0b8f21e61db61b943aa0c8b-153-Sickle-cell-disease-in-children.pdf
1. Marchant WA, Walker I. Anaesthetic management of the child with sickle cell disease. *Paediatr Anaesth* 2003;13:473-89.
 2. Nandi R, Martin R. Haematology. In: Bingham R L-TA, Sury M, editor. *Hatch and Sumner's Textbook of Paediatric Anaesthesia*. 3rd ed, 2008.
 3. Firth PG, Head CA. Sickle cell disease and anesthesia. *Anesthesiology* 2004;101:766-85.
 4. Frietsch T, Ewen I, Waschke KF. Anaesthetic care for sickle cell disease. *Eur J Anaesthesiol* 2001;18:137-50.
 5. Hirst C, Williamson L. Preoperative blood transfusions for sickle cell disease. *Cochrane Database Syst Rev* 2012;1:CD003149.
 6. http://www.sicklecellsociety.org/websites/123reg/LinuxPackage22/si/ck/le/sicklecellsociety.org/public_html/news/view/22
 7. Davies SC, Oni L. Management of patients with sickle cell disease. *BrMed J* 1997;315:656-60.

HIV in children and anaesthesia

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It is estimated that 3.3 million children under 15 years of age are living with HIV, the vast majority of which are in sub-Saharan Africa. Due to improved HIV prevention a 35% reduction in numbers from 2009 means that in 2012 approximately 260,000 children under 15 were diagnosed with HIV and 647,000 children were receiving antiretroviral therapy (ART).

A significant number of these children will require surgery for elective or emergency procedures, either directly related or unrelated to HIV. For this reason, it is essential for us to fully understand the implications for anaesthesia. In order to safely anaesthetise an HIV-infected child, we require a reasonable understanding of HIV infection: its pathophysiology, multisystem complications and the pharmacology of ART. An accurate assessment of the child with HIV can potentially impact upon your choice of anaesthetic agents, whether to use regional anaesthesia, how to manage pain, approaches to infection control and general issues surrounding peri-operative care.

PATHOGENESIS, AETIOLOGY AND CLASSIFICATION

HIV is a single-stranded RNA virus with HIV-1 and HIV-2 types and multiple subtypes recognised. These subtypes express differences in geographical prevalence as well as disease progression and transmission rates. Like other retroviruses, HIV contains the enzyme *reverse transcriptase* that enables viral RNA to be transcribed to DNA, which then becomes incorporated into the host cell genome and is able to replicate freely. Inhibition of this viral replication process is the target of ART. HIV preferentially infects T helper lymphocytes (CD4+ T cells) and leads to their progressive quantitative and qualitative destruction. This makes the host increasingly immunocompromised, and thus more susceptible to opportunistic infections and malignancies.

More than 80% of HIV infections in children are due to vertical transmission (ie. trans-placental exposure to maternal HIV during the perinatal period). Perinatal transmission can occur during any one of three phases: in utero, during the peripartum

period, or during breastfeeding. The risk of perinatal acquisition without intervention (such as maternal ART, caesarean delivery, avoidance of breast milk) is 25-40%. Other routes of transmission, as for adults, are sexual or via contaminated blood through the administration of blood products, organ donations or by sharing contaminated needles during intravenous drug use.

The Centers for Disease Control and Prevention (CDC) classification system (Table 1) clearly shows the increasing severity of symptoms as HIV infection progresses.

MULTISYSTEM INVOLVEMENT

In order to perform a thorough preoperative assessment of the child with HIV, it is important that you appreciate which organ systems may be affected, either as a direct consequence of HIV infection (opportunistic infection / malignancy), or indirectly such as from side effects of ART, chemotherapy or anti-infective agents. Awareness of these effects allows us to appropriately adapt our anaesthetic.

Haematological system

The haematological system can be greatly affected during HIV infection; anaemia, neutropaenia and thrombocytopenia are common. Persistent generalised lymphadenopathy may be a feature and may also be the target of surgery for the affected child in the early stages of disease for diagnostic purposes. Haematological malignancies are often seen as are coagulation abnormalities, with obvious implications for surgical and anaesthetic interventions.

Cardiovascular system

The cardiovascular system may be affected in a number of ways in an HIV-infected child. The pericardium, myocardium or endocardium may be involved or there may be vascular lesions or neoplasms. Important and common cardiovascular complications that have major implications for patient management are:

- Pulmonary hypertension
- Pericardial effusions

Summary

It is estimated that 3.3 million children under 15 years of age are living with HIV, the vast majority of them in sub-Saharan Africa. A significant number of these children will require surgery. It is essential to understand the implications for anaesthesia in order to formulate an appropriate anaesthetic management plan.

In order to safely anaesthetise an HIV-infected child, we require a reasonable understanding of HIV infection: its pathophysiology, potential multisystem complications and the pharmacology of ART. An accurate assessment of the child with HIV may affect your choice of anaesthetic agents, how to manage pain including whether to use regional anaesthesia, approaches to infection control, and general issues surrounding peri-operative care.

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Table 1. CDC classification of paediatric HIV infection

Category	Associated symptoms
N - Asymptomatic	No symptoms 1 category A symptom only.
A - Mild symptoms	2 or more of the following symptoms: Lymphadenopathy Hepatomegaly Splenomegaly Dermatitis Parotitis Recurrent or persistent upper respiratory infection, sinusitis, or otitis media.
B - Moderate symptoms	Anaemia, neutropenia, or thrombocytopenia Bacterial meningitis, pneumonia, or sepsis Candidiasis, oropharyngeal (thrush), Cardiomyopathy Cytomegalovirus infection, with onset before 1 month Diarrhoea, recurrent or chronic Hepatitis Herpes simplex virus (HSV) stomatitis, recurrent HSV bronchitis, pneumonitis, or oesophagitis Herpes zoster (shingles) Leiomyosarcoma Lymphoid interstitial pneumonia (LIP) or pulmonary lymphoid hyperplasia complex Nephropathy Nocardiosis Persistent fever Toxoplasmosis Varicella, disseminated (complicated chickenpox).
C - Severe symptoms:	Serious bacterial infections, multiple or recurrent Candidiasis, oesophageal or pulmonary Coccidioidomycosis, disseminated Cryptococcosis, extrapulmonary Cryptosporidiosis or isosporiasis Cytomegalovirus disease Encephalopathy Herpes simplex virus infection Histoplasmosis, disseminated Kaposi's sarcoma Lymphoma Mycobacterium tuberculosis, disseminated or extrapulmonary Mycobacterium, other species disseminated Mycobacterium avium complex or Mycobacterium kansasii Pneumocystis carinii pneumonia Progressive multifocal leukoencephalopathy Salmonella (nontyphoid) septicemia, recurrent Toxoplasmosis of the brain.

- Endocarditis and valvular lesions
- Vasculitis
- Cardiomyopathies.

Respiratory system

Both the upper and lower airway can be involved in HIV infection in children, and respiratory symptoms are a common presenting complaint. These include:

- Upper respiratory tract infections (chronic otitis media)
- Bacterial pneumonia
- Atypical infections (commonly tuberculosis, non-tuberculous mycobacteria, and fungal infections)
- Lymphocytic interstitial pneumonia (LIP)
- Bronchitis
- Sinusitis
- Airway obstruction.

Gastrointestinal system

Commonly encountered complications of the gastrointestinal tract associated with HIV infection and its treatment can have major effects on the metabolism of our anaesthetic agents and any other drugs used. These effects include:

- Poor nutritional status with resultant delayed healing times
- Difficulty or pain on swallowing
- Increased gastric emptying times.

Acute or chronic diarrhoea is common, with associated dehydration and electrolyte dysfunction and in addition, there can be hepatobiliary and pancreatic impairment.

Renal system

Acute and chronic renal disease can be associated with HIV. Potential causes of renal impairment include:

- Drug induced nephrotoxicity
- Hypertension
- Diabetes
- HIV-associated nephropathy.

It is important to avoid nephrotoxic drugs where possible in these children and to dose adjust renally excreted drugs and adequately hydrate the child in order to prevent further renal damage.

Neurological system

HIV can affect the neurological system directly (HIV encephalopathy) or indirectly via opportunistic infections, neoplasms or immune deficiency. These can involve all structures including the meninges, brain, spinal cord, peripheral nerve or muscle. Neurocognitive impairment, developmental delay, encephalopathy, autonomic

neuropathy and seizures are all recognised complications of HIV which have important implications in our clinical management.

Full neurological examination pre-operatively with appropriate documentation is essential especially if you plan to use regional anaesthesia.

Endocrine and metabolic system

ART has common endocrine side effects including lipodystrophy, insulin resistance (metabolic syndrome), hypothalamic-pituitary-adrenal axis dysfunction, hypo- or hyperthyroidism and lactic acidosis.

ANTIRETROVIRAL THERAPY

The use of a combination ART or highly active antiretroviral therapy (HAART) has been a major advance in the treatment of HIV infection. These drugs are classified into the following five classes according to the mechanisms of inhibition of viral replication (Table 2):

- Reverse transcriptase enzyme inhibitors
- Protease enzyme inhibitors
- Integrase inhibitors
- Entry inhibitors
- Fusion inhibitors.

Adherence to antiretroviral therapy is of paramount importance; adherence levels below 95% are associated with increases in viral load and drug resistance. This naturally has implications for interruption of ART due to perioperative fasting. Fasting times should be kept to an absolute minimum.

Many adverse side effects are associated with ART and you should look for these during preoperative assessment. They can be divided broadly into four groups:

- **Allergic reactions:** skin rashes and hypersensitivity responses
- **Bone marrow suppression:** anaemia, neutropaenia and thrombocytopaenia.
- **Metabolic abnormalities:** fat maldistribution and change in body habitus, dyslipidaemia, hyperglycaemia and insulin resistance, bone disorders e.g. osteopaenia, osteoporosis and osteonecrosis
- **Mitochondrial dysfunction:** lactic acidosis, hepatic toxicity, pancreatitis, peripheral neuropathy.

Anaesthetic drugs may interact with ART. Anaesthetic agents may induce pharmacodynamic changes to affect their efficacy and toxicity via cytochrome p450 induction/inhibition, and the pharmacokinetic effects of ART can affect the absorption, distribution, metabolism and elimination of anaesthetic and analgesic drugs.

Pharmacodynamic interactions can be managed by avoiding anaesthetic agents such as halothane or methoxyflurane that cause

hepatic or renal dysfunction. Propofol and NRTIs may both potentially promote mitochondrial toxicity and lactic acidosis, so it may be wise to avoid propofol infusions in patients receiving ART due to the already potentially compromised mitochondrial function.

Pharmacokinetic interactions are more complex and are primarily due to liver enzyme induction or inhibition, particularly of the CYP450 3A4 enzyme. Protease inhibitors (PIs) and NNRTIs are the most commonly implicated groups of drugs. Enzyme induction or inhibition can affect the action of several classes of anaesthetic drugs:

- **Opioids.** The effects of fentanyl may be enhanced by ritonavir due to both liver enzyme inhibition and induction. Enzyme inhibition reduces fentanyl clearance and enzyme induction increases metabolism to active metabolites such as normeperidine.

- **Benzodiazepines** Saquinavir may inhibit midazolam metabolism.
- Local anaesthetics such as lignocaine may have increased plasma levels due to enzyme inhibition with direct implications for toxic doses.
- Neuromuscular blocker effects may be prolonged, even after a single dose.
- Calcium channel blockers may have enhanced hypotensive effects due to enzyme inhibition.

These interactions are complicated and multiple and databases exist that describe these interactions in detail (such as www.hiv-druginteractions.org). Evidence for interactions specifically with anaesthetic drugs is relatively sparse.

Table 2. ART

Drug class	Available drugs
1. Reverse transcriptase inhibitors	
Nucleoside/nucleotide analogues (NRTIs)	Abacavir (ABC) Didanosine (ddI) Emtricitabine (FTC) Lamivudine (3TC) Stavudine (d4T) Zidovudine (AZT, ZDV)
Non-nucleotide reverse transcriptase Inhibitors (NNRTIs)	Delavirdine (DLV) Efavirenz (EFV) Etravirine (ETR) Nevirapine (NVP)
2. Protease inhibitors (PIs)	Atazanavir (ATV) Darunavir (DRV) Fosamprenavir (FPV) Indinavir (IDV) Lopinavir (LPV) Nelfinavir (NFV) Ritonavir (RTV) Lopinavir/ritonavir (LPV/r) Saquinavir (SQV) Tipranavir (TPV) Amprenavir (APV)
3. Integrase inhibitors	Raltegravir (RAL) Dolutegravir
4. Entry inhibitors	
CCR 5 antagonists	Maraviroc (MVC)
5. Fusion inhibitors	Enfuvirtide (ENF, T-20)

Due to the risk of developing resistance if doses are missed, it is recommended that ART be continued throughout the perioperative period if at all possible. Naturally this needs to be compatible with the proposed surgery and with the patient's gastrointestinal function. Some drugs are available in liquid form enabling administration via nasogastric feeding tube or gastrostomy. Parenteral preparations are limited to zidovudine and enfuvirtide only.

REGIONAL ANAESTHESIA

The presence of HIV infection is not an absolute contraindication to regional anaesthesia and there is no evidence that HIV progression is increased by central neuraxial blockade. Regional anaesthesia may be relatively contraindicated with several pathologies associated with HIV infections such as myelopathy, vertebral or spinal neoplasms, CNS infections and coagulopathies, as well as in the severely immunocompromised patient. You must conduct a full preoperative neurological assessment and document any pre-existing neurological deficit if you plan a regional anaesthetic technique.

BLOOD TRANSFUSION

There is evidence that allogeneic blood transfusion in the HIV infected patient can lead to transfusion-related immunomodulation (TRIM) and can result in an increase in HIV viral load. Blood should therefore only be transfused where unavoidable to maintain patient safety.

PAIN

Pain is common in advanced HIV disease and can be very difficult to treat. The aetiology of this pain can be multifactorial, including:

- Opportunistic infections such as herpes simplex
- HIV-related arthralgia
- Peripheral neuropathy
- Drug-related pain.

Pre-existing pain and its treatment can affect the treatment of postoperative pain and will necessitate a multimodal approach.

INFECTION CONTROL

Infection control is an important issue both for the protection of the immunocompromised child from opportunistic infections, as well as for the professionals caring for the child during any procedures. Use a strict aseptic technique for all procedures.

The cumulative risk of contracting HIV over an anaesthetic career can be as high as 4.5%. This can occur due to a needlestick injury (transmission risk of 0.3%); the risk is increased if there is a higher volume of blood injected, such as with hollow needles or deep punctures. Risk of transmission via the mucocutaneous route (splashing of a mucosal surface or broken skin by body fluid) is extremely low (0.03%).

There are universal precautions that should be taken to reduce the risk of HIV transmission to healthcare workers:

- Dispose of sharps safely (into a rigid locking container prior to incineration)
- Do not re-sheath needles
- Wear gloves
- Cover any cuts or broken skin with an impermeable dressing
- Strongly consider eyeshields and facemasks
- Use disposable equipment where possible; clean reusable equipment properly.

Post-exposure prophylaxis for healthcare workers should be available and should commence as soon as possible after potential high risk exposure (ideally within the first 1-2hrs).

If tuberculosis is suspected or likely in a patient known to have HIV, all healthcare workers should wear a tight fitting facemask to reduce the risk of transmission (ideally a high quality particulate mask if available e.g. N95 or HEPA).

APPROACH TO FORMULATING AN ANAESTHETIC MANAGEMENT PLAN FOR THE HIV-INFECTED CHILD

A multisystem and multidisciplinary approach is recommended.

Thorough preoperative assessment for status of HIV infection:

- History, including risk factors (for child & parents)
- Physical examination
- Laboratory tests
- Assess organ involvement
- Drug history and side effects.

Investigations ideally include:

- Full blood count
- Clotting function to exclude coagulation abnormalities (consider use of TEG/platelet mapping if available)
- Biochemical tests including glucose, electrolytes, renal & liver function to exclude possible metabolic, liver or renal disturbances
- Viral load and CD4 count
- Chest radiography to screen for opportunistic infections and tuberculosis
- Cardiac evaluation with electrocardiography and echocardiography (if possible) to screen for cardiomyopathy.

Preparation of operating theatre and personnel:

- Infection control preparation including universal precautions with gloves, aprons, visors available

- Sharp object collection devices with appropriate sharps handling (no re-sheathing of needles)
- Staff fully aware of protocols in the event of occupational exposure
 - Rinse and wash affected area with soap & water
 - Recipient lab tests: HIV, acute hepatitis panel
 - Determine infectious status of source
- Availability of post exposure prophylaxis to be started as soon as possible following accidental high risk exposure (ideally within 1 hour of exposure)
 - HIV PEP protocol with 3 or more antiretroviral drugs if known HIV positive donor or high-risk patient or with 2 or more if low risk. ART is given for 4 weeks or until source is found to be negative for HIV
 - Follow up with counselling and HIV testing for at least 6 months post exposure (tests done at baseline, 6 weeks, 12 weeks and 6 months)
 - Hepatitis B immune globulin +/- hepatitis B vaccine
 - Achieve early identification of chronic hepatitis C disease.

Perioperative considerations for the child with HIV:

- Minimise interruptions in ART as far as possible to diminish the risk of developing drug resistance
- Consider drug interactions between ART and drugs affected by hepatic enzyme inhibition and/or induction
- Exercise strict aseptic technique as HIV infected children are immunocompromised and are susceptible to opportunistic infections
- Additional emotional and psychological support may be necessary as primary caregivers may have been affected by HIV/AIDS
- The anaesthetic plan should of course be tailored to the individual child and the type of surgery to be undertaken.

REFERENCES AND FURTHER READING

Bayer R: Discrimination, informed consent, and the HIV infected clinician. *British Medical Journal* 1997; **314**: 915-6.

Centers for Disease Control and Prevention. HIV/AIDS Surveillance Report 2004. Vol. 16. Atlanta, Ga: US Department of Health and Human Services, Centers for Disease Control and Prevention; 2005. 1-46.

Diprose P, Deakin CD and Smedley J. Ignorance of post-exposure prophylaxis guidelines following HIV needlestick injury may increase the risk of seroconversion. *British Journal of Anaesthesia* 2000; **84**: 767-70.

Horlocker TT and Wedel DJ. Regional Anaesthesia in the Immunocompromised Patient. *Regional Anaesthesia and Pain Medicine* 2006; **31**: 334-45.

Joint WHO/United Nations programme on HIV/AIDS; 2008 report on global AIDS epidemic. www.unaids.org

Kharasch ED, Mitchell D, Coles R and Blanco R. Rapid clinical induction of hepatic cytochrome P4502B6 activity by ritonavir; . *Antimicrobial Agents and Chemotherapy* 2008; **52**:1663-9.

Kuczkowski KM. Anaesthetic considerations for the HIV-infected pregnant patient. *Yonsei Medical Journal* 2004; **45**:1-6.

Leelanukrom R: Anaesthetic considerations of the HIV-infected patients. *Current Opinion in Anaesthesiology* 2009, **22**: 412-418.

Leelanukrom R and Pancharoen C. Anaesthesia in HIV-infected children. *Pediatric Anaesthesia* 2007; **17**: 509-19.

Parthasarathy S and Ravishankar M: HIV and anaesthesia; *Indian Journal of Anaesthesia* 2007; **51**: 91-99.

Prout J and Agarwal B: Anaesthesia and critical care for patients with HIV infection. *Continuing Education in Anaesthesia Critical Care and Pain* 2005; **15**: 153-6.

Schwartz D, Schwartz R, Cooper E and Pullerits J: Anaesthesia and the child with HIV infection. *Canadian Journal of Anaesthesia* 1991; **38**: 626-33.

Wilson S: HIV and Anaesthesia. *Update in Anaesthesia* 2010; **25**: 25-9. Available at: http://www.wfsahq.org/components/com_virtual_library/media/25b2de5bdb19692a50cc78dbbbab9a9c-HIV-and-Anaesthesia--Update-25-1-2009-.pdf

World Health Organization. Global summary of the AIDS epidemic: 2009. Available at http://www.who.int/hiv/data/2009_global_summary.png.

Anaesthesia for non-cardiac surgery in children with congenital heart disease

Based in part upon: Story E. Recognising cardiac disease in children *Anaesthesia Tutorial of the Week* 93 (2008)¹

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SUMMARY

Children with congenital heart disease provide a challenge to the anaesthetist, but with careful planning, most can be anaesthetised safely. This article covers pathophysiology, recognition, and principles of anaesthesia for children with congenital heart disease, including anaesthesia for specific cardiac lesions.

INTRODUCTION

The risk of perioperative complications is higher in children with congenital heart disease (CHD) for both minor and major surgery, particularly in neonates and those with complex lesions.² Although CHD is uncommon, it is important for the anaesthetist to understand how to recognise a child with CHD, and the principles of anaesthetic management for children with both unrepaired and repaired lesions.

INCIDENCE OF CONGENITAL HEART DISEASE

The incidence of CHD is approximately 8:1000 live births. The frequency of different lesions varies between populations, with ventricular septal defect (VSD) being the most common in all populations (see Table 1).

Most CHD is untreated in low- or middle-income countries (LMIC). Babies with complex cardiac

disease are likely to die due to lack of access to corrective or palliative surgery, in particular those with duct dependent lesions. A child with untreated heart failure due to high pulmonary blood flow is more likely to die from pneumonia, which is the most common cause of mortality in the under 5 age group. If a child with high pulmonary blood flow remains untreated they may develop Eisenmenger's syndrome, which is associated with high mortality in childhood or early adult life.

However, as surgical systems improve, there are an increasing number of programmes to treat children with CHD in LMIC, many developed in partnership with international outreach teams, so that the outlook for children born with simple conditions such as VSD, atrial septal defect (ASD), patent ductus arteriosus (PDA), coarctation of the aorta or Tetralogy of Fallot is much more optimistic.

Table 1. The incidence of different types of congenital heart disease in children in the UK

Condition	Incidence
Ventricular septal defect (VSD)	32%
Patent arterial duct (PDA)	12%
Pulmonary stenosis (PS)	8%
Coarctation of the aorta (CoA)	6%
Atrial septal defect (ASD)	6%
Tetralogy of Fallot (TOF)	6%
Aortic stenosis (AS)	5%
Transposition of the great arteries (TGA)	5%
Hypoplastic left heart syndrome (HLHS)	3%
Hypoplastic right heart syndromes	2%
Atrioventricular septal defects (AVSD)	2%
Truncus arteriosus (common arterial trunk)	1%

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CAUSE OF CONGENITAL HEART DISEASE AND ASSOCIATED ABNORMALITIES

The cause of most CHD is unknown, but may be related to chromosomal abnormalities, exposure of the mother to teratogens (e.g. alcohol, or drugs such as anti-epileptics or warfarin), or maternal disease (rubella, diabetes).

Recognisable chromosomal abnormalities are present in 25% of children with CHD. The diagnosis of a chromosomal abnormality should lead to assessment of the child for CHD. The most common chromosomal abnormality is Down syndrome (trisomy 21) – 40% of children with Down syndrome have CHD, most commonly atrioventricular septal defect (AVSD) or VSD. Chromosomal abnormalities associated with cardiac lesions are shown in Table 2.

PATHOPHYSIOLOGY OF CONGENITAL HEART DISEASE

It is useful to classify CHD according to the pathophysiology of the major heart lesion, in particular, whether the child is acyanotic (i.e. the child is 'pink'), or the lesion is associated with cyanosis (i.e. the child is 'blue'). Most CHD is acyanotic. Measuring oxygen saturation by pulse oximetry shortly after birth is a useful screening test to identify cyanotic lesions such as tetralogy of Fallot, transposition of the great arteries (TGA), total anomalous pulmonary venous drainage (TAPVD) or duct dependent lesions such as hypoplastic left heart syndrome. However, the most common cause of cyanosis in the neonatal period is due to a respiratory problem. A description of common lesions is shown in Table 3.

Left to right shunting lesions: ASD, VSD, AVSD, PDA

The child with acyanotic heart disease presents with signs and symptoms of cardiac failure. The severity of the symptoms depends on the volume of the shunt:

- *Atrial septal defect, ASD.* (See Figure 1). A secundum ASD is associated with a low volume left to right shunt with right heart overload. The child may present with recurrent chest infections, failure to thrive, or may be asymptomatic.
- *Ventricular septal defect, VSD.* A 'restrictive VSD' is small, and causes a low volume shunt with few symptoms. An 'unrestrictive VSD' is large, associated with a high volume left to right shunt and symptoms of severe cardiac failure in infancy
- *Atrioventricular septal defect, AVSD.* (See Figure 2). There is an abnormal atrioventricular valve, an atrial septal defect ('primum ASD'), +/- a ventricular septal defect. This results in a high volume shunt and signs of severe cardiac failure in infancy leading to early pulmonary hypertension. Untreated, children with AVSD develop Eisenmenger's syndrome (see later) by the age of 1-2 years, with 4% survival beyond 5 years.
- *Patient ductus arteriosus, PDA.* (See Figure 3). If the duct is restrictive the child may present with relatively few symptoms initially, or signs and symptoms of cardiac failure. Premature neonates with a large unrestrictive PDA may have severe cardiac failure and be ventilator dependent.

Table 2. Chromosomal abnormalities associated with cardiac lesions

Abnormality	Clinical features and associated conditions	Typical cardiac lesion
Down syndrome Trisomy 21	Typical facial appearance, single palmar crease, short stature, learning difficulties, lax joints (including cervical spine instability), hypothyroidism, obstructive sleep apnoea, leukaemia, duodenal atresia.	AVSD, VSD
DiGeorge syndrome ('Catch 22') 22q11 deletion	Learning difficulties, cleft palate, hypocalcaemia, absent thymus (frequent respiratory infections), typical facial appearance	Aortic arch abnormalities, VSD
Marfan's syndrome	Abnormally tall stature, long fingers, scoliosis, abnormal shaped chest, high arched palate, retinal detachment, inguinal hernia, spontaneous pneumothorax	Aortic root dilatation and dissection
Apert syndrome	Craniosynostosis (premature fusion of cranial sutures), syndactyly (fused fingers), deafness	PS, VSD
Charge association	Abnormal iris (coloboma), choanal atresia (abnormal nasal passageway), developmental delay, abnormal genitalia, ear deformity	Variety, including VSD, AVSD
VATER	Vertebral abnormalities, anal atresia, tracheo-oesophageal fistula, renal abnormalities	VSD, TOF
Goldenhar syndrome	Hemifacial microsomia (poorly developed maxilla/mandible), difficult intubation, ear abnormalities, cleft palate	VSD, TOF

Table 3. Classification of common congenital heart lesions

Acyanotic congenital heart disease – ‘pink’ babies		
	Common examples	Comment
Left-to-right shunt ‘Restrictive’ lesions ‘Non-restrictive’ lesions	Small ASD, VSD, PDA Large VSD, PDA, AVSD, common arterial trunk	Large (‘non-restrictive’) lesions are associated with severe congestive cardiac failure in infancy. If unrepaired, may lead to pulmonary hypertension and reversal of shunt (Eisenmenger’s syndrome).
Obstructive lesions	Aortic stenosis, coarctation of the aorta, pulmonary stenosis	Severity of lesion determines age at presentation – neonates with severe obstruction may be critically ill with a duct dependent circulation
Cyanotic lesions – ‘blue’ babies		
Right-to-left shunt	Tetralogy of Fallot: • VSD with aortic override • Right ventricular outflow tract obstruction • Right ventricular hypertrophy,	May present with severe cyanosis and hypercyanotic ‘spells’; or if unrepaired in an older child, with cyanosis, fatigue and a history of ‘squatting’.
Transposition of the Great Arteries (TGA)	TGA may be associated with ASD, VSD, PDA	Long term survival requires intervention in early infancy (arterial switch operation)
Single ventricle physiology	Hypoplastic left heart syndrome (HLHS), hypoplastic right heart syndrome, tricuspid atresia.	Duct dependent circulation; survival requires intervention in the neonatal period
Common mixing	Total anomalous pulmonary venous drainage (TAPVD).	Present with heart failure and/or cyanosis – survival requires intervention in the neonatal period

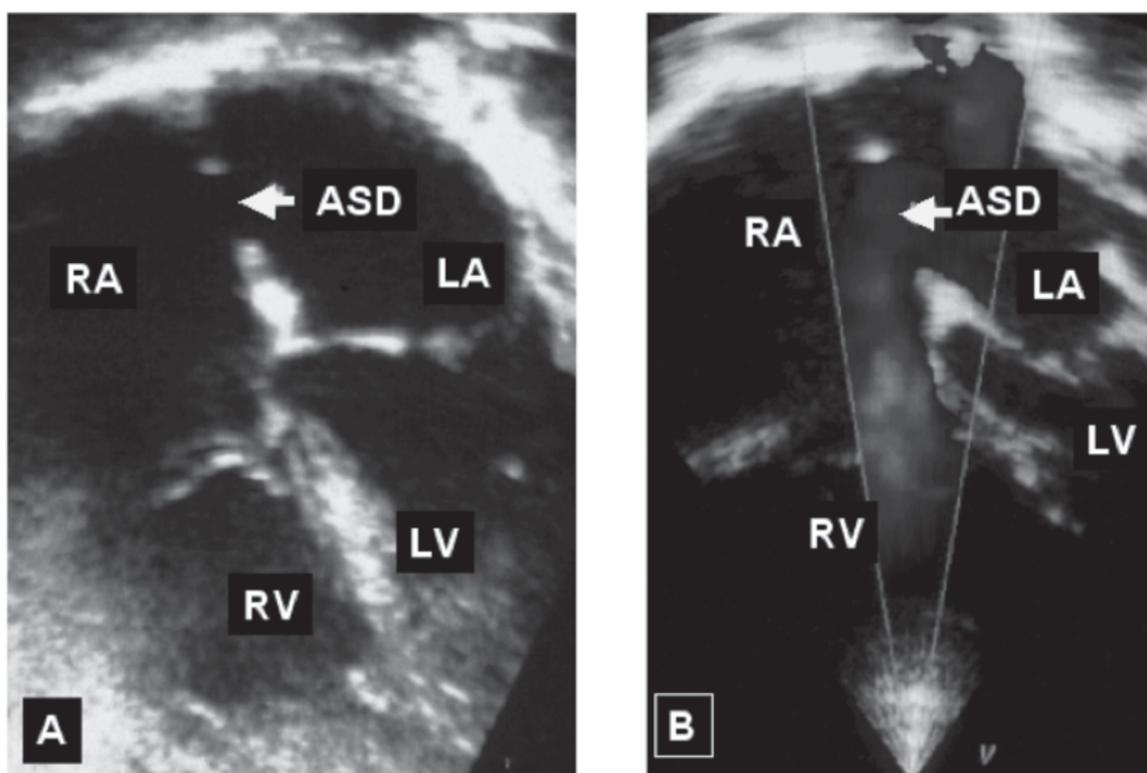


Figure 1. Echocardiographic appearance of secundum ASD with left-to-right flow

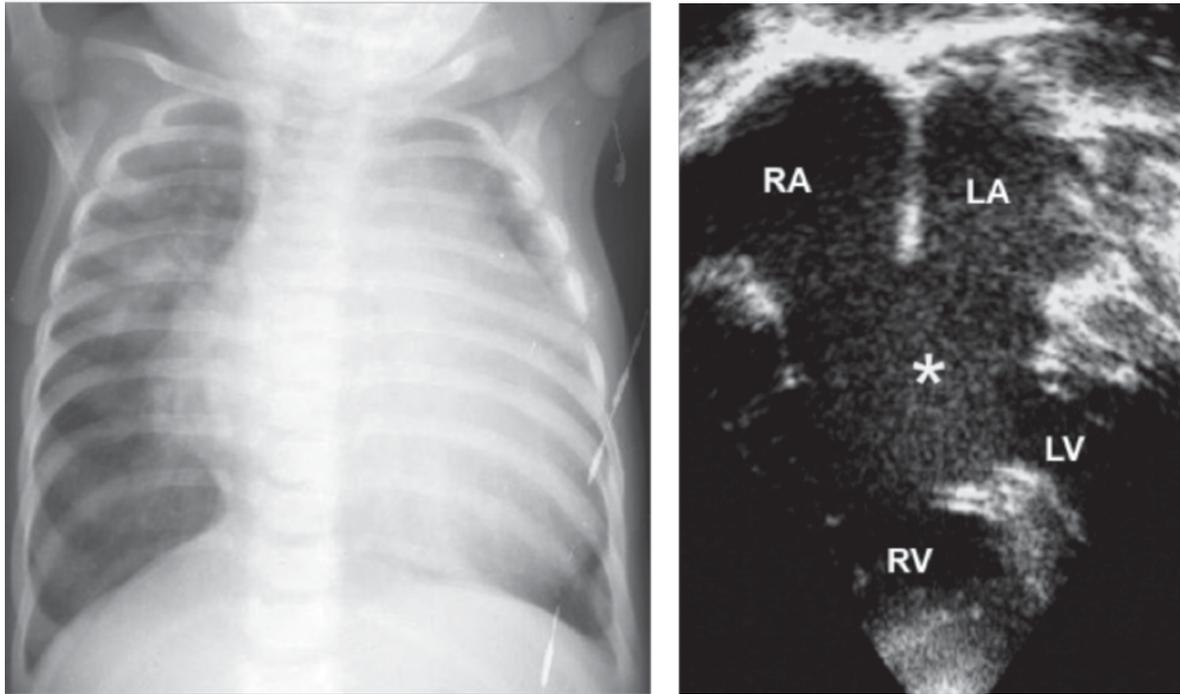


Figure 2. CXR and echocardiographic appearance in a child with heart failure secondary to AVSD

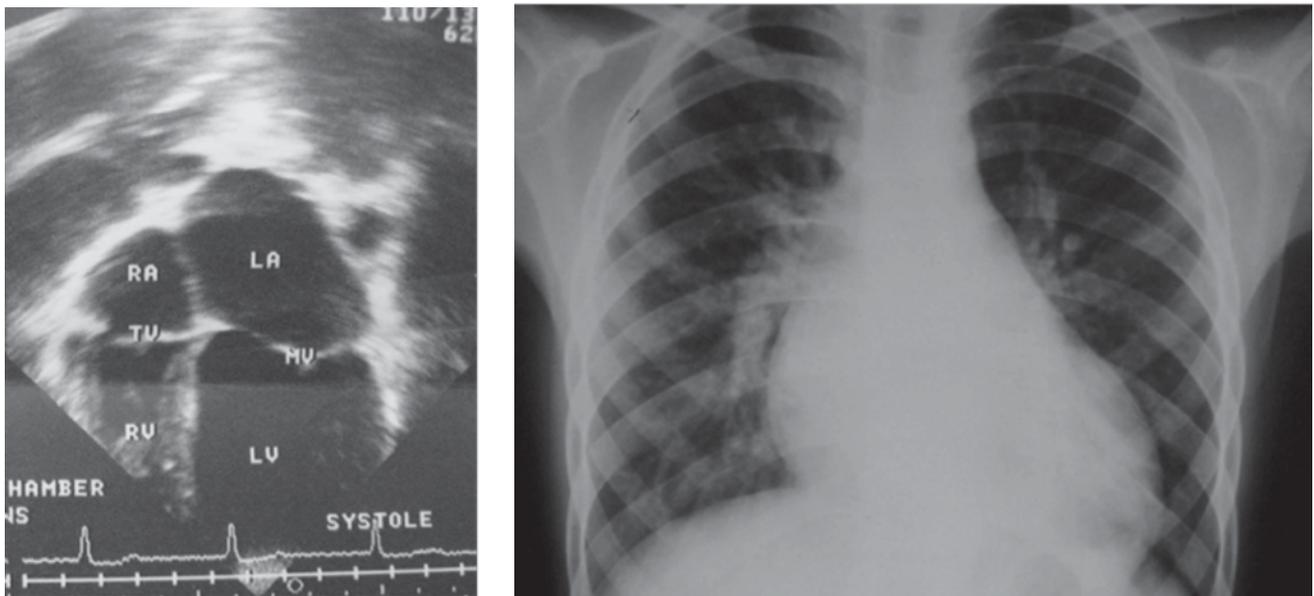


Figure 3. Echocardiographic appearance and CXR in a child with a PDA, showing left heart volume overload and prominent pulmonary vasculature

Tetralogy of Fallot

Tetralogy of Fallot (TOF) is associated with a right to left shunt. The abnormality in TOF is due to:

- VSD with aortic override.
- Right ventricular outflow tract obstruction (RVOTO) (muscle bundles below the pulmonary valve; valvar or supra-valvar obstruction).
- Right ventricular hypertrophy due to the RVOTO.

The child presents with cyanosis from birth and may develop cyanotic 'spells' in infancy, due to spasm of muscle bundles in the right ventricular outflow tract in response to adrenergic stimulation, for instance when the child is upset or cries. An older child who has not had corrective surgery may 'squat' when tired to increase pulmonary blood flow (by increasing systemic vascular resistance), and will have clubbing. (See Figure 4).

A modified Blalock-Taussig (mBT) shunt may be placed as a palliative procedure in the first few weeks of life to provide a secure



Figure 4. Finger clubbing in a child with Tetralogy of Fallot

blood supply to the lungs before corrective surgery is performed. This is an arterial shunt between the innominate artery and the right pulmonary artery. The shunt is 'restrictive' so that the pulmonary blood supply is adequate, but not too high; the ideal SpO₂ is around 85%. If the shunt is too large (unrestrictive), the child will have too high a pulmonary blood flow, causing signs of cardiac failure, with low systemic pressure due to excessive 'runoff' from the innominate artery to the lungs.

Surgery for TOF should ideally be at a few months of age. Complete correction involves closure of the VSD and relief of the obstruction at the right ventricular outflow tract. The surgical course may be complicated, but the saturation will be normal after surgery and the long-term outlook is usually good. Some patients may develop pulmonary regurgitation, right heart dilation and arrhythmias if a patch has to be placed across the pulmonary annulus.

Duct dependent circulation

In utero, the placenta is the main site for gas exchange for the developing foetus and the blood flow to the foetal lung is minimal. Blood from the right ventricle bypasses the lungs and passes directly to the aorta via a foetal vessel called the arterial duct. After birth, a number of changes occur in transition from the foetal to the newborn circulation, one of which is closure of the arterial duct so that blood now perfuses the lungs. (See Basic Science article, this edition of *Update*, page 4)

There are certain severe congenital cardiac lesions that are only compatible with life if the arterial duct remains open – the circulation is said to be 'duct dependent'. If undiagnosed at birth, these babies typically present with acute collapse as the duct closes within the first 5 days of life (the differential diagnosis is septic shock).

Examples of duct dependent circulations:

- Critical coarctation of the aorta (duct dependent systemic circulation). There is extreme narrowing of the aorta where the arterial duct joins the aorta, and blood supply to the lower half of the body is only possible if blood passes from

the pulmonary artery to the descending aorta via the duct. The blood in the pulmonary artery is deoxygenated, so the oxygen saturation in the feet ('post-ductal') will be lower than the oxygen saturation in the right hand ('pre-ductal')

- Pulmonary atresia (duct dependent pulmonary circulation). In pulmonary atresia, the only blood supply to the lungs is that which passes from the aorta to the pulmonary artery via the duct.

Continued survival of these babies requires infusion of prostaglandin E1 to keep the duct open until urgent cardiac surgery can be performed.

Eisenmenger's syndrome

A large ('unrestricted') VSD allows blood to pass from the left ventricle to the right ventricle ('left-to-right shunt'), which results in high pulmonary blood flow and congestive cardiac failure. The normal physiological response to high pulmonary blood flow is for the resistance in the pulmonary vessels ('pulmonary vascular resistance', PVR) to increase. With time, the PVR rises until eventually it exceeds the systemic vascular resistance, and the flow across the VSD reverses - this is known as Eisenmenger's syndrome. Clinically, this is associated with an initial improvement in symptoms of cardiac failure as pulmonary blood flow reduces, followed by increasing cyanosis as the shunt reverses to become right-to-left. Surgical closure of the shunt is not possible at this stage as the resistance to flow through the pulmonary circulation is too high and right ventricular failure will occur. Individuals with Eisenmenger's syndrome are deeply cyanosed with clubbing of fingers; they may develop haemoptysis, endocarditis or cerebral abscess, and will eventually die from cardiac failure.

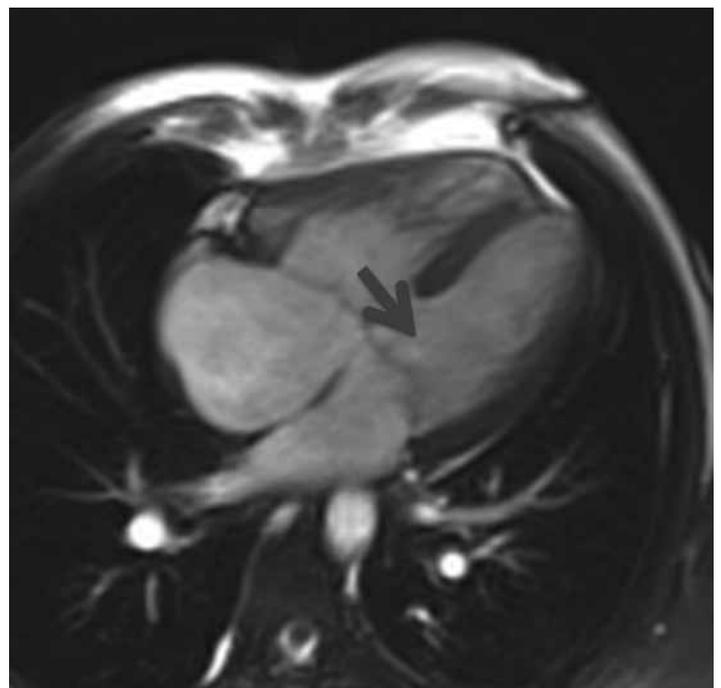


Figure 5. Cardiac MRI of patient with AVSD and right-to-left shunt in Eisenmenger's syndrome

Corrective cardiac surgery must be undertaken before the onset of pulmonary vascular disease to avoid Eisenmenger's syndrome. The ideal age for surgery is critically dependent on the underlying lesion. Eisenmenger's syndrome occurs before the age of 1 year in children with very high pulmonary blood flow, such as unrepaired AVSD, but may occur at 40-50 years in adults with unrepaired ASD who have had moderately increased pulmonary blood flow over many years. (See Figure 5).

RECOGNISING CONGENITAL HEART DISEASE IN CHILDREN

It is important to recognise the child with CHD so that they can be prepared for surgery:

- Treat cardiac failure (e.g. VSD, AVSD).
- Recognise a condition that may be associated with acute decompensation during surgery (hypercyanotic 'spell' in Tetralogy of Fallot; loss of cardiac output in aortic stenosis, coarctation; cardiomyopathy; pulmonary hypertensive crisis in Eisenmenger's syndrome).
- Ensure antibiotic prophylaxis is given to children at risk of endocarditis.

Children rarely present with the symptoms classically associated with heart disease in adults (i.e. chest pain, shortness of breath, swollen ankles); they usually present with symptoms such as failure to thrive, frequent chest infections, or unexplained 'funny turns'. A careful history and examination is key, as are special investigations such as CXR, ECG, SpO₂, echocardiogram, or occasionally cardiac catheterisation. Cardiac MRI is increasingly used.

History

Ask about the following:

Pregnancy: Maternal disease, drug and alcohol intake

Birth history: A history of prematurity is associated with PDA. Birth asphyxia is associated with persistent foetal circulation (persistent pulmonary hypertension of the newborn, PPHN)

Cardiac symptoms

- Cyanosis - central cyanosis is an important cardiac symptom that is difficult to detect and may often be missed by parents. Central cyanosis is seen as blue discoloration of the tongue and lips and may be confirmed using pulse oximetry.
- Hypercyanotic 'spell'. This is a classic symptom of Tetralogy of Fallot (see Table 3). The child may become deeply cyanosed when upset or crying, and may become limp and unresponsive; this is a sign of acute reduction in pulmonary blood flow associated with sudden increase in the dynamic obstruction to the right ventricular outflow tract. Older children with uncorrected TOF may 'squat' to increase pulmonary blood flow – this position increases the systemic vascular resistance and reduces the right-to-left shunt across the VSD. Cyanotic spells are uncommon

in the newborn. Older babies may be placed in the knee to chest position in response to a cyanotic spell. The differential diagnosis for episodic cyanosis is fits or respiratory problems such as croup or asthma.

Respiratory symptoms. Breathlessness due to increased pulmonary blood flow is a common respiratory symptom in children with cardiac failure, for instance due to a large VSD or AVSD. In babies, this presents as slow feeding, breathlessness, cold clammy sweatiness and poor weight gain. An older child may have limited exercise tolerance and not keep up with their peers. Frequent respiratory tract infections and poor weight gain are common in older children with an ASD, although there may not be overt breathlessness.

Funny turns and chest pain – funny turns are an unusual presentation for cardiac disease in children, more commonly associated with simple faints, or neurological disease such as epilepsy. Sudden collapse may be due to arrhythmias, and collapse with exercise is a very worrying sign in a child with significant left ventricular outflow tract obstruction such as aortic stenosis. Most chest pain in children is due to musculoskeletal problems, especially in older children. Coronary artery abnormalities, and hence chest pain due to angina, is rare. A young infant with angina due to Kawasaki disease or Anomalous origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) may present with 'quiet' episodes associated with reduced activity and sweatiness – more commonly with breathlessness and listlessness due to left ventricular failure.

Poor weight gain – this is common in conditions causing heart failure or associated with increased pulmonary blood flow such as VSD. Older children with acquired heart disease such as endocarditis or cardiomyopathy may have anorexia and weight loss. Conditions associated with cyanosis and reduced pulmonary blood flow such as TOF are not usually associated with poor weight gain.

General enquiry – this may reveal other symptoms suggestive of a complex congenital disorder such as Down's syndrome, a family history of cardiac disease, or symptoms suggestive of acquired heart disease such as rheumatic fever or endocarditis.

Examination

Examine the child using the standard routine of inspection, palpation, percussion and auscultation:

- Look for dysmorphic features, for instance suggestive of Down's syndrome, or the presence of an associated congenital abnormality such as cleft palate or spinal deformity.
- Signs of poor weight gain and failure to thrive should be sought (weigh the child and compare to standard growth charts).
- Signs of breathlessness will be apparent as increased respiratory rate, and in the infant, intercostal and sub-costal recession, nasal flaring and grunting. In an older child, chest deformity may be associated with longstanding ventricular enlargement.

- Peripheral cyanosis is common in children at any age and is not important. Central cyanosis is always important but may be missed in a child with severe anaemia or children with pigmented skin. Look at the colour of the tongue – if blue, it suggests the SpO₂ is <85%. Saturation must be confirmed using a pulse oximeter. Long standing cyanosis may be associated with ‘clubbing’ of the nails of the hands and feet. Children who have long standing cyanosis develop compensatory polycythaemia and possibly complications such as cerebral thromboembolism.
- The jugular venous pulse is very difficult to see in children <5 years who have chubby necks and who move around a great deal – the liver size gives a much better estimate of venous pressure.
- Palpate the pulses to assess rate, rhythm, volume and character. Radio-femoral delay or absent femoral pulse is seen in coarctation; differential right and left radial pressures are seen in aortic arch interruption.
- A suprasternal ‘thrill’ may be felt in aortic stenosis, occasionally in pulmonary stenosis or other causes of aortic arch anomaly leading to a ‘palpable’ murmur. A palpable ‘heave’ indicates ventricular hypertrophy.
- The normal position of the cardiac apex is the 4th intercostal space inside the nipple line in a child <5 years, the 5th intercostal space at the nipple line in a child >5 years.
- Estimate the liver size by palpation. The normal neonate may have 1cm of liver palpable, an older child may have a liver edge palpable – anything more may indicate increased right atrial pressure, usually due to heart failure, or a non-cardiac cause of hepatomegaly.
- Dependent peripheral oedema is a late sign of cardiac failure in children, and may be felt by palpation over the sacrum.
- Percussion may be useful to estimate liver size and the presence of ascites (rarely due to cardiac failure in children).

Heart murmurs

Innocent cardiac murmurs

The most common murmur in children is a functional, innocent or physiological heart murmur, which is heard in 10% of normal children. The classical innocent murmur in children is known as the ‘Still’s’ murmur. Innocent murmurs may also be due to flow murmurs associated with increased cardiac output, heard in children with a fever or anaemia. The heart is structurally normal in all children with an innocent murmur. A murmur in a child may be classified as innocent if the child has no other signs or symptoms of cardiac disease, and the murmur has certain characteristic features:

- Soft (no thrill)
- Systolic and short (never pansystolic)
- Asymptomatic

- Best heard at the left sternal edge, no radiation
- Changes with posture – softer when standing.

Pathological cardiac murmurs

Cardiac murmurs associated with a left to right shunt such as a VSD may not be very obvious in the newborn period when the pulmonary vascular resistance is high. The pulmonary vascular resistance falls in the first few days of life - the loud systolic murmur due to a VSD will become apparent and the child may develop signs of heart failure as the flow increases across the shunt. A murmur is the result of turbulent flow and is graded as soft, moderate, or loud; a very loud murmur may also be felt (‘thrill’). See Table 4 and Figure 6 for description and location of pathological cardiac murmurs.

Investigations

Special investigations include the Chest Xray, ECG, echocardiography and cardiac catheterisation.

Chest Xray. Consider the age of the patient and if the film was taken in the sitting or lying position. Evaluate the chest Xray systematically:

- A - Adequacy and alignment. The film should be sufficiently penetrated to just visualise the disc spaces of the lower thoracic vertebrae through the heart shadow. At least 5 anterior rib ends should be seen above the diaphragm on the right hand side. Alignment can be assessed by ensuring that the medial ends of both clavicles are equally spaced about the spinous processes of the upper thoracic vertebrae.
- B – Bones. Check the ribs, clavicles and vertebrae. (Rib notching is sometimes seen in severe coarctation of the aorta).
- C - Cartilage and soft tissues.
- Lungs - Compare side-to-side and upper, middle and lower thirds of the chest. Look for pleural effusions, pneumothorax, vascular markings that are increased or decreased (plethoric or oligoemic), fluid in the fissures, white lung areas which could be consolidation or haemorrhage.

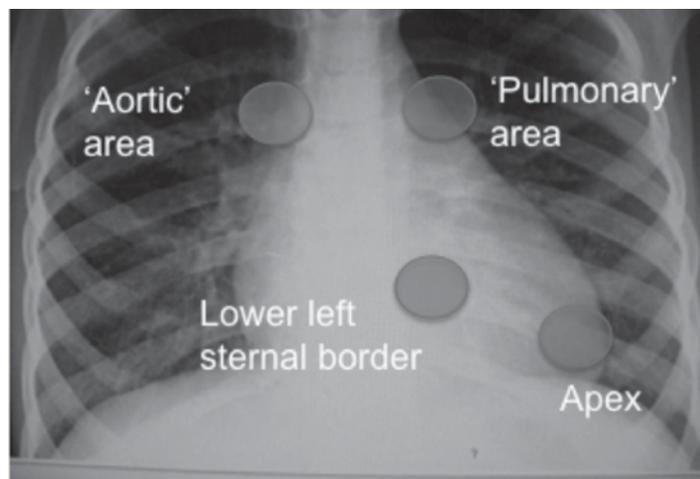


Figure 6. Best location to hear cardiac murmurs

Table 4. Pathological murmurs in children

Murmur	Location murmur heard best
Systolic murmurs	
<i>'Soft' = grade 2/6; 'Moderate' = grade 3/6; 'Loud with thrill' = grade 4/6</i>	
Ejection systolic	<i>Aortic stenosis: Upper right sternal edge +/- carotid thrill Pulmonary stenosis or ASD: Upper left sternal edge, no thrill TOF: Long harsh murmur (in a child with cyanosis)</i>
Pansystolic	<i>VSD: Lower left sternal edge +/- thrill Mitral regurgitation (NB. rheumatic fever): Apex</i>
Diastolic murmurs (unusual in children)	
<i>'Soft' = grade 2/4; 'Moderate' = grade 3/4; 'Loud' = grade 4/4</i>	
	<i>Aortic regurgitation (NB. endocarditis): Lower left sternal edge, sitting forward, collapsing pulses</i>
Continuous murmurs	
	<i>Patent ductus arteriosus (PDA): Left infraclavicular region Arteriovenous malformation: Any site (head, shoulder, lungs).</i>

- Heart: Look at the size of the heart; is it enlarged? Is the shape unusual? (See below). In normal infants the heart is up to 60% of the thoracic diameter, 50% thereafter. (See Figure 7). Remember that a normal cardiac shadow does not rule out cardiac disease.
- The upper mediastinum: In children under the age of 18 months, the normal thymus may simulate superior mediastinal widening (above the level of the carina) (See Figure 7).
- D – Diaphragms. The border between the heart and the diaphragm and between the diaphragm and the ribs (cardiophrenic and costophrenic angles) should be clear on both sides. Loss of definition of the left diaphragm behind the heart suggests left lower lobe collapse, an abnormal hump suggests diaphragmatic rupture, a hazy diaphragm suggests effusion or collapse in the bordering lung segment, and an elevated diaphragm suggests phrenic nerve palsy

There are some classical appearances of the chest Xray in children:

- 'Egg-on-side' = Transposition of great arteries in a neonate
- Boot shaped heart = Tetralogy of Fallot (right ventricular hypertrophy and reduced pulmonary markings (See Figure 8))
- 'Snowman in a snow storm' = Obstructed total anomalous pulmonary venous connection in a neonate
- Globular heart. Usually associated with pericardial effusions, may be secondary to pericarditis or dilated cardiomyopathy

- Situs. In the normal situation (situs solitus) the heart is on the left with the gastric bubble on the left and the liver on the right. In situs inversus these relations are reversed
- Oligoemic lung fields are seen in conditions associated with reduced pulmonary blood flow such as TOF and pulmonary atresia
- Plethoric lung fields are seen in children with left to right shunts, especially VSD and AVSD.

The electrocardiogram (ECG)

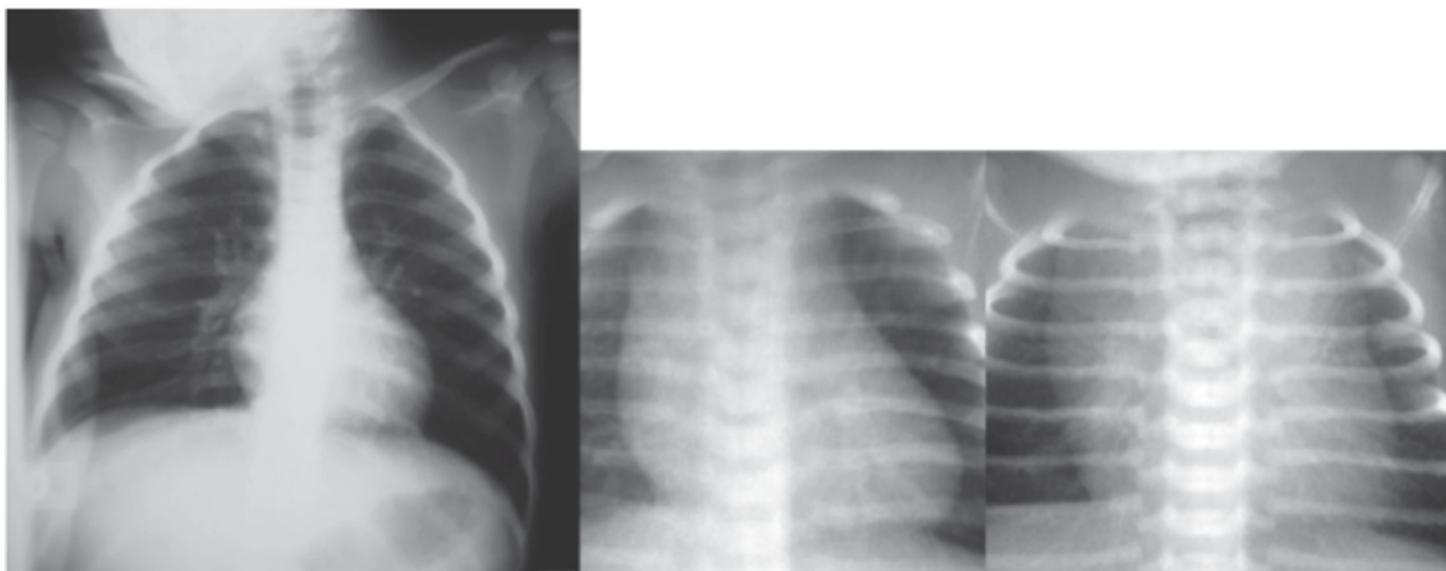
The ECG may be useful to investigate rhythm and conduction abnormalities, as well as assessing chamber hypertrophy and strain. Interpretation of the paediatric ECG is complex and must take the child's age into account, with comparison to tables of normal values.

Echocardiography

Echocardiography is a form of cardiac imaging that uses reflection of ultrasound pulses from interfaces between tissue planes. It can be used to generate detailed real time images of the cardiac anatomy. It has become the standard investigation for all patients with valvular heart disease, congenital heart disease, myocardial and pericardial disease, and to assess myocardial function.

The heart is examined in a systematic way, deciding first the orientation of the heart within the body (normal = solitus); the connections between the atria and the ventricles (normal = concordant), ventricles and major vessels (normal = concordant); and the side of the aortic arch (normal = left). The normal position and connections is often described in short hand as 'SCCL'. The echocardiologist then describes the ventricular function, the valves, the shunts, and the size of the major vessels.

Figure 7. Normal chest Xrays in children



Normal 3 year-old

Normal newborn

Doppler ultrasound may be used to estimate pressure gradients across valves and shunts using the following formula:

$$\text{Pressure} = 4 v^2, \text{ where } v \text{ is the velocity measured by Doppler m.sec}^{-1}$$

The 'z score' is used to describe the heart compared to normal values for the age of the child; if the aortic valve has a z score of -2, this suggests the valve is 2 standard deviations smaller than the normal average size.

Cardiac catheterisation

Cardiac catheterisation is used to answer specific diagnostic questions in children with congenital heart disease. A catheter can be passed into the heart chambers under Xray control to measure intracardiac pressures and oxygen saturations, or for radiological imaging by injection of contrast media. Interventional cardiology is increasingly providing definitive treatment for a number of conditions, for instance closure of ASD or PDA by insertion of an occlusion device, balloon dilatation of pulmonary stenosis, or diathermy ablation of abnormal conduction pathways.

PRINCIPLES OF ANAESTHESIA IN CHILDREN WITH CONGENITAL CARDIAC DISEASE

Unnecessary surgery should be avoided due to the increased risk associated with anaesthesia, particularly in neonates, and children with complex (cyanotic) disease. It is important to optimise the condition of the child preoperatively, to understand the physiology of the lesion, and know how to avoid acute decompensation during surgery.

The general principles of anaesthesia are as follows:

- Avoid deep (halothane) anaesthesia
- Support ventilation where possible

- Avoid air emboli, particularly for cyanotic children with right-to-left shunts; make sure there are no air bubbles in the drugs or fluids given
- Consider endocarditis prophylaxis
- Consider the effect of anaesthesia on the pulmonary blood flow, and the balance between systemic and pulmonary blood flow.

Pulmonary blood flow

Pulmonary blood flow may be low due to increased pulmonary vascular resistance (PVR), which may be due to excessive muscularisation of the pulmonary arterioles secondary to long-standing high pulmonary blood flow. Pulmonary vascular resistance may be increased acutely, particularly in neonates, due to:

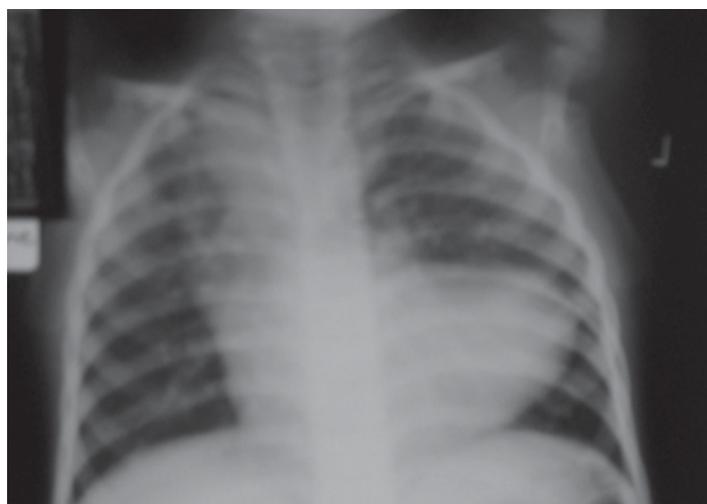


Figure 8. Chest Xray of a child with tetralogy of Fallot and right-sided aortic arch showing a 'boot shaped' heart

- Hypoxia
- Acidosis
- Over/under inflation of the lungs.

The most powerful pulmonary vasodilator available therapeutically is oxygen. The effect of oxygen is particularly evident in the first weeks of life when the pulmonary vasculature remains very reactive. It is sometimes important to avoid giving too much oxygen. For instance, if a patient with high pulmonary blood flow is given 100% oxygen to breathe (e.g. a baby with a large unrepaired AVSD), this will cause the pulmonary vascular resistance to fall, pulmonary blood flow will increase and the symptoms of cardiac failure, such as breathlessness, will get worse. A saturation of 94% is adequate in children with unrepaired AVSD, and excessive oxygen should be avoided.

Other pulmonary vasodilators include:

- Nitric oxide, an inhaled pulmonary vasodilator, used for the treatment of pulmonary hypertension in neonates
- Sildenafil, a cGMP-specific phosphodiesterase type 5 inhibitor
- Bosentan, an endothelin receptor antagonist.

If a patient has impaired pulmonary blood flow due to pulmonary hypertension, for instance, a child with sepsis and persistent pulmonary hypertension of the newborn, they should be treated with oxygen, nitric oxide +/- sildenafil if available.

ANAESTHESIA FOR SPECIFIC CARDIAC LESIONS

Children who have had corrective surgery e.g. for ASD, VSD or PDA, may be treated the same as any other child. Children with uncorrected cardiac lesions may be more challenging.

Uncorrected left-to-right shunt; ASD, VSD, AVSD, PDA

Problems

- Increased pulmonary blood flow
- Cardiac failure.

Anaesthesia for uncorrected left-to-right shunt

- Careful preoperative assessment; if possible, treat cardiac failure with diuretics (furosemide, spironolactone PO). In older children, exclude signs of Eisenmenger's syndrome and shunt reversal
- Avoid deep halothane anaesthesia
- Avoid excessive oxygen; keep FiO_2 at a minimum, accept SpO_2 approx. 94% (but see below for Eisenmenger's syndrome)
- Support the ventilation to reduce the work of breathing (increased pulmonary blood flow causes lung compliance to fall).

Eisenmenger's syndrome

Irreversible pulmonary hypertension has developed as a consequence of long-standing or significantly raised pulmonary blood flow. There is now right-to-left shunting or bidirectional shunting and the

patient is cyanosed. The mortality from Eisenmenger's syndrome is 75% at 30 years of age. (See figure 5).

Problems

- Shunt reversal
- Cyanosis

Anaesthesia for Eisenmenger's syndrome

The patient is severely unwell, and may present with the following signs and symptoms:

- Limited exercise tolerance
- Polycythaemia, hyperviscosity syndrome (fatigue, headache, blurred vision, cerebral thrombosis, cerebral abscess)
- Coagulopathy
- Thromboembolic events
- Endocarditis.

This is very high-risk surgery, with an estimated perioperative mortality of 10-14% even for minor surgery. The risks and benefits of proceeding with surgery have to be considered carefully. The following is recommended:

- Polycythaemia is associated with hyperviscosity. Consider venesection if the haematocrit is >60. Do not let the patient become dehydrated; start IV fluids preoperatively.
- Give high inspired oxygen at all times.
- Use a careful balanced general anaesthetic and close monitoring. Induce anaesthesia with small incremental doses of midazolam and fentanyl IV, and a small dose of induction agent (ketamine, etomidate).
- Support ventilation to reduce the work of breathing, but do not over/under inflate the lungs (increases PVR).
- Avoid increases in pulmonary vascular resistance, as this will make the cyanosis worse.
- Do NOT use a spinal, as this will cause a fall in systemic vascular resistance, and will encourage the right to left shunt.
- Maintain oxygen carrying capacity; do not allow the patient to become anaemic, and maintain haemoglobin 12-14g.dl⁻¹ (Hct 36-42).
- Consider a pulmonary vasodilator if possible – sildenafil or inhaled NO

Management of a pulmonary hypertensive crisis

In the event of a pulmonary hypertensive crisis with falling SpO_2 and blood pressure, you must intervene to reduce the PVR and increase cardiac output:

- Give 100% oxygen and hyperventilate the patient
- Give analgesia
- Treat metabolic acidosis

- Support the circulation – adrenaline infusion
- Start a pulmonary vasodilator if available (inhaled NO, sildenafil via a nasogastric tube).

The patient must be monitored in a high dependency area or ICU after surgery.

Tetralogy of Fallot

Problems

- Reduced pulmonary blood flow due to obstruction at the right ventricular outflow tract (NB PVR is LOW)
- Hyperviscosity syndrome and polycythaemia due to long-standing cyanosis.

Anaesthesia for children with unrepaired TOF

- Sedative premedication; keep the child calm
- Balanced anaesthesia with good analgesia; avoid dehydration, keep well-filled, avoid sympathetic stimulation to avoid hypercyanotic spells.

Management of a hypercyanotic 'spell' during surgery in unrepaired TOF

- Increase the FiO_2 .
- Deepen anaesthesia.
- Give a bolus of Ringer's $20ml.kg^{-1}$ to increase cardiac output and blood flow to the lungs.
- Increase the systemic vascular resistance to reduce the right-to-left shunt; phenylephrine bolus $1mcg.kg^{-1}$ IV, titrated

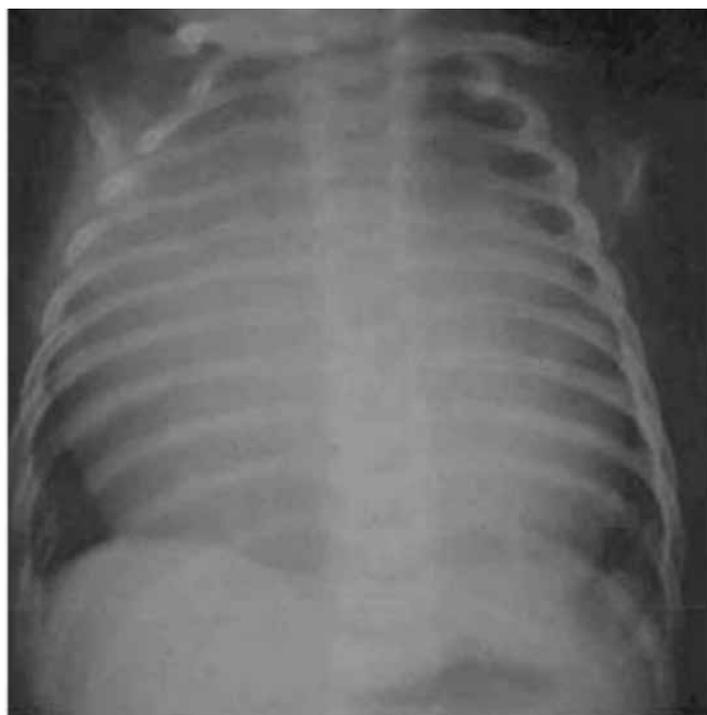


Figure 9. Large pericardial effusion in a child

to effect. Do NOT use adrenaline as this will worsen the right ventricular outflow tract obstruction (β_1 -receptor agonist effect).

- Consider a β -blocker – esmolol or propranolol IV.

Anaesthesia for children with a modified BT shunt (mBT shunt)

- Check the shunt is patent before the start of surgery – SpO_2 should be around 85%, and a shunt murmur should be audible (continuous murmur in the second intercostal space, mid-clavicular region on the right).
- Avoid dehydration as this may result in the shunt becoming blocked.
- Keep the blood pressure up; if the blood pressure falls, pulmonary blood flow will also fall and the oxygen saturation will drop.

High-risk lesions

There are a number of high-risk lesions that should ring alarm bells; these 'red flag' patients must be assessed and optimised before surgery, and the risk/benefits of non-cardiac surgery considered very carefully.

The following are high-risk lesions:

- Eisenmenger's syndrome. (See above)
- Aortic stenosis. Avoid a fall in SVR – do not use spinal anaesthesia
- Cardiomyopathy. May be dilated or restrictive, and the child will have limited reserve. Ketamine is useful for induction, and the child should be monitored carefully postoperatively
- Pericardial effusion. Must be drained by the cardiologist before surgery (See Figure 9)
- Endomyocardial fibrosis. Manage as for restrictive cardiomyopathy.

ENDOCARDITIS PROPHYLAXIS

Children who have had bacterial endocarditis, who have a prosthetic valve, and who have congenital heart disease are at high risk of bacterial endocarditis. The current advice from the American Heart Association is to give prophylactic antibiotics in the following situations:

- High risk dental surgery e.g. involving suturing or dental infection
- Give antibiotics as indicated for the surgical procedure.

ANAESTHESIA FOR A CHILD WITH AN UNDIAGNOSED CARDIAC LESION

There may be situations in LMIC where the anaesthetist is presented with a child with a murmur and an undiagnosed cardiac condition. The following approach is advised:

Elective surgery

Refer for investigation if possible.

Emergency surgery:

- 'Well' child, pink, breathless with cardiomegaly. Likely to be a condition associated with left to right shunt such as an ASD, VSD or PDA. The child may have heart failure with increased work of breathing. Support ventilation during surgery to reduce the work of breathing.
- Young child, 'well', but has always been blue. Likely to be tetralogy of Fallot. The problem is reduced pulmonary blood flow. Give high-inspired oxygen and avoid sympathetic stimulation.
- Any neonate, or older child with poor function, funny turns +/- cyanosis. These cases are high risk and only life-saving surgery should be contemplated. Use a balanced anaesthetic technique with positive pressure ventilation. Ketamine is a good agent to use in these high-risk cases.

All children with undiagnosed cardiac disease should be referred to a cardiologist as soon as possible for investigation.

CONCLUSION

Children with CHD may provide a challenge to the anaesthetist, but with careful planning, most can be anaesthetised safely. It is important to understand the physiology of the particular cardiac lesion, to prepare in advance, and to do the simple things well.

ACKNOWLEDGEMENT

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REFERENCES

1. Story E. Recognising cardiac disease in children. *Anaesthesia Tutorial of the Week* 93 (2008). Available at: http://www.wfsahq.org/components/com_virtual_library/media/4bd1c43b45969453c47a9a1c96723d46-64f3a3069773de361efae976bb23f931-93-Recognising-Cardiac-Disease-in-Children.pdf
2. Baum VC, Barton DM, Gutgesell HP. Influence of congenital heart disease on mortality after non-cardiac surgery in hospitalized children. *Pediatrics* 2000; **105**: 332-5.
3. Paediatric cardiology: an introduction. Archer N, Burch M. Chapman Hall Medical. Philadelphia 1998.

Anaesthesia in patients with asthma, bronchiolitis and other respiratory diseases

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ASTHMA

Introduction

Asthma is a leading cause of morbidity in children throughout the world.¹ In urban hospitals, it is the most common reason for hospital admission. The prevalence among children in Western countries is between 2 and 10%.¹ Asthma is a lung disease characterized by three distinct features: airway obstruction, airway inflammation and airway hyper-responsiveness. Symptoms manifest as episodes of recurrent wheezing, chest 'tightness', dyspnoea and dry cough. Typically episodes result in variable obstruction of airflow that resolves either spontaneously or with treatment. Asthma has no radiologic, histologic or confirmatory blood test. It is characterized by reversible symptoms, findings on examination and pulmonary function tests. While the characteristic symptom of asthma is wheezing (usually expiratory), it is important to realize that wheezing is produced simply by airflow passing through a sufficiently narrowed airway. Wheezing can occur during different phases of the respiratory cycle depending on the site of the airway obstruction and its cause. The differential diagnosis of wheezing in children is extensive and includes: asthma, foreign body, bronchiolitis, inhalational injury, pneumothorax, endobronchial intubation, herniated endotracheal tube cuff, cardiac failure, cystic fibrosis, sickle cell disease, recurrent aspiration, mediastinal mass, tracheomalacia, vascular ring, tracheal web/stenosis, bronchial stenosis and roundworm infestation.²

An acute exacerbation of asthma can quickly progress to severe respiratory distress and may ultimately lead to respiratory failure defined by hypoxaemia/hypercarbia. It is important to note that as an acute asthma attack worsens and the child fatigues, wheezing may become diminished or be completely absent. Many children with chronic asthma also have chronic inflammatory changes that may be associated with permanent alterations in their airway structure. These patients may not be responsive to commonly used treatments.

Epidemiology

The median age of onset for asthma is 4 years old with

more than 20% of children developing symptoms within the first year of life.¹ However, the diagnosis of asthma in an infant is unusual and other causes for wheezing should be investigated. Asthma most likely results from an interaction between both an inherited modifier of inflammation and environmental influences. A number of risk factors have been identified but the best researched include gender, atopy, allergens, infections, obesity, tobacco smoke, and perinatal factors.

Childhood asthma tends to occur predominantly in males until puberty. After age 20 the prevalence is equal between the sexes. The association between asthma and other atopic diseases has been well researched. An "atopic march" has been identified which starts as atopic dermatitis in the infant, followed by allergic rhinitis and asthma in the older child or adolescent. Indoor allergens have been shown to play a significant role in the development of asthma. The dust mite, *Alternaria* mould, cockroach allergens, as well as cat and dog allergens have all been implicated.

Smoke exposure from cooking on open wood fires is an important risk factor in many developing countries. Viral and bacterial infections are well known triggers of asthma exacerbations but their causal relationship remains unproven. Several large studies have suggested that patients with an elevated body mass index (BMI) or that are actively smoking are at increased risk of developing asthma. Lastly, perinatal risk factors have been studied extensively but have so far found few strong correlations. This is most likely due to the inherent difficulty in controlling for confounders between study groups.¹

Pathophysiology

The chronic airway obstruction seen in asthmatic patients is caused by inflammation and hypertrophied bronchial smooth muscle leading to hyperinflation and air trapping. This results in decreased lung compliance and increased work of breathing. Chronic airway obstruction leads to a ventilation/perfusion mismatch and dead space ventilation which are clinically evident as hypoxaemia and further increased work of breathing.

Summary

Respiratory illnesses are common in the paediatric population. As a result, children often present for surgery and anaesthesia with respiratory symptoms. The child with well controlled asthma or minimal symptoms of respiratory infection may proceed with anaesthesia. The dilemma lies in the decision to cancel or proceed when symptoms are more than minimal. This article describes respiratory conditions in children as they relate to anaesthetic perioperative management.

Our goal is to inform anaesthesia providers of safe practices, and the risks and benefits of administering anaesthetics to these patients.

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Preoperative assessment

Assessment of acute asthma

Important questions when evaluating a child with asthma include the following:

- frequency of symptoms,
- amount and purulence of sputum,
- use and effectiveness of medications,
- asthma triggers,
- activity level, previous history of surgery and anaesthesia, recent upper respiratory infections,
- hospitalizations and emergency department attendance.²

Investigate hospitalizations to determine if ICU admission or intubation were required. Use the physical examination to assess for wheezing, a prolonged expiratory phase, use of accessory muscles of breathing (e.g. intercostal and subcostal retractions), respiratory rate, cyanosis, and drowsiness. Oxygen saturation (SpO₂) is useful to have preoperatively.

Preparation for surgery

Instruct parents to administer all asthma medications, including on the morning of surgery to ensure optimal treatment. Children with well controlled asthma who are scheduled for elective surgery should not have wheezing on the morning of surgery. Preoperative wheezing is predictive of perioperative complications. Stabilize patients in conjunction with the paediatric medical team and anaesthetise only when determined to be stable by a senior anaesthetist.

Treatment

Understanding the patient's asthma severity is critical to administering the appropriate preoperative treatment. Table 1 defines each severity classification based on frequency of daytime symptoms, frequency of night time symptoms, ratio of Peak Expiratory Flow Rate (PEFR) to Forced Expiratory Volume over 1 second (FEV₁) and the PEFR variability. In medical settings where these pulmonary function tests are unavailable, spirometry is a useful alternative for preoperative assessment. Spirometry offers an easy method of determining the presence and severity of airway obstruction. The patient is asked to place on a nose clip and then take a deep breath, exhaling into the spirometer as hard and for as long as possible. This is immediately followed by a rapid inhalation and results in a diagnostic flow-volume loop.

Preoperative treatment for mild intermittent or mild persistent asthma involves administering a nebulized β_2 -adrenergic agonist, such as salbutamol, 1 to 2 hours prior to surgery.

Preoperative treatment for moderate persistent asthma involves additional optimization with any inhaled anti-inflammatory agent (i.e. cromolyn, theophylline, montelukast - all act by suppressing the inflammatory mediators that cause airway hyperreactivity) and by consistent use of nebulized β_2 agonists in the week prior to surgery. Preoperative treatment for severe persistent asthma involves a visit to their primary care physician or pulmonologist prior to surgery in order to optimize treatment. Some children benefit from short-term oral corticosteroid therapy, such as prednisone (2mg.kg⁻¹ PO once daily for 3-5 days, max of 60 mg.dose⁻¹) or oral dexamethasone (0.6mg.kg⁻¹ PO once daily for 2 days, max of 16mg.dose⁻¹) prior to surgery.

These preoperative treatments have all been shown to be both effective and safe with a low incidence of side effects.¹ However, determining the severity of the patient's asthma is not sufficient. It is also important to determine the patient's control of their disease. A patient with severe asthma may nevertheless be well controlled, whereas a patient with mild asthma may be very poorly controlled. It is also important to note the difference between poorly controlled asthma and severe persistent asthma. Poorly controlled asthma (i.e. non-compliant with preventative or therapeutic medications) can occur within all severity classifications of chronic asthma. However, to be categorized as having severe persistent asthma the patient must meet the specific qualifications of symptom frequency and measured PEFR and FEV₁ values. The potential for perioperative complications (e.g., bronchoconstriction, bronchospasm, laryngospasm and reduced FEV₁) is present with either of these scenarios. Most patients with asthma should be seen by the physician who manages their disease one week prior to surgery for appropriate and timely preoperative management.

For the child with asthma, base your decision whether to proceed with surgery on the stability of asthma symptoms and whether symptoms are optimally managed. The possible intraoperative complications include bronchospasm and laryngospasm. Warner and colleagues³ identified three factors that correlate with perioperative bronchospasm:

1. Use of bronchodilators
2. Recent asthma exacerbation

Table 1. Severity classification of chronic asthma

	Days with symptoms	Nights with symptoms	PEFR/FEV ₁ % predicted	PEFR variability
Mild intermittent	≤ 2 / week	≤ 2 / month	≥ 80%	≤ 20%
Mild persistent	3-6 / week	3-4 / month	≥ 80%	20-30%
Moderate persistent	Daily	≥ 5 / month	60-80 %	> 30%
Severe persistent	Continuous	Frequent	≤ 60%	>30%

3. Recent visit to a medical facility to treat asthma.

This makes an accurate asthma history crucial. The intraoperative goal is to depress airway reflexes with appropriate anaesthetic drugs. This will prevent bronchoconstriction of the patient's hyperreactive airway that can occur with airway manipulation. Stimuli that would not usually cause an airway response in a patient without asthma may initiate a serious episode of bronchoconstriction in a patient with asthma.

Intraoperative management

Anaesthetic technique

Consider regional anaesthesia as the sole technique since this may prevent the need for airway instrumentation. This is particularly important in patients undergoing emergency surgery when there is insufficient time to optimize their pulmonary status. When choosing to do a general anaesthetic, a thorough understanding of the physiological effects of each anaesthetic drug and its interaction with an asthmatic patient is crucial. These are described below.

Intravenous drugs

Midazolam does not alter bronchial tone and is therefore a safe choice for preoperative anxiolysis.

Propofol inhibits bronchoconstriction and increases airway dilation by directly relaxing the airway smooth muscle. These actions decrease the risk of bronchospasm during induction and propofol is considered safe for patients with asthma. This agent may not be suitable for haemodynamically unstable patients.

Ketamine has sympathomimetic bronchodilatory properties, with a direct relaxing action on bronchial smooth muscle. These actions decrease the possibility of bronchospasm with induction so that ketamine has been proposed as a good choice for patients with severe persistent asthma. Ketamine also increases bronchial secretions and should be given simultaneously with an anticholinergic drug such as glycopyrrolate or atropine. If possible, ketamine sedation may also be considered as an alternative to general anaesthesia.

Thiopental use has been associated with bronchospasm, making it a poor first choice for induction of anaesthesia.

Lidocaine, when given intravenously, significantly increases the histamine threshold and blocks the cough reflex. It may be given to decrease the airway responses associated with endotracheal intubation.

Morphine causes histamine release which may result in bronchospasm. Fentanyl, if rapidly administered in large doses, can result in chest rigidity which could be mistaken for bronchospasm. However, fentanyl administered more judiciously is preferable to morphine.

Inhalational anaesthetics

Halothane, enflurane and isoflurane are potent bronchodilators that act via β -adrenergic receptor stimulation. They have been shown to decrease airway responsiveness and help ease histamine-induced bronchospasm. Many studies have shown their effectiveness in the treatment of status asthmaticus. Sevoflurane has controversial effects in asthmatics: some studies show an increase in airway resistance

and others show no change. Desflurane is a very pungent agent that is irritant to the airway and has been shown to increase secretions, coughing, and laryngospasm. Halothane and sevoflurane remain good choices for inhalational induction.

Muscle relaxants

Gallamine, pipecuronium and rapacuronium are all neuromuscular blockers that bind and stimulate M2 muscarinic receptors more than M3 muscarinic receptors, causing bronchoconstriction. The neuromuscular blockers that stimulate the M2 and M3 muscarinic receptors evenly, such as vecuronium, rocuronium, cisatracurium and pancuronium, do not cause bronchoconstriction. Atracurium, mivacurium and suxamethonium dose-dependently release histamine and thus trigger bronchoconstriction. The rapid onset and short duration of action of suxamethonium continue to make it a useful agent for a rapid sequence induction.

Intubation technique

Airway manipulation, particularly in a light plane of anaesthesia, is a potent stimulus of bronchospasm, laryngospasm, coughing and breath-holding. Use of an inhaled β_2 -agonist before intubation may reduce bronchospasm. In general, avoidance of airway stimulation seems sensible. For short cases a face mask may be sufficient. Whenever possible, avoidance of tracheal intubation is preferred and a laryngeal mask airway (LMA) should be used since it may be less stimulating than an endotracheal tube.² However, in cases that require airway protection (for instance patients with severe gastro-oesophageal reflux disease), and endotracheal intubation is necessary, ensuring a deep level of anaesthesia prior to airway instrumentation reduces the risk of bronchoconstriction.² An endotracheal tube that is too long and touches the carina is a potent cause of bronchospasm; the length of the endotracheal tube should therefore be assessed carefully after intubation. The Morgan formula, (endotracheal tube length measured at the front upper teeth [cm] = 0.10 x height [cm] + 5), is a useful gauge of appropriate endotracheal tube depth in children. Another alternative which is simple to calculate for oral endotracheal tube depth (in cm) is endotracheal tube (internal diameter) size x 3. Neither formula uses age in its calculations, since this can be misleading in poorly-nourished populations.

Maintenance

Maintenance of anaesthesia with inhalational agents such as halothane helps to dilate bronchioles and prevent bronchospasm. It is important to have a method to deliver a bronchodilator (e.g. salbutamol) intraoperatively after the airway is secured. A simple technique is to remove the plunger from a 60ml syringe and place a bronchodilator metered dose inhaler (MDI) into the barrel. The plunger is then replaced and when depressed will trigger the MDI to spray through the Luer Lock connector of the syringe. This can easily be incorporated into the anaesthesia circuit at the elbow connecting the endotracheal tube. Synchronize the MDI with inspiration to increase its administration. Commercially available adaptors also exist which optimize timing of drug administration with inspiration and further improve efficiency.⁴

You must be vigilant for the signs of intraoperative bronchospasm in order to be able to treat it. Intraoperative bronchospasm presents as a prolonged expiratory phase/wheeze with increased airway pressure,

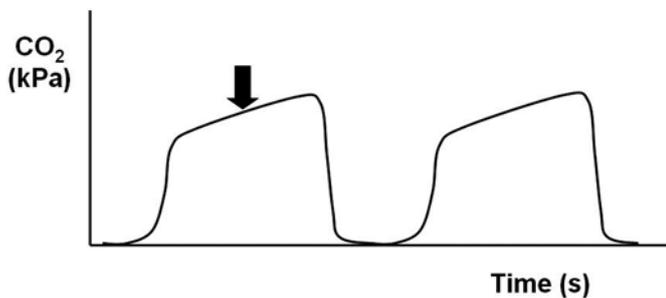


Figure 1. Typical up-sloping capnograph trace seen with bronchospasm

up-sloping in the end-tidal CO₂ waveform and hypoxia, and may result in difficulty with ventilation. You must rule out other causes of airway obstruction (e.g. equipment malfunction) and wheeze (e.g. pulmonary aspiration) before starting treatment:

Treatment of bronchospasm under general anaesthesia

1. Remove causative stimulus, while increasing the FiO₂
2. Deepen the volatile anaesthetic
3. Inhaled salbutamol
4. SC or IV adrenaline (10mcg.kg⁻¹ SC or 1mcg.kg⁻¹ IV, caution with IV adrenaline)
5. IV steroids
6. Paralysis (if not already)
7. IV ketamine (consider).

The end-tidal CO₂ waveform is an early indicator of the effectiveness of these interventions. Do not be overly concerned with treating a high CO₂ which may lead to excessive ventilation, elevated peak inspiratory pressures and possible pneumothorax.

Extubation

For patients undergoing elective surgery and where no other contraindications exist, deep extubation is the best option. The depth of anaesthesia should be sufficient to prevent laryngo-/

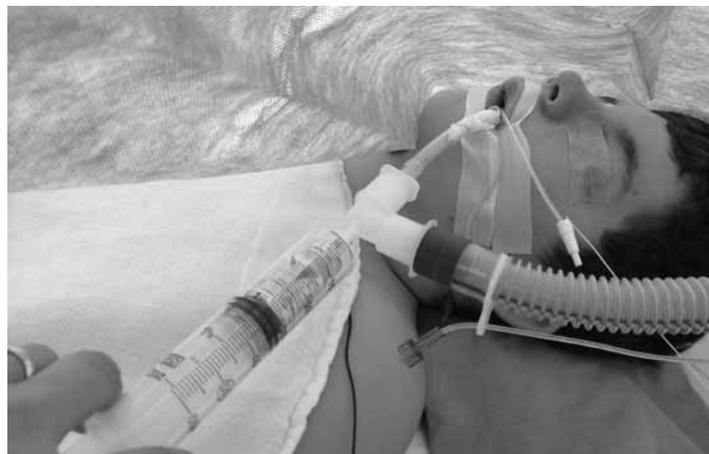


Figure 1: MDI incorporated into anaesthesia circuit using a 60ml syringe

bronchospasm when the endotracheal tube is removed. For patients undergoing emergency surgery, with a potentially full stomach, awake extubation is a more sensible option. An awake extubation occurs after upper airway reflexes have returned and the patient can therefore protect their airway. Sufficient amounts of opioid and re-dosing of inhaled β₂-agonists may also be useful prior to extubation to decrease the reflex bronchoconstriction that may occur. It is important to remember when reversing muscle relaxants that both neostigmine and physostigmine cause increased secretions and bronchial hyperreactivity.

Postoperative management

Analgesia

When possible and where indicated, postoperative regional analgesia is preferable. A functioning epidural will block afferent pathways mediating pain from the abdominal viscera/incision and therefore help to maintain the patient's respiratory muscle function. This leads to more adequate tidal volume and vital capacity and helps to preserve diaphragm function. A regional-only anaesthetic technique also eliminates the need for airway manipulation, thus preventing potential airway irritation. Avoidance of both meperidine (pethidine) and morphine is preferable, given their ability to release histamines which can result in bronchospasm. As mentioned above, fentanyl is the best opioid alternative. An infusion of ketamine has also been used in the postoperative period since it is capable of providing analgesia with the direct advantage of preventing bronchospasm.⁴ Use nonsteroidal anti-inflammatory drugs (e.g. ibuprofen and ketorolac) with caution in patients with asthma given that a small proportion of children (1 in 50) may have a bronchospastic response.

Monitoring

Standard postoperative monitors are indicated while waiting for the patient to regain normal airway/breathing function. The head up position is preferred to help clear secretions and prevent atelectasis. The early control of sputum and respiratory symptoms helps to prevent postoperative complications.

BRONCHIOLITIS

Introduction

Bronchiolitis is a lower respiratory tract infection with a spectrum of clinical presentations ranging from minimal symptoms to fulminant respiratory failure requiring mechanical ventilation. It is usually a clinical diagnosis; routine laboratory or radiologic studies are not recommended. A child usually presents with symptoms of a respiratory infection (e.g. cough, nasal congestion). This may be followed by fever, poor appetite and lethargy. Clinical symptoms peak around day 3 to 4 of illness and bronchiolitis is usually a self-limiting disease.⁶ Those at higher risk of clinical deterioration include infants with chronic lung disease, congenital heart disease, and/or prior prematurity.

Epidemiology

Viral bronchiolitis is the most common cause of respiratory disease in children under two years of age requiring hospitalization. The most common cause (80%) is respiratory syncytial virus (RSV). Less common causes include rhinovirus, parainfluenza, influenza, human metapneumovirus and adenovirus. Annual epidemics of RSV

bronchiolitis occur during the winter and spring months. Risk factors include prematurity (gestation < 37 weeks), low birth weight, age < 6–12 weeks, chronic lung disease, haemodynamically significant congenital heart disease, immune deficiency, neurologic disease and anatomical airway abnormalities.⁷ Outcome is generally favourable, although a significant number will develop reactive airway disease.

Pathophysiology

The virus penetrates the terminal bronchiole's lining of epithelial cells, causing necrosis, ciliary disruption and peri-bronchiolar lymphocyte infiltration. Pathological changes are detectable 18 to 24 hours after infection. Subsequently oedema, excess mucus and sloughed epithelial cells lead to obstruction of small airways and atelectasis. Ventilation-perfusion mismatch leads to hypoxaemia. Infants are more susceptible than adults due to proportionally smaller diameter airways.

Preoperative management

Assessment

When you assess a child with bronchiolitis prior to surgery, it is important to assess the severity of illness to determine the course of action. Postponing surgery may well be safest. Usually the child with mild bronchiolitis presents with nasal congestion. With increasing severity, hallmark findings include rhonchi and crepitations, with occasional expiratory wheezes.⁸ More severe disease is associated with respiratory distress (increased work of breathing and tachypnea), cyanosis, hypoxaemia ($\text{SpO}_2 < 95\%$ on room air), dehydration and restlessness or lethargy. Severe cases often involve infants younger than 3 months old, who appear ill or toxic, and likely will have atelectasis on chest X-ray.⁹ The most significant clinical parameters in determining severity of illness are respiratory rate, work of breathing, and hypoxia. Possible indications for medical management in the intensive care unit include recurrent apnoea, slow irregular breathing, reduced level of consciousness, shock, exhaustion, hypoxia despite high levels of inspired oxygen, and respiratory acidosis ($\text{pH} < 7.20$).⁴ Continuous positive airway pressure can be effective; however, the majority of deteriorating infants require mechanical ventilation.

In the setting where the child with bronchiolitis presents for surgery, you must evaluate the urgency of the surgical procedure. If it is an elective case, postpone it and reschedule after 4 to 6 weeks. If the procedure is an emergency or urgent, the risks associated with anaesthesia may be outweighed by the benefits of surgery. These risks include:

- Airway hyperreactivity - this may result in bronchospasm, laryngospasm and wheezing
- Supplemental oxygen
- Excessive secretions - may cause airway obstruction, necessitating frequent suctioning
- Apnoea and respiratory failure - RSV causes significant apnoea in infants by an unknown mechanism. In one retrospective review, 21% of infants who were hospitalized with RSV presented with apnoea. This may be worsened by the use of inhalation anaesthetics, sedatives and opioids.

As a result, children with bronchiolitis undergoing general

anaesthesia have a higher risk of requiring mechanical ventilation in the postoperative period.

Preoperative treatment

Preoperative treatment goals for the child with active symptoms of bronchiolitis, who requires urgent surgery include optimizing the respiratory status and rehydration. The key therapeutic intervention is oxygen administration to maintain $\text{SpO}_2 > 92\%$. Oxygen can be administered via nasal cannula, head box or facemask. Nasopharyngeal suctioning is often effective and can relieve upper airway obstruction, increase comfort and decrease work of breathing. Intravenous fluids are often administered starting with a crystalloid bolus of 10ml.kg^{-1} until deficits are replaced. Fluid requirements are increased due to fever, tachypnoea and decreased oral intake. Consider nasogastric feeding for nutritional support until feeding improves prior to surgery, provided that it meets fasting time requirements prior to surgery.

Therapeutic interventions that have been used include bronchodilators, corticosteroids, antiviral agents, antibacterial agents, chest physiotherapy and decongestant drops. None of these have demonstrated significant impact on illness duration, severity or clinical outcomes.

The use of bronchodilators in the routine treatment of bronchiolitis is not recommended by the American Academy of Pediatrics. Their efficacy is uncertain and published results have been variable.¹⁰ Given the excellent safety profile of this medication, you may consider a trial of bronchodilators, but continue only if a response is documented. Adrenaline and salbutamol are used most commonly (refer to Table 2), although adrenaline should be reserved for hospitalized patients only.

Table 2. Recommended doses of medications

Salbutamol:

0.15 mg.kg^{-1} (minimum 2.5mg, maximum 5mg) diluted in 2.5 to 3ml saline and administered over 5 to 15 minutes via nebuliser (requires oxygen/driving gas); or 4 to 6 puffs via MDI with spacer and facemask dependent upon size/age of child.

Adrenaline:

0.05ml.kg^{-1} of 2.25% adrenaline diluted in 3ml normal saline and administered via nebuliser.

Corticosteroids should not be used as they have not been shown to reduce the length of stay or disease severity. In addition, they have a well-established undesirable side effect profile. Chest physiotherapy should also be avoided as it will not improve the diffuse regions of inflammation involved in bronchiolitis. Likewise, ribavirin and antibacterial agents are not recommended for routine use.

Intraoperative management

Choice of anaesthetic technique

Preoxygenation is essential prior to induction of anaesthesia. Where IV access is available it might be the preferable route for induction. It may be better to use agents that are associated with less bronchoconstriction and suppress airway reflexes (e.g. propofol), as

for the asthmatic child. Wherever possible, avoidance of opioids will decrease the risk of postoperative apnoeic episodes. When IV access is unavailable, proceed with cautious inhalation induction (using halothane or sevoflurane) recognizing that there is an increased risk of bronchospasm and laryngospasm. Avoid nitrous oxide as it may worsen hypoxaemia.

Intubation technique

It is essential to ensure a deep plane of anaesthesia before attempting intubation. IV lidocaine can reduce airway reflex responses. Endotracheal tube placement provides advantages of a definitive airway and allows for frequent suctioning. Once the airway is secured, the goals of ventilation are to minimize air-trapping and lung distention, and prevent barotrauma. If available, employ a pressure-controlled mode of mechanical ventilation (or manual ventilation) to minimize the risk of dangerously high inspiratory pressure. A preset long respiratory phase will allow full expiration.

Intraoperative management

Maintenance of anaesthesia using volatile agents which bronchodilate may be preferable (e.g. halothane). Drying of secretions with atropine or glycopyrrolate may be useful intra-operatively but can exacerbate mucus plugging postoperatively. Avoid histamine-releasing agents which may contribute to bronchospasm. Consider regional and neuraxial anaesthesia for analgesia. These techniques should reduce opioid use and decrease the risk of postoperative apnoea and hypoventilation.

Intraoperatively, monitor airway pressure closely for subtle changes in lung compliance. A sudden elevation of airway pressure may be due to mechanical problems such as kinking or mucus plugging of the endotracheal tube, pneumothorax, or endobronchial intubation. If obstruction of the endotracheal tube from mucus plugging is thought to be the cause, suctioning using soft catheters may reduce obstruction, or replacement may be required. The use of humidifiers may reduce the risks of inspissated mucus plugs. If the cause is thought to be bronchospasm, inhaled salbutamol (4-8 puffs every 20 minutes x 3 doses) can be introduced into the anaesthesia circuit. Further interventions may include subcutaneous adrenaline (10mcg.kg⁻¹ SC every 20 minutes x 3 doses, up to 500mcg.dose⁻¹) or intravenous adrenaline (1mcg.kg⁻¹ IV every 20 minutes). Given its cardiovascular side effects, use intravenous adrenaline cautiously and reserve it for cases when other treatments fail to alleviate symptoms. Capnography is a useful monitor as the changing shape of the capnograph trace (slower upstroke) is an early indicator of bronchospasm and the effectiveness of treatment.

Extubation

For some elective surgery patients, the trachea may be extubated under deep inhalational anaesthesia to avoid triggering bronchospasm. Close monitoring into the postoperative recovery phase is mandatory. For emergency surgery with a potentially full stomach, extubate the trachea once the child is awake and the upper airway reflexes have returned. The use of anticholinesterases for neuromuscular blockade reversal is recommended if muscle relaxants were used to optimize respiratory function postoperatively. In some circumstances, you may prefer to leave the child electively intubated at the conclusion of surgery. This is particularly true in patients with a history of

apnoea, hypoxaemia, hypercapnia (PaCO₂ > 55mmHg/7kPa) prior to surgery, haemodynamically significant heart disease and chronic lung disease.

Postoperative management

Analgesia

Avoid opioids or titrate them carefully. Multi-modal analgesia, using acetaminophen (paracetamol), non-steroidal anti-inflammatory drugs and regional anaesthesia techniques are useful to supplement postoperative analgesia.

Monitoring

Postoperative monitoring with continuous pulse oximetry and apnoea monitoring for 24 hours minimum is recommended. If frequent monitoring with pulse oximetry is impractical, the child should be located such that there is direct visual monitoring of the patient (e.g., close to the nursing station).

OTHER RESPIRATORY DISEASES

Overview

The spectrum of respiratory tract infections ranges from the uncomplicated upper respiratory infection (URI) to pneumonia and pneumonitis. The most frequently encountered by the anaesthesia provider is the child with an uncomplicated URI, who presents with nasal secretions and cough. Provided that there is no fever, no lethargy or decreased appetite, and no findings of respiratory disease on physical examination (e.g. lack of wheeze or crepitus on auscultation), consider the child as having an uncomplicated URI. If the child presents with the above symptoms, a complicated URI (e.g. pneumonia) must be considered. The more common etiologies include viral (influenza, parainfluenza, adenovirus, human metapneumovirus, rhinovirus) and bacterial (*Streptococcus pneumoniae*, *Mycoplasma pneumoniae*, *Chlamydia pneumoniae*) causes. In immunocompromised children, additional etiologies include tuberculosis, fungal and less common bacterial and viral causes.

Management of respiratory diseases will depend on the severity of infection and the etiology. In uncomplicated viral URIs, no additional measures are usually required. However, in the case of more severe disease, antibiotics targeting the organisms involved, oxygen, nasal pharyngeal suction, rehydration, and respiratory support may be necessary.

Upper respiratory infections

A longstanding controversial issue is whether to proceed with anaesthesia in a child who presents with an uncomplicated URI. As a general rule, elective surgery should be postponed 4 to 6 weeks from the end of symptoms to allow acute airway reactivity to resolve. The risks of anaesthesia in the setting of increased airway reactivity include severe coughing, breath holding, bronchospasm, laryngospasm, apnoea with more rapid oxygen desaturation and post operative oxygen desaturation.¹¹ In a study by Cohen and Cameron, patients with URI symptoms preoperatively were 2 to 7 times more likely to experience respiratory related intraoperative complications and 11 times more likely if intubated.¹²

However, a child may experience 6 to 8 URIs per year and to postpone surgery for 4 weeks after symptom resolution may make scheduling of a procedure logistically impractical. Furthermore, in otherwise healthy children, the problems encountered intraoperatively can generally be easily handled without serious complications.¹³ Ungern-Sternberg and colleagues reported that increased perioperative risk of adverse respiratory events only occurred when URI symptoms were present on the day of surgery or had occurred within the preceding two weeks.¹⁴ Thus, the decision whether or not to proceed with an elective procedure should be made on a case by case basis, keeping in mind the anaesthesiologist's experience, level of comfort, and potential for management of possible complications.

SUMMARY

Asthma

- 3 distinct features: airway obstruction, airway inflammation, airway hyperresponsiveness
- Clinical diagnosis
- Optimize medical treatment before surgery, per the severity classification of chronic asthma
- Induction agents of choice: propofol, ketamine, halothane, sevoflurane
- Avoid intubation if possible, and consider deep extubation.

Bronchiolitis

- Wide spectrum of severity
- Most common cause is RSV
- Usually self limiting course
- Treatment is generally supportive with oxygen, nasopharyngeal suctioning and rehydration
- Recommend postoperative monitoring with pulse oximeter and apnoea monitor.

Upper respiratory tract infections

- Common presentation in children
- General management depends on severity of infection and etiology
- Postpone elective surgery 4 to 6 weeks to allow acute airway hyperreactivity to resolve
- In otherwise healthy children with uncomplicated URI, intraoperative complications can generally be managed without serious complications
- Proceeding with anaesthesia in a child with an uncomplicated URI is a case by case decision.

REFERENCES and FURTHER READING

- Liston DE. Childhood asthma and anaesthesia. *Anaesthesia Tutorial of the Week* **187** (2010).
1. Eggleston P. Chapter 420: Asthma. In: McMillan J, Feigin, R, DeAngelis, C, Jones M, eds, *Oski's Pediatrics - Principles and Practice* (4th edition) Philadelphia: Lippincott Williams & Wilkins, 2006: 2405-10.
 2. Firth P, Haver K. Chapter 11: Essentials of Pulmonology. In: Cote C, Lerman J, Todres I, eds. *A Practice of Anesthesia for Infants and Children* (4th edition) Philadelphia: Saunders Elsevier, 2009: 229-33.
 3. Warner DO, Warner MA, Barnes RD, Offord KP, Schroeder DR, Gray DT, Yunginger JW. Perioperative respiratory complications in patients with asthma. *Anesthesiology* 1996; **85**:460-7.
 4. Everett L, Fuzaylov G, Todres I. Chapter 37: Pediatric Emergencies. In: Cote C, Lerman J, Todres I, eds. *A Practice of Anesthesia for Infants and Children* (4th edition) Philadelphia: Saunders Elsevier, 2009: 776-78.
 5. Jahangir S, Islam F, Aziz L. Ketamine Infusion for Postoperative Analgesia in Asthmatics: A Comparison with Intermittent Meperidine. *Anesth Analg*, 1993; **76**:45-49.
 6. Hanson I, Shearer W. Chapter 229: Bronchiolitis. In: McMillan J, Eigin R, Deangelis C, Jones M, eds. *Oski's Pediatrics - Principles and Practice* (4th edition) Philadelphia: Lippincott, Williams & Wilkins, 2006: 1391.
 7. Piedra P, Stark A. Bronchiolitis in Infants and Children: Clinical Features and Diagnosis. *Up To Date*, September 3, 2009.
 8. Mcnamara D, Nixon G, Anderson B. Methylxanthines for the treatment of apnea associated with bronchiolitis and anesthesia. *Pediatr Anesth* 2004; **14**: 541-50.
 9. American Academy of Pediatrics Clinical Practice Guideline. Diagnosis and Management of Bronchiolitis. *Pediatrics* 2006; **118**:1774-93.
 10. Piedra P, Stark A. Bronchiolitis in Infants and Children: Treatment; outcome; and prevention. *Up To Date*, June 5, 2009.
 11. Tait AR, Malviya S, Voepel-Lewis T, et al. Risk factors for perioperative adverse respiratory events in children with upper respiratory tract infections. *Anesthesiology* 2001; **95**:299-306.
 12. Cohen MM, Cameron CB. Should you cancel the operation when the child has an upper respiratory tract infection? *Anesth Analg* 1991; **72**:282-8.
 13. Tait AR, Malviya S. Anesthesia for the child with an upper respiratory tract infection: still a dilemma? *Anesth Analg* 2005; **100**:59-65.
 14. Ungern-Sternberg BS, Boda K, Chambers N, Rebmann C, Johnson C, Sly PD, Habre W. Risk assessment for respiratory complications in paediatric anaesthesia: a prospective cohort study. *Lancet* 2010; **376**:773-83.

The preparation of children for surgery

Based in part upon: Langford R. The preparation of children for surgery. *Anaesthesia Tutorial of The Week* 132 (2009)

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INTRODUCTION

Good preoperative assessment and preparation of children for surgery is essential. This article reviews the preparation of children for elective and emergency surgery, including psychological preparation.

WHY DO CHILDREN REQUIRE A DIFFERENT APPROACH?

The physiological, psychological and social differences between children and adults necessitate a tailored approach to preoperative preparation.

Physiological

Most children presenting for elective surgery are systemically well and require little in the way of physiological assessment and investigation. Some children can present with complex congenital disease or unusual syndromes that may require specific preoperative investigation and preparation. Previously fit children presenting for emergency surgery may become very unwell, very quickly – this group of children must be recognised so that their condition is optimised prior to surgery.

Psychological

In comparison to adults, children are more likely to demonstrate behavioural issues at induction of

anaesthesia. Psychological development is related to the age of the child (Table 1).

Behavioural issues can result in a stormy anaesthetic induction and post-operative psychological difficulties such as nightmares, phobias, fears and negativism. Post-operative psychological problems are more likely to occur in:

- Children aged between two and three years old
- Children displaying a withdrawn affect pre-operatively
- A difficult, stressful induction
- A child with a history of multiple procedures.

It is important to be aware of these factors and to manage them effectively. This will improve the peri-operative experience for the child, and help shape long term attitudes to healthcare.

Safeguarding children

All staff working with children should be trained in basic child protection. Children are inherently vulnerable by virtue of their dependant status. Evidence of abuse or neglect may be encountered by the anaesthetist in a variety of ways, including direct disclosure. It is important to remember that the child's safety should have primacy over all other considerations. Act upon

Summary

Preparation of children for surgery is essential.

All children should be weighed and examined carefully prior to surgery.

For elective procedures, a pre-assessment visit may be helpful.

Children presenting for emergency surgery may be critically ill and require thorough assessment and resuscitation prior to induction of anaesthesia.

Table 1. Psychological developmental milestones

Age	Psychological developmental stage
<9 months	Babies are able to accept surrogates to parent and respond well to physical contact, talking and rocking.
1-3 years	Children are 'bonded' to parents, and more likely to display separation anxiety. Stormy inductions are most frequent and post-operative behavioural problems are more likely.
3-6 years	Children are more likely to display literal thinking: use euphemisms and metaphors with care. A clear explanation of events and description of procedures will reduce post-operative anxiety.
7-12 years	Children can think logically about real objects, but have trouble understanding hypothetical concepts. They are more independent and should be given simple honest explanations and the opportunity to participate (e.g. hold their own mask for induction)
Adolescents	Reasoning and mature 'adult-like' defence mechanisms have developed. Clear explanation and the opportunity to make decisions are essential to minimise anxiety.

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any suspicions by discussing concerns with colleagues and if possible, involving professionals specifically trained in child protection.

ELECTIVE SURGERY

Children are frequently admitted on the day of surgery. Day case surgery has been shown to reduce post-operative behavioural problems and should be encouraged where possible. Over 60% of paediatric surgery in the USA is now completed as a day case procedure.

Day surgery minimises disruption to families but leaves little time for preparation or for the child to adjust to their surroundings. Pre-admission programmes are now used in the majority of UK children's hospitals to help prepare for surgery.

Common approaches to pre-assessment include:

- Telephone screening - this is particularly useful if the child lives far away from the hospital.
- Nurse-led pre-assessment clinics - for children who live close to the hospital.

Many institutions have produced a standard proforma for pre-assessment as part of the pre-operative pathway, which allows a thorough history to be taken. Hospital patient records, including clinic letters should be retrieved for review. The child can be screened for suitability for day surgery, the requirement for further investigation, and the precautions required for anaesthesia in order to reduce day-of-surgery cancellations, also whether an anaesthetist needs to see the child for further assessment prior to surgery.

Table 2 (following page) lists a selection of common issues encountered in pre-operative screening and their implications. It is helpful to use this table during pre-assessment to identify high-risk patients.

The child's social situation is important when assessing suitability for day surgery. The family should have access to suitable transport from the hospital with one carer free, (i.e. not driving), should ideally live less than one hour from the hospital, and must have access to a telephone. An adult should be present at home for 24 hours after surgery. It is advisable for the family to purchase suitable analgesics in advance.

Pre-assessment visit

Explanation of procedures

Allow time to explain the peri-operative sequence of events to the parent and child. Preparation of the parent is crucial in reducing parental and therefore the child's anxiety. Parents should be encouraged to ask questions about any concerns they may have. If the parents are anxious, the child is more likely to display signs of anxiety themselves.

Explain using:

- Videos
- Play
- Booklets and written instructions
- Face-to-face discussions.

If using interpreters, check for understanding and supplement by written explanation and instructions in the patient's own language whenever possible.

Preoperative instructions

Written instructions used in addition to verbal information reduce confusion and increase compliance. Give particular emphasis to peri-operative fasting and instructions regarding regular medication.

There is controversy as to suitable fasting limits for breast and formula milk, resulting in a lack of uniformity between institutions. Even the concept of solids and liquids is difficult to fully appreciate. For example gelatine (jelly) is ingested as a solid, but turns to liquid in the stomach; cows milk is ingested as a liquid, but turns to solid (curds) in the stomach. There is some evidence that human milk and whey-based formula empties from the stomach faster than cows-milk (casein) based formula. This may be due to the higher protein content of casein formulas. There is great variability in gastric emptying. Table 3 shows some standard fasting guidelines for elective surgery.

Table 3. Preoperative fasting times

	Time before surgery
Solids	6 hours
Milk (formula)	6 hours
Milk (breast)	4 hours
Clear fluids	2 hours

In recent years, there has been a greater emphasis on preventing unnecessary pre-operative fasting in children. There are many reasons to encourage clear fluids up to 2 hours before surgery.

- Infants and small children are less able to tolerate dehydration, especially in hot environments.
- Nausea and vomiting is more frequent in children starved for a long period of time.
- Hypoglycaemia can be avoided.
- Paradoxically, there may be an increase in gastric contents through increased secretion if starvation is prolonged.
- Pre- and postoperative behaviour is improved by minimising fasting times.

Some units require the parent to sign a form at pre-assessment to ensure that they have understood pre-operative instructions.

Consent

The pre-operative visit is the ideal opportunity to gain surgical consent for the procedure if this has not been obtained already. Consent should be obtained from the parent with parental rights, and child if they are old enough to understand. Anaesthetists should understand the local rules about who may give and obtain consent.

In some countries, an older child's consent can be accepted provided they can fully understand

- The procedure
- Potential complications
- The implications of not having the procedure.

Often a child younger than sixteen years is assumed to lack such understanding, but the child may want to countersign the consent

Table 2. Common issues encountered in pre-operative screening and their implications

Problem	Implication for anaesthesia and surgery
Potential airway problems	Airway management in most children is not usually problematic. Abnormal airway anatomy associated with difficult ventilation or intubation may be found in syndromes such as Pierre Robin (airway improves with age), Treacher Collins and the mucopolysaccharidoses (airway worsens with age). Intubation can be extremely challenging in these patients.
Asthma [see article “Anaesthesia in patients with asthma, bronchiolitis and other respiratory disease”, page 58]	<p>Markers for poorly controlled and brittle asthma:</p> <ul style="list-style-type: none"> • Recurrent admission to hospital (including HDU or PICU) • Regular oral steroids and nebulised bronchodilators. <p>Previous use of NSAIDs should be established: while some children's asthma control is worsened by NSAIDs, the majority are not.</p>
Behavioural issues/ additional needs	<p>Attention deficit hyperactivity disorder (ADHD) does not usually present peri-operative problems.</p> <p>Autism: Pre-operative visits may be difficult due to the child's requirement for familiarity and routine.</p> <ul style="list-style-type: none"> • A quiet room with toys helps keep the child calm pre-operatively • Parents will often have successful coping methods • Discuss with the parents: <ul style="list-style-type: none"> - Sedative premedication - Alternative approaches – e.g. successful distraction techniques for the child - When to abort anaesthesia - Possible use of ‘therapeutic restraint’ – this should be avoided if possible. <p>Patients with additional needs/ learning disabilities may need reasonable adjustments to the perioperative routine to allow them to tolerate the hospital experience. These should be discussed case by case with the parents/ carers.</p>
Congenital heart disease [see article page 46]	<p>This may be cyanotic, acyanotic, corrected or palliated.</p> <p>Further investigation and optimisation will depend on functional reserve and previous history and liaison with the cardiologist or paediatrician is essential.</p> <p>Medications should be carefully documented and, in general, continued. Anticoagulants require careful consideration. Consider endocarditis prophylaxis.</p>
Heart murmur	<p>Innocent murmurs tend to be quiet, early in systole, unaccompanied by abnormal signs or symptoms.</p> <p>Pathological murmurs are diastolic, pansystolic or late systolic, loud or continuous, often associated with suggestive signs or symptoms:</p> <ul style="list-style-type: none"> • Failure to thrive • Recurrent chest infections • Reduced exercise tolerance compared to their peers • Hypertension • Radio-femoral pulse delay • Syncope • Cyanotic episodes. <p>If there is any doubt as to the nature of a murmur, it must be further assessed before surgery goes ahead. Give antibiotic prophylaxis as dictated by the surgery being undertaken and current guidelines.</p>

Diabetes	<p>Ask about normal blood sugar and current insulin regimen. Involve the diabetic nurse if possible.</p> <p>Agree careful instructions with respect to peri-operative insulin doses and fasting times.</p> <p>Book first on the list if possible. Regular testing of blood sugar is required peri-operatively.</p>
Down's syndrome [see article , page 27]	<p>Pay particular attention to co-existing cardiac conditions.</p> <p>Seek history suggestive of atlanto-axial subluxation or tracheal stenosis (neck pain, noisy breathing). These may be difficult to detect pre-operatively - maintain a high index of suspicion.</p> <p>Increased incidence of: behavioural problems, obstructive sleep apnoea, leukaemia, thyroid abnormalities, duodenal atresia, Hirschsprung's disease, epilepsy.</p>
Ex-premature babies	<p>Chronic lung disease (including requirement for postoperative oxygen), developmental delay, seizure disorders, gastro-oesophageal reflux and failure to thrive are common. Venous access may be difficult.</p> <p>Peri-operative apnoea and bradycardia are more frequent.</p> <p>Ex-premature babies less than 60 weeks post conceptual age (PCA) should not be discharged home on the day of surgery, and should ideally be monitored with a pulse oximeter for 12 hours postoperatively.</p>
Immunisations	<p>It is essential that the immunisation schedule is not interrupted by surgery. Occasionally, the child may be pyrexial after vaccination, in which case elective surgery may be postponed if the child is unwell.</p>
Obstructive sleep apnoea (OSA)	<p>Signs of OSA include snoring, nocturnal apnoea, daytime somnolence, behaviour disturbances, failure to thrive or obesity.</p> <p>OSA may be due to adenotonsillar hypertrophy or congenital abnormalities such as Aperts syndrome or Pierre Robin sequence, or follow interventions such as pharyngoplasty. Use opioids cautiously and monitor post-operative respiratory function, ideally with a pulse oximeter for 12 hours postoperatively.</p>
Previous bad experiences	<p>Peri-operative behavioural problems especially at induction are more frequent.</p> <p>Plan for alternative approaches: involve parents, play therapy if available, consider sedative premedication.</p>
Repeat procedures and anaesthetics	<p>These patients often have an established peri-operative routine.</p> <p>Anxiety may be minimised by keeping to this routine.</p>
Sickle cell disease [see article, page 35]	<p>This may be evident from the family history or a Sickledex test.</p> <p>Patients with severe disease (frequent painful crises, chest crises, or stroke) or major surgery may require a pre-operative top-up transfusion.</p> <p>The child should be first on the list to avoid excessive dehydration. Give intravenous peri-operative fluids and avoid hypothermia during surgery. Adequate analgesia and early mobilization are essential.</p>
Thalassaemia	<p>Suspect if there is a family history of anaemia in patients from endemic areas.</p> <p>FBC and cardiac investigation may be required.</p> <p>Pre-operative blood transfusion is sometimes necessary.</p>

form to demonstrate assent, when their parents have signed consent. This can give them some positive feelings of control of their situation.

The process becomes complicated in the infrequent event of the child consenting whilst the parent refuses. In many countries, a child under the age of 16 may not refuse consent against their parent's wishes. The legal implications are complex, depending on the country.

Play therapy

Young children learn and process experiences through playing. Some hospitals employ play specialists to assist in preparing children for surgery. The aims are to:

- Establish a rapport through normal play.
- Explore the child's interpretation of their operation.
- Prepare the child for their peri-operative experience, for example, by use of a doll to simulate anaesthetic induction.
- Identify children that fail to engage or those with particular anxiety issues that might be improved with targeted sessions prior to the date for surgery.

Investigations

Most children do not require routine pre-operative screening investigations unless there is a specific clinical indication such as anaemia or sickle cell disease.

Day of surgery

Conduct on the day of surgery will be dictated by whether there has been the opportunity for a pre-assessment visit. It is essential that all children are identified by name-band, consent is signed, and the operation site marked by the surgical team, as recommended by the WHO Safe Surgery Saves Lives team.

All patients should be assessed by the anaesthetist immediately prior to surgery.

The value of the pre-operative visit cannot be overstated: it is a final opportunity to consider the child's fitness for anaesthesia and surgery, and to prepare for the peri-operative period.

The main points to address include:

Fasting

Establish the fasting status. Children who are not adequately fasted should not undergo elective procedures. See above for fasting recommendations.

Preparation of child and parent

Before surgery, the anaesthetist should see the child and parent and conduct a pre-operative assessment.

Wear appropriate dress. An anaesthetist who looks unprofessional will not inspire confidence.

- Speak directly to the child using simple and age-appropriate language in the presence of the parents to help ensure a fuller understanding of proceedings.
- Aim to put yourself at the child's eye level, adapted as best as possible to the child's developmental stage and personality.
- Give an opportunity for the child and parents to ask questions and discuss options.

- Re-affirm or obtain consent.

Parents may be keen to hear about all the treatment alternatives and risks, while young children are often disinterested in these discussions. They tend to focus much more on the practicalities, for example, how long the operation takes, how they are going to feel afterwards, and what activities they may be temporarily excused from.

Clinical assessment of the child

Review the pre-assessment record and note any new history such as a recent upper respiratory tract infection (URTI), gastroenteritis or change in medications. The anaesthetist should examine all children pre-operatively, in the presence of the parents, with particular attention to the cardiorespiratory system. This examination also helps with establishing rapport with the child and can help determine whether the child trusts you and will let you near them!

Upper respiratory tract infection (URTI)

Colds are very common in children, but they may be associated with a 2-10 fold increase in peri-operative respiratory complications such as infection, laryngospasm and wheeze.

Elective surgery should be postponed 2-4 weeks if the child is 'unwell', or if there is any suggestion that the child has a lower respiratory tract infection:

- Fever > 38°C
- General malaise
- Chest signs on auscultation
- Productive cough
- The child is under one year old
- Wheezy child
- Major surgery
- Surgery involving the airway.

Investigations

It is essential that all children are weighed on the day of surgery.

Special investigations are not usually required on the day of surgery. Exceptions include:

- Malaria parasites/full blood count in malaria endemic areas
- Peak expiratory flow rate (for brittle asthmatic children or those with an exacerbation of asthma)
- Blood sugar level (for diabetic children only).

Prescription of premedication if required (see below)

Other Issues

Preparation of environment

Consider a 'child friendly' environment with brightly coloured pictures on the walls and toys (be wary of soft toys for infection control reasons). Devices such as mobile phones with music may provide useful distraction. If possible, allow children to enter the theatres in their own clothes to help maintain normality and dignity.

Premedication

Consider premedication for:

- Anxiolysis
- Analgesia
- Antiemesis
- Antisialagogues
- Antacids
- Topical anaesthesia of suitable veins (using local anaesthetic cream) to reduce the pain associated with cannula placement and to help facilitate an intravenous induction.

There has been a reduction in routine anxiolytic pre-medication over the last decade. Appropriate preparation of the child combined with parental presence at induction is sufficient in the majority of cases. There are still situations where anxiolytics are appropriate:

- Multiple/repeated procedures
- Learning difficulties
- Extreme anxiety
- Uncooperative children.

Anxiolytics should be used with caution if there is a history suggestive of obstructive sleep apnoea due to the increased risk of post-operative apnoea.

Midazolam (0.5mg.kg⁻¹ PO (maximum 20mg), or 0.3mg.kg⁻¹ buccally) is the most commonly used pre-operative anxiolytic. The child is sleepy and cooperative after 15-30 minutes and it has minimal effect on wake-up times. Midazolam tastes bitter, which can be disguised by mixing with a small amount of paracetamol elixir or fruit flavoured drink.

Paracetamol is commonly used PO preoperatively and is preferable to perioperative suppositories as rectal absorption of paracetamol is unpredictable. A small amount of water with tablets or a minimal volume of analgesic elixir does not significantly increase the risk of pulmonary aspiration. NSAIDs such as ibuprofen or diclofenac may also be given.

Induction

The presence of parents at induction has become more common in recent years and reduces peri-operative anxiety in both the parent and child. You will need to prepare the parent with a description of what to expect, their role, and when to leave.

There are certain situations when it may not be appropriate to allow parents to be present at induction:

- Neonates and babies - there is little benefit to children under 6 months in having a parent present.
- Many parents may prefer not to be there for the acute management of a critically ill child.
- The anaesthetist or other staff may find that working under the scrutiny of the parent impairs their ability to treat the child in stressful situations.

EMERGENCY SURGERY

There are additional psychological and physiological issues to consider when preparing children for emergency surgery.

Psychological issues

By their nature, these admissions are unplanned. The child may be quiet and withdrawn due to pain, fear, hypotension or sepsis, with little interest in interacting with their environment (state of passivity). Be alert to a child presenting in this manner - never assume that the child is simply frightened. Immediately examine the child to exclude septic or hypovolaemic shock.

It is important to consider the parent. Parental anxiety can present in a spectrum of behaviour from silence and crying to open aggression. This can often be attenuated by a full explanation of the peri-operative plan.

Physiological issues

All of the conditions outlined in table 2 above can also be present in the acutely ill child and the principles of management are the same. Often patient notes are not immediately available. Balance the need for further investigation of pre-existing disease against the urgency for surgery.

Children presenting for emergency surgery may be critically ill and will require thorough assessment and, if necessary, resuscitation prior to induction. Key aspects of the assessment of the critically ill child are outlined in Table 4.

Inadequate resuscitation of a critically ill child prior to induction can result in severe peri-operative haemodynamic instability. Pre-operative shock may be due to hypovolaemia (e.g. trauma, dehydration, acute abdomen) or sepsis (e.g. abscess, appendicitis). The principles of management of the shocked child should follow the ABC framework, and are discussed further on page 223.

Surgery should only proceed if resuscitation is incomplete for haemostasis in trauma or other resuscitation situations.

Anaesthetic induction and maintenance in emergency situations is difficult – use extreme caution. A rapid sequence induction using judicious (lower than normal) drug doses is usually most appropriate.

More thorough reviews of the management and assessment of the critically ill child, including cardiopulmonary resuscitation, are elsewhere in this *Update* (pages 209, 223, 264).

Fasting

Patients awaiting emergency surgery should be 'nil by mouth' for solids for 6 hours pre-op and clear fluids 2 hours, as for elective surgery. However, a risk-benefit decision may be required depending on surgical urgency (e.g. trauma). Beware the use of pre-operative opioids, which can lead to a significant delay in gastric emptying. Intubation is often prudent in this situation. If in doubt, you should choose intubation over an LMA or facemask to protect the airway. If the child is unable to take oral fluids, consider starting pre-operative IV fluids.

Consent

This should be obtained where possible. Proceed in the child's best interests if emergency life-saving surgery is required and there is no parent or guardian available.

Table 4. Recognition of the critically ill child

	Clinical signs of concern
Airway and breathing: Respiratory rate Respiratory effort Efficacy Effects on other organs	A very sensitive sign. Normal rate varies with age. Tachypnoea may indicate pain, respiratory or circulatory compromise. Recession, grunting, accessory muscle use and gasping are especially concerning. Look for poor expansion or reduced breath sounds on auscultation. Heart rate, skin colour, conscious level will alter if the child is hypoxic.
Circulation: Heart rate Blood pressure Pulse volume Capillary refill Peripheral temperature	May be raised due to shock (septic or hypovolaemic), pain, or anxiety. Bradycardia is a worrying sign and may be pre-terminal. Hypotension is a late sign of shock. Age-normal blood pressure is NOT a good measure of volume status. May be bounding in early septic shock or reduced in severe shock. Five seconds of pressure on the sternum should result in capillary refill in 2-3 seconds. Prolonged refill is a sensitive measure of hypoperfusion. The peripheries may be warm in early septic shock, cold in established shock, or simply cold due to a cool ambient temperature.
Neurological status: Conscious level	Reduced consciousness or lethargy is concerning and may be due to hypovolaemia, sepsis or have a primary neurological cause (e.g. head injury, meningitis).

Investigations

Depending on the pathology, pre-operative blood tests are much more important for emergency surgery. Significant electrolyte imbalance (e.g. pyloric stenosis, bowel obstruction) or anaemia (trauma) may be present. Blood group typing or cross match may also be necessary. Always measure blood sugar concentration in a shocked child.

CONCLUSION

Preparation of children for surgery is essential to ensure a safe perioperative course and pleasant experience for the child. All children should be weighed and examined carefully prior to surgery.

For elective procedures, a pre-assessment visit may be helpful to smooth the preoperative process and avoid cancellations on the day.

Children presenting for emergency surgery may be critically ill and require a thorough physiological assessment to ensure that resuscitation measures are taken prior to induction of anaesthesia.

REFERENCES and FURTHER READING

- Langford R. The preparation of children for surgery. ATOTW (2009) 132. Available from: http://www.wfsahq.org/components/com_virtual_library/media/64cd6bb5b4307b722eedf89c4b4bd7d0-db600de236b211016cf0b4647da75d42-132-preparation-of-children-for-surgery.pdf
- Advanced Life Support Group. Advanced paediatric life support. The practical approach (APLS) 5th Edition. John Wiley & Sons (Wiley-Blackwell), 2011.
- Association of Paediatric Anaesthetists of Great Britain and Ireland. APA Consensus guideline on perioperative fluid management in children. v1.1 September 2007. www.apagbi.org.uk/sites/default/files/Perioperative_Fluid_Management_2007.pdf
- Brennan LJ, Prabhu AJ. Paediatric day-case anaesthesia. *BJA CEPD Review* 2003; **8**: 134-8.
- Ferrari LR, Rooney FM, Rockoff MA. Preoperative fasting practices in pediatrics. *Anesthesiology* 1999; **90**: 978-80.
- McGraw T. Preparing children for the operating room: psychological issues. *Can J Anaesth* 1994; **41**: 1094-103.
- Tait, AR, Malviya, S. Anaesthesia for the Child with an Upper Respiratory Tract Infection: Still a Dilemma? *Anesth Analg* 2005; **100**: 59-65.

Update in Anaesthesia

Perioperative analgesic pharmacology in children

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Summary

It is both desirable and possible to achieve safe, effective analgesia for children of all ages using individualised pain management planning and combined multimodal analgesic techniques.

INTRODUCTION

The inadequate treatment of pain in children following surgery was first highlighted over 20 years ago. A survey at the time found that 40% of paediatric surgical patients experienced moderate or severe postoperative pain and that 75% had insufficient analgesia. Since then increased interest in this area has led to a better understanding of the developmental neurobiology of pain and analgesic pharmacology and, consequently, allowed for the development of safer and more effective analgesic techniques for children of all ages.

PAIN PERCEPTION

During foetal, neonatal and infant life the nervous system is continually evolving. This allows structural and functional changes to occur continuously in response to the child's needs as it grows and develops. The pain pathways mirror these changes with different components developing along differing time frames. For instance the structural components required to perceive pain are present from early foetal life whereas pathways involved in modifying pain perception are still developing during infancy. Also opioid and other receptors vary in their number, type and distribution between early life and adulthood.

The challenge of treating pain in these young age groups is to understand how this changing nervous system affects the child's perception of pain and the efficacy and safety of analgesic treatments. The field is the subject of much research but there are many gaps in our knowledge that need to be filled. Pain is a subjective experience and is thus difficult to assess if communication is not possible. Assessment relies on using non-specific behavioural and hormonal signs of distress/stress. It has been shown in neonates and infants that the use of adequate perioperative analgesia will modify behavioural and hormonal stress responses and reduce morbidity.

pain in children, to minimise moderate and severe pain safely in all children, to prevent pain where it is predictable, to bring pain rapidly under control, and to continue pain control after discharge from hospital.

Once a pain management plan is implemented it should be regularly reassessed and changes made as required. Appropriate pain assessment is vital to aid this. This should involve clinical assessment of the child and the use of an appropriate pain scoring tool to identify discomfort and monitor the efficacy of any analgesic intervention. Due to the subjective nature of pain and the lack of a reliable measure many different tools are available. If the child is able to communicate their pain then a self reporting score, such as the "pain faces" (see figure 1) should be used. If the child cannot communicate then other tools using behavioural and physiological signs should be appropriate, the one chosen from the many available will depend on the age of the child, their neurological and cognitive state, and local preference.

The plan should also include provision for the rapid control of pain that is not alleviated by the original treatment (breakthrough pain) and the identification and treatment of side-effects. In established paediatric centres with high level of resources a dedicated paediatric pain service is the standard of care. Where this is not available significant improvements in pain management can be made by the establishment of clinical routines and protocols for the treatment and assessment of postoperative pain and a network of interested medical and nursing staff to provide ongoing education.

MULTIMODAL ANALGESIA

Multimodal, or balanced, analgesia, involves the simultaneous use of a number of analgesic interventions to achieve optimal pain management. Our increased understanding of pain biology has allowed us to use analgesic techniques that modify nociceptive transmission at different points along the pain pathway. This produces analgesia using minimal doses of drugs, thereby reducing side-effects.

Using a multimodal approach effective pain management is achievable for most cases and the technique can be

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Successful pain management is based on the formulation of a sensible analgesic plan for each individual patient. It is best to take a practical and pragmatic approach dependent on the patient, the type of surgery and the resources available. Realistic aims are to recognise

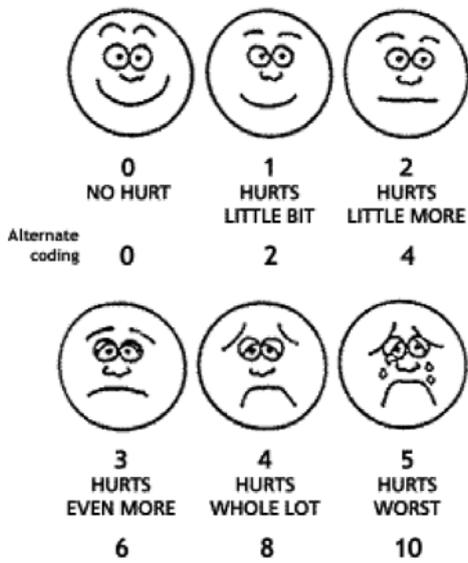


Figure 1. Wong-Baker Faces Pain Rating Scale. Reproduced from Hockenberry MJ, Wilson D, Winkelstein ML: *Wong's Essentials of Pediatric Nursing*, ed. 7, St. Louis, 2005, p. 1259.

adapted for day cases, major cases, the critically ill child, or the very young. In current practice most analgesic techniques are based on differing combinations of four main classes of analgesics: paracetamol, non-steroidal anti-inflammatory drugs (NSAIDs), opioids, and local anaesthetics. Though it is not uncommon for other agents, e.g. NMDA antagonists and alpha-2 agonists, and non pharmacological analgesic methods to be used in addition. Unless there is a contraindication to do so, a local/regional analgesic technique should be used in all cases. For many minor and day case procedures this, in combination with paracetamol and NSAIDs, may allow opioids to be omitted.

Paracetamol (Acetaminophen)

Paracetamol has a mainly central mode of action producing both antipyretic and analgesic effects. It has been shown to inhibit

prostaglandin synthesis in the hypothalamus, reduce hyperalgesia mediated by substance P and reduce nitric oxide generation involved in spinal hyperalgesia induced by substance P or NMDA.

It is the most widely prescribed drug in paediatric hospitals and has become the mainstay base analgesic for almost all procedures. The analgesic potency is relatively low and on its own it is only really effective against mild pain. In combination, however, with NSAIDs and weak opioids it has been shown to be effective for moderate pain and it has also been shown to have an opioid sparing effect when used in tandem with the more potent opioids.

The oral bioavailability of paracetamol is very good as it is rapidly absorbed from the small bowel. Rectal absorption is slow and incomplete, except in neonates. Though the formulations of different brands of suppository vary and the more lipophilic the better the bioavailability. Even so, if paracetamol is given rectally at the start of a short procedure (<1hour) it is unlikely to reach therapeutic plasma levels by the time the child wakes in the recovery room. Thus, if possible, paracetamol should be given orally and pre-operatively. Intravenous paracetamol preparations are becoming widely available in many countries. Studies suggest that it may have a higher analgesic potency than either the oral or rectal preparations though the time to peak analgesia is still 1–2 hours. Irrespective of the route of administration a regular rather than an “as required” post-operative prescription has been shown to provide better analgesia.

In recent years the accumulation of a large body of evidence concerning the use of paracetamol in children has led to the revision of dosing schedules to ensure a balance between efficacy and safety. Smaller doses and longer dosage intervals are required in the neonate or sick child.

Non-steroidal anti-inflammatory drugs (NSAIDs)

NSAIDs act mainly peripherally by inhibiting prostaglandin synthesis and thus reducing inflammation, although central effects have also been postulated involving the opioid, serotonin and nitric oxide pathways. They are highly efficacious in their own right in the treatment of mild to moderate pain in children. They have a reported

Table 1. Dosing guide for oral and rectal paracetamol

Age	Oral: Loading dose	Maintenance dose	Rectal: Loading dose	Maintenance dose	Maximum daily dose	Duration at maximum dose
Pre term <32 weeks	20mg.kg ⁻¹	15mg.kg ⁻¹ up to 12 hourly	20mg.kg ⁻¹	15mg.kg ⁻¹ up to 12 hourly	45mg.kg ⁻¹ .day ⁻¹	48h
32 weeks – 1 month	20mg.kg ⁻¹	15mg.kg ⁻¹ up to 6 hourly	30mg.kg ⁻¹	15mg.kg ⁻¹ up to 6 hourly	60mg.kg ⁻¹ .day ⁻¹	48h
>1 month	20mg.kg ⁻¹	15mg.kg ⁻¹ up to 4 hourly	30-40mg.kg ⁻¹	20mg.kg ⁻¹ up to 6 hourly	90mg.kg ⁻¹ .day ⁻¹	72h

Table 2. Dosing guide for intravenous paracetamol

Age/Weight	Dose	Maximum daily dose
1 month – 50kg	15 mg.kg ⁻¹ up to 6 hourly	60 mg.kg ⁻¹ /day
>50kg	1g up to 6 hourly	4 mg.kg ⁻¹

Table 3. Dosing guide for NSAIDs

Drug	Route	Dose
Ibuprofen	Oral	<20kg: 5-10mg.kg ⁻¹ up to 6 hourly >20kg: 200mg 6 hourly
Diclofenac	Oral/Rectal	1-3mg.kg ⁻¹ up to 3mg.kg ⁻¹ in divided doses

opioid-sparing effect of 30 – 40 % when used in combination with opioids and also reduce opioid-related adverse effects as well as facilitating more rapid weaning of opioid infusions. They have also been shown to be highly effective in combination with local or regional nerve block. Combination with paracetamol produces better analgesia than either drug alone.

There are limitations to their use in paediatric populations. At present, in general, their use is not recommended for children less than 6 months of age, although in the UK Ibuprofen is now available over the counter in formulations for ages 3 months and above. Care should also be taken in those patients who are asthmatic, have a known aspirin or NSAID allergy, are hypovolaemic or dehydrated, renally-impaired, coagulopathic or where there is a significant risk of haemorrhage. A careful history of previous use of NSAIDs should be taken in every case. Different NSAIDs have different side-effect profiles and the relative risks associated with each of the contraindications given will differ between drugs. In the UK the Committee on the Safety of Medicines has classified Ibuprofen and Diclofenac as having the best side-effects profile.

Pharmacokinetic studies have indicated a higher than expected dose requirement in children if scaled by body weight from adult doses. Rectal and oral bioavailability are both good though again for short cases they are best given orally preoperatively. Other routes of administration are available, such as intravenous and topical, though their use in children has been limited so far.

Opioids

As with adults, opioids, and morphine in particular, remain the mainstay of analgesic treatment for the majority of all but minor surgical procedures. The choice of which opioid to use will depend on the patient's medical history, the type of surgery, drug availability, any locally devised protocols and, often, individual anaesthetic preference. The pharmacology of these agents changes during early life and these changes are not consistent between different drugs. When using a particular opioid in neonates and infants it is important to understand the pharmacology of that particular drug in those age groups to ensure both efficacy and safety.

Morphine remains the most commonly used opioid and, consequently, is the most studied. Morphine clearance is decreased and the elimination half-life is increased in neonates compared with infants and older children. Also in neonates the glucuronidation pathways, the main metabolic pathway for morphine, are still developing, slowing morphine metabolism and giving a relatively increased production of morphine-6-glucuronide, an active metabolite of morphine. These differences may to some extent account for the increased efficacy

of morphine seen clinically in neonates. Codeine however, another popular opioid in neonates and infants, works via metabolism to morphine. The cytochrome P450 enzyme responsible for this conversion shows markedly reduced activity at these ages compared with that seen in older children and adults. Thus it may be that little or no morphine is produced from a dose of codeine. This may explain codeine's good safety profile in young children but may also suggest that the analgesic efficacy is questionable. Recently there has been increased awareness of children suffering respiratory depression following codeine, this is thought to be due to ultra-fast metabolism converting codeine into its active metabolite, morphine. In UK it is no longer recommended to use codeine in any child under 12 years old for these reasons. It is not possible to predict who will metabolise codeine quickly or poorly.

Many routes for the administration of opioids are available in children. During surgery, with an IV cannula in situ, the intravenous route is the easiest. In terms of bioavailability and consistency of effect it is also the most reliable. For these reasons it is usually the route of choice postoperatively, especially after major surgery. Safety, however, must always be a priority. Potentially serious complications such as over-sedation and respiratory depression can occur even when using well constructed protocols for opioid use. With infusion or bolus techniques safe practice must include the presence of appropriately educated staff and regular observation of sedation and respiratory rate. Oxygen and opioid antagonists should be easily accessible in case of emergency. If these are not available then other routes of administration or analgesic strategies are indicated. It is sensible for each institution to devise protocols for opioid use dependent on their own local resources.

Oral and rectal formulations of many opioids are available and have a place for some patients in the perioperative period, especially if safety is a prohibitive local issue for other methods of delivery. If the IV route is available postoperatively, continuous IV infusions and intermittent intravenous boluses have been shown to be safe and effective. Where local resources allow, IV opioids are usually administered via patient (PCA) or nurse (NCA) controlled delivery systems which demonstrate good efficacy and safety in all age groups, even neonates. The subcutaneous route remains an alternative to IV administration but absorption is not as reliable and it should not be used in hypovolaemic patients. Intramuscular injection demonstrates slow absorption and unreliable effect and is considered undesirable in the awake child due to the pain and distress of the injection. Other routes of administration are available, e.g. sublingual, transdermal and intranasal and may be appropriate in specific cases or where local resources dictate their use. Opioids are also commonly used as adjuncts to local anaesthetics.

The extensive study and use of morphine in all age groups has led to the formulation of safe and effective dosing strategies in children, provided appropriate monitoring is used:

Titration loading dose of IV morphine

50mcg.kg⁻¹ increments, repeated up to x4

Ongoing bolus dose of IV morphine

20mcg.kg⁻¹ per bolus, repeated to effect

Oral morphine*

80mcg.kg⁻¹ 4hrly (1 month – 1yr)

200 – 400mcg.kg⁻¹ 4 hourly (>1yr)

IV or s.c. morphine infusion

10 – 40mcg.kg⁻¹.hr⁻¹

PCA with morphine

Bolus dose 10 – 20mcg.kg⁻¹

Lockout interval 5min

Background infusion 0 – 4mcg.kg⁻¹.hr⁻¹

NCA with morphine

Bolus dose 10 – 20mcg.kg⁻¹

Lockout interval 20 - 30min

Background infusion 4 – 20mcg.kg⁻¹.hr⁻¹**

* Can be used in neonates at 80mcg/kg but may need to increase dosage interval to 6 hourly and child must be closely monitored.

**For neonates a background dose is omitted to reduce the possibility of a prolonged effect of the morphine. This allows the carer to use the bolus function to titrate the analgesia and anticipate painful episodes.

Local Anaesthetics (see chapters on specific local anaesthetic blocks)

Local anaesthetic agents work by blocking the conduction of nociceptive stimuli along the pain pathway. This can be achieved by many different routes, the commonest being central/regional blocks, plexus blocks, peripheral nerve blocks, infiltration at the site of injury, and topical application. Some method of nerve blockade is appropriate for nearly all surgical procedures and forms an important part of a balanced analgesic technique. Though, once again, the technique used will depend on the patient’s medical history, the type of surgery, any locally devised protocols and, often, individual anaesthetic preference.

Regional analgesia produces excellent perioperative analgesia for major surgery at all age groups, even preterm neonates, and has been shown to decrease postoperative complications. The evidence for this, however, is limited as is the evidence for the risks associated with paediatric epidural analgesia. In general these techniques should only be performed by experienced practitioners with appropriately trained staff and protocols and monitoring guidelines available for the postoperative period. Plexus or peripheral nerve blocks have been shown to have good efficacy and safety for limb and head and neck surgery. In some procedures involving the lower limbs the analgesia obtained from a peripheral nerve block has a much longer duration of action than that obtained from a single shot central block.

For many years bupivacaine has been the local anaesthetic of choice in paediatric practice. It has been extensively studied and safe-dosing guidelines have been established which have greatly reduced the incidence of systemic toxicity. Neonates demonstrate decreased clearance and decreased protein binding of local anaesthetic agents. Therefore, at this age, there is a risk of systemic toxicity and dosing schedules have to be adjusted. Although thankfully rare, bupivacaine still carries the risk of significant cardiotoxicity and this has led to the increasing introduction of the newer agents, ropivacaine and levo-bupivacaine (chirocaine), into current practice. There are now sufficient paediatric data to recommend either of these agents. Adjuncts such as opioids, adrenaline, ketamine and clonidine are commonly added to local anaesthetic agents. This is mainly seen with regional/central blockade. They allow for increased duration and spread of the block with minimal increase in side-effects, due to the low doses used.

DISCHARGE FROM HOSPITAL

A sensible analgesic plan should include provision for analgesia to continue once the child has been discharged from hospital. Clear and easy to follow instructions should be given to aid compliance and thus efficacy. Inadequate pain relief at home, as in hospital, will lead to unacceptable distress for both child and carer and the potential for complications. In general an assessment is needed of the likely severity and duration of the pain. Again a multi-modal approach should be adopted and regular dosing schedules are superior to “as required” prescriptions when the pain is likely to be significant.

If a single bolus local anaesthetic technique has been used then the time that this will wear off must be anticipated and analgesic provision made. Often analgesia is required for a significant period of time after discharge. For instance it is well described that children still experience significant pain 7-10 days following adenotonsillectomy.

Table 4. Dosing guide for bupivacaine, levobupivacaine and ropivacaine:

<p>Single bolus injection</p> <p>Neonates</p> <p>Children</p>	<p>Maximum dosage</p> <p>2mg.kg⁻¹</p> <p>2.5mg.kg⁻¹</p>
<p>Continuous Infusion</p> <p>Neonates</p> <p>Children</p>	<p>Maximum infusion rate</p> <p>0.25mg.kg⁻¹.hr⁻¹</p> <p>0.5mg.kg⁻¹.hr⁻¹</p>

SUGGESTED READING

Williams G. Perioperative analgesic pharmacology in children. ATOTW (2006) 23. Available from: http://www.wfsahq.org/components/com_virtual_library/media/e558e631b7d38a387f407ae65805690c490d426becffb3927dcc16bd60ef9805-23-perioperative-analgesic-pharmacology-for-children.pdf

1. Lonnqvist P-A, Morton NS. Postoperative analgesia in infants and children. *Br J Anaesth* 2005; 95: 59 – 68.
2. Berde CB, Sethna NF. Analgesics for the Treatment of Pain in Children. *N Engl J Med* 2002; 347: 1094 – 103.
3. Howard RF, Lloyd-Thomas A. Pain management in children. In: Sumner E and Hatch DJ eds. *Paediatric Anaesthesia*. London, U.K. Arnold. 1999.
4. <http://onlinelibrary.wiley.com/doi/10.1111/pan.2012.22.issue-s1/issuetoc#group1>. Good Practice in Postoperative and Procedural Pain Management, 2nd Edition, 2012 (79 page special issue, free to download).
5. Association of Paediatric Anaesthetists & Royal College of Paediatrics and Child Health. Guidance for the administration of codeine and alternative opioid analgesics in children. 2013. http://www.apagbi.org.uk/sites/default/files/images/Codeine_Nov2013.pdf.

Paediatric procedural sedation

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INTRODUCTION

Sedation is the use of drugs for reduction of fear and anxiety, induction of drowsiness or sleep and comfort.

All sedation techniques carry risk and many procedures done under sedation are performed outside operating theatres. Sedation in children needs special considerations, and some drugs used for sedation are not as reliable as those for anaesthesia. Its safety and success depends upon skill and judgement.

DEFINITIONS OF SEDATION

Sedation is a continuum from the awake state. The American Society of Anaesthesiologists uses the following definitions for levels of sedation:

Minimal sedation (formerly known as anxiolysis) is a drug-induced state during which patients respond normally to verbal commands. Although cognitive function and coordination may be impaired, respiratory and cardiovascular stability is unimpaired.

Moderate sedation (formerly known as conscious sedation) is a drug-induced depression of consciousness during which patients respond purposefully to verbal commands, either alone or accompanied by light tactile stimulation. It is important to remember that reflex response to a painful stimulus is not a purposeful response. No interventions are required to maintain a patent airway, and spontaneous ventilation is adequate. Cardiovascular stability is usually maintained.

Deep sedation/analgesia is a drug-induced depression of consciousness during which patients cannot be easily roused but respond purposefully following repeated or painful stimulus. Patients may require assistance in maintaining a patent airway, and spontaneous ventilation may be inadequate. Cardiovascular stability is usually maintained.

In the UK deep sedation is considered to be a part of the spectrum of general anaesthesia.

GOALS OF SEDATION

The goals of sedation in the paediatric patient for diagnostic and therapeutic procedures are to:

- Guard the patient's safety and welfare
- Minimize physical discomfort and pain

- Control anxiety, minimize psychological trauma and maximize the potential for amnesia
- Control behaviour and/or movement to allow the safe completion of the procedure
- Return the patient to a state in which safe discharge from medical supervision is possible.

PATIENT SELECTION

You must carefully assess the patient with detailed history and clinical examination; this is important to identify potential risk factors. [See also article on 'Preparation of children for surgery' in this edition of *Update*, page 65] Previous sedation history is important as previously failed sedation may indicate a need for general anaesthesia. Although the ASA classifications are not totally appropriate for paediatrics, the Scottish Intercollegiate Guidelines Network advises that only patients in ASA classes I and II should be considered suitable for sedation as outpatients. Patients in classes III to V should be regarded as high risk patients who should only be managed in a hospital setting with the involvement of an anaesthetist trained in paediatric sedation, anaesthesia and resuscitation.

INDICATIONS FOR SEDATION

Painless procedures:

- Transthoracic echocardiography
- Radiotherapy
- Computed tomography
- MRI
- Electroencephalography.

Painful procedures:

- Minor, painful oncology procedures
- Interventional radiology
- Dental procedures
- Wound care, including burns dressings
- Cardiac angiography
- Fracture manipulation.

Summary

Sedation in children needs special considerations, and some drugs used for sedation are not as reliable as those for anaesthesia. Its safety and success depends upon skill and judgement. If you act as a sedation practitioner, you should be trained in sedation techniques.

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CONTRAINDICATIONS TO SEDATION

Children with any of the following should not normally be sedated

- Abnormal airway. This includes adenotonsillar hypertrophy causing obstruction to breathing when asleep (Obstructive Sleep Apnoea, or OSA), or any other anatomical abnormality of upper and lower airway
- Raised intracranial pressure
- Depressed conscious level
- History of sleep apnoea (OSA)
- Respiratory failure
- Cardiac failure
- Neuromuscular disease
- Bowel obstruction
- Active respiratory tract infection
- Known allergy to sedative drug / previous adverse reaction
- Child too distressed despite adequate preparation
- Older child with severe behavioural problems
- Refusal by the parent / guardian / child.

The following subgroup of patients should not be sedated with nitrous oxide

There are specific contraindications to the use of nitrous oxide due to its ability to diffuse into enclosed air spaces causing them to expand or increase in pressure, or in the case of pulmonary hypertension, as it increases pulmonary vascular resistance.

- Intracranial air (e.g. after skull fracture)
- Pneumothorax, pneumopericardium
- Bowel obstruction
- Pneumoperitoneum
- Pulmonary cysts or bullae
- Lobar emphysema
- Severe pulmonary hypertension.

Extra caution should be exercised when sedating children who have any of the following conditions

- Neonates, especially if premature or ex-premature
- Children with cardiovascular instability or impaired cardiac function
- Renal impairment
- Hepatic impairment
- Severe respiratory disease
- Gastro-oesophageal reflux
- Impaired bulbar reflexes

- Emergency cases who are not adequately starved
- Anticonvulsant therapy
- Children receiving opioids and other sedatives
- Children receiving drugs that potentiate the action of sedatives (e.g. macrolide antibiotics such as erythromycin potentiate and prolong the sedative effects of midazolam).

PATIENT PREPARATION AND MONITORING

During procedural sedation when protective airway reflexes are lost, gastric contents may be regurgitated into the airway. Therefore, patients with a history of recent oral intake or with other known risk factors for aspiration should not be sedated.

Appropriate intake of food and liquids before elective sedation

<i>Ingested Material</i>	<i>Minimum Fasting Period (Hours)</i>
Clear liquids: water, fruit juices without pulp, carbonated beverages, clear tea, black coffee	2
Breast milk	4
Infant formula / non human milk	6
Solids	6

Systematic approach to sedation

It is important to use a systematic approach to sedation techniques so as to not overlook an important drug, piece of equipment, or monitor that may be required at the time of a developing emergency. To avoid this problem, it is helpful to use an acronym that allows the same setup and checklist for every procedure (see Figure 1).

WHO SHOULD ADMINISTER SEDATION?

If you act as a sedation practitioner, you should be trained in sedation techniques. You should be competent to obtain consent, prescribe and administer sedative drugs, understand the pharmacology of the agents used and be capable of providing Paediatric Basic Life Support and preferably Paediatric Advanced Life Support. For deep sedation this should be an anaesthetist or a practitioner with training in anaesthesia.

DRUGS USED FOR SEDATION

A drug can only be considered 'safe' after experience in hundreds and thousands of cases; however, few drugs have been studied to this extent. Good protocols are important for the safety and success of sedation.

Choral hydrate and Triclofos

Choral hydrate and triclofos are effective oral sedatives and are metabolised to trichlorethanol. Chloral hydrate has an unpleasant taste and causes gastric irritation; triclofos is more palatable but is slower and less potent (1g triclofos = 600mg chloral hydrate). Respiratory complications, vomiting and paradoxical reactions can occur. Deaths have occurred in unattended children. Small children are calmed by 'sub-sedation' doses.

Figure 1: A system for planning sedation

A commonly used acronym that is useful in planning and preparation for a procedure is SOAPME:

S	Suction	Size-appropriate suction catheters and a functioning suction apparatus (e.g. Yankauer-type suction)
O	Oxygen	Adequate oxygen supply and functioning flow meters/other devices to allow its delivery
A	Airway	Size-appropriate airway equipment (nasopharyngeal and oropharyngeal airways, laryngoscope blades and handles [checked and functioning], tracheal tubes, stylets/bougies, face mask, bag-valve-mask or equivalent device [functioning])
P	Pharmacy	All the basic drugs needed to support life during an emergency, including antagonists as indicated
M	Monitors	Functioning pulse oximeter with size appropriate oximeter probes. Other monitors as appropriate for the procedure (e.g., non-invasive blood pressure, end-tidal carbon dioxide, ECG, stethoscope)
E	Equipment	Special equipment or drugs for a particular case (e.g. defibrillator)

This table summarizes the expected clinical responses and the ideal monitoring requirements for different levels of sedation:

	Minimal sedation	Moderate sedation	Deep sedation
<i>Responsiveness</i>	Normal	Purposeful to light stimulation	Purposeful to painful stimulation
<i>Airway</i>	Unaffected	No intervention	(±) Intervention
<i>Ventilation</i>	Unaffected	Adequate	(±) Inadequate
<i>Cardiac stability</i>	Unaffected	Maintained	(±) Maintained
<i>Monitoring</i>	Observation & intermittent assessment	Pulse oximetry Heart rate Intermittent recording of RR and BP	Pulse oximetry ECG – continuous BP every 3-5 minutes (±) EtCO ₂ , precordial stethoscope

Benzodiazepines

Midazolam

Midazolam induces anxiolysis, sedation and amnesia; it is absorbed enterally and via oral and nasal mucosa. By mouth, 0.5mg.kg⁻¹ (maximum 20mg, 30 min beforehand) reduces crying during induction of anaesthesia, but occasionally dizziness, dysphoria and paradoxical reactions occur. Its bitter taste needs masking with a sweetening agent. In the emergency department, 0.5-1mg.kg⁻¹ orally is useful to calm children for suture of lacerations. Intranasal drops 0.2 mg.kg⁻¹ effectively calms irritable infants but this method is unpleasant and causes crying - an atomizer may be better. Absorption is so rapid that apnoea and desaturation occur occasionally.

Sublingual administration is more pleasant, equally rapid and effective, but requires co-operation. Rectally, 0.3-1mg.kg⁻¹ may cause moderate sedation.

IV titration is best but effects are variable, unpredictable and depend upon the discomfort of the procedure (0.05-0.2mg.kg⁻¹ for moderate sedation). Co-administration of opioids increases the risk of apnoea while co-administration of macrolide antibiotics may result in prolonged unconsciousness due to inhibition of hepatic metabolism. Occasionally children may develop paradoxical excitation and anxiety (confusion/disinhibition).

Diazepam

Intravenous diazepam (Diazemuls) is 4-5 times less potent than midazolam. Despite a longer elimination half-life, recovery profiles are similar (usually by 2h). Dose- 200-300microgram.kg⁻¹ orally and 100-200microgram.kg⁻¹ IV.

Temazepam

Temazepam tablets are preferred to the taste of the elixir and oral doses of 0.5-1mg.kg⁻¹ cause minimal sedation and sleep.

Reversal of benzodiazepine sedation

Flumazenil 20-30microgram.kg⁻¹ IV can be used to reverse benzodiazepine sedation. There may be a risk of fitting from sudden benzodiazepine withdrawal. As the half-life of flumazenil is less than that of some benzodiazepines, there is a risk of re-sedation.

Barbiturates

Thiopental

Intravenous thiopental is too potent for non-anaesthetists to use safely. When given rectally in children, thiopental 25-50mg.kg⁻¹ produces sedation after 30 min. Airway obstruction can occur and recovery takes between 30 and 90 min.

Pentobarbital and quinalbarbital

Quinalbarbital (7.5-10mg.kg⁻¹ orally) makes 90% of children (<5 yr) sleep but older children may have paradoxical excitement.

For painless imaging, pentobarbital 2-6mg.kg⁻¹ IV is very successful but 1-3% of children have airway obstruction or paradoxical reactions. Pentobarbital is not available in the UK.

Propofol

The short action and lack of side effects make propofol the best of all the IV agents but, because apnoea and desaturation are common, it is not recommended for non-anaesthetists. Sedation is induced by 2-4mg.kg⁻¹ and usually maintained by an infusion of 6-8mg.kg⁻¹.h⁻¹; recovery is pleasant and occurs within a few minutes. Tolerance and behavioural disturbances are reported.

Melatonin

Natural sleep may be induced successfully in 55% for MRI and 80% for EEG. Doses range from 2-10mg orally.

Opioids

Morphine

Morphine is useful for painful procedures such as wound care. A dose of 60microgram.kg⁻¹ IV has been used in combination with midazolam 0.05mg.kg⁻¹ IV without major respiratory effects.

Meperidine

Meperidine 0.5-1mg.kg⁻¹ IV combined with midazolam 0.05-0.1mg.kg⁻¹ IV provides effective sedation for endoscopy. However, oxygen desaturation has been reported in cases.

Fentanyl

The potency of fentanyl increases the risk of apnoea. For example, 5% of children given IV midazolam and fentanyl (1-6mcg.kg⁻¹) for gastroscopy required reversal with naloxone. Fentanyl is absorbed from the mucosa of the mouth and oral transmucosal fentanyl citrate is available both as a lozenge and a palatable lollipop; side effects include vomiting (30%) and desaturation.

Reversal of opioid-induced respiratory depression

Opioid-induced respiratory depression can be reversed with naloxone. The usual dose is 10microgram.kg⁻¹ IV, repeated as necessary.

Major tranquillizers

Trimeprazine

Trimeprazine 3-4 mg.kg⁻¹ orally causes sleep in 50% of children before anaesthesia. However, because of reports of hypotension, the maximum recommended dose is 2mg.kg⁻¹. At this dose, it can be combined with morphine 0.2mg.kg⁻¹ IM for sedation of children >15kg for MRI.

Chlorpromazine and promethazine

Chlorpromazine and promethazine have been combined together with meperidine (pethidine) to form pethidine compound (1ml contains 25 mg meperidine, 6.25mg chlorpromazine and 6.25mg promethazine). It is for IM administration only and combines analgesia, anxiolysis and sedation; effective doses are between 0.06 and 1ml.kg⁻¹. This powerful combination can cause apnoea.

Nitrous oxide

Nitrous oxide provides valuable analgesia and sedation in cooperative children for a wide variety of painful procedures. Loss of consciousness can occur when combined with other sedatives or when used alone in concentrations over 50%.

Ketamine

This anaesthetic drug causes a 'dissociative' sedation or anaesthesia with analgesia. In maintaining cardio-respiratory function, ketamine (IV or IM) is extremely useful when other methods of anaesthesia are unavailable or impractical. If non-anaesthetists use ketamine they must be prepared for laryngospasm and apnoea. Apnoea has occurred following 4mg.kg⁻¹ IM and is more likely if ketamine is combined with opioids. Nausea and vomiting can occur in 15-33% and distressing hallucinations in 3% even when combined with midazolam. For needle-phobic children, 5mg.kg⁻¹ orally causes variable sedation after 10-20min, and 10mg.kg⁻¹ makes 50% of children unconscious; recovery can take up to 2 hours.

KEY REFERENCES

Sethi DS, Smith J. Paediatric Sedation. ATOTW (2008) 105. Available at: http://www.wfsahq.org/components/com_virtual_library/media/477f2ba f45cc2bb9a4494be0d922870a-788e4c95dcdadbf9e5a5af36b465ebff-105-Paediatric-sedation-v2.pdf

SIGN. Safe sedation of children undergoing diagnostic and therapeutic procedures. A national clinical guideline. Scottish Intercollegiate Guidelines Network, 2004 <http://www.sign.ac.uk/guidelines/fulltext/58/index.html>

Editorial note - guideline has been withdrawn by SIGN since original publication of this article.

Charles J. Cote, Stephen Wilson and the Work Group on Sedation, Guidelines for Monitoring and Management of Pediatric Patients During and After Sedation for Diagnostic and Therapeutic Procedures: An Update. *Pediatrics* 2006;118;2587-2602 <http://pediatrics.aappublications.org/content/118/6/2587.full.pdf+html>

Cote CJ, Notterman DA, Karl HW, Weinberg JA, McCloskey C. Adverse sedation events in pediatrics: a critical incident analysis of contributing factors. *Pediatrics* 2000; 805-814

M R J Sury Paediatric Sedation, British Journal of Anaesthesia CEPD, Number 4, Volume 4 August 2004.

M R J Sury, Anaesthesia and sedation outside operating theatres, Chapter 44 in: Hatch and Sumner's Textbook of Paediatric Anaesthesia (2007 Edition). Publisher – Hodder Arnold.

Perioperative fluids in children

Reprinted with revisions from: Wilson CM. Perioperative fluids in children. *Update in Anaesthesia* 2005; 19: 36-8.

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INTRODUCTION

Starvation, surgery and anaesthesia cause stress and alter physiology. Intravenous fluids are administered perioperatively to maintain homeostasis during this period. Water and electrolytes are required to correct deficits and ensure adequate intravascular volume, cardiac output and ultimately tissue oxygen delivery. Calories in the form of dextrose may be needed to prevent hypoglycaemia.

The majority of healthy children undergoing minor surgery (e.g. circumcision, hernia repair) will be able to drink in the early postoperative phase and will not need intravenous fluids before, during or after surgery. Fasting times should be observed so that children are not left without fluid intake for longer than necessary. The fasting guidelines for elective surgery are shown in Table 1.

Patients undergoing longer or more major procedures, or anyone compromised by underlying problems, will need intravenous fluids during surgery.

Fluids are given for three reasons:

- Resuscitation: to correct pre-existing hypovolaemia or dehydration.

- Maintenance: to provide water, electrolytes and glucose during starvation.
- Replacement: of ongoing losses due to evaporation from an open wound or via the humidification of dry inspired gases, bleeding, pyrexia, gastrointestinal and third space losses (fluid leak into tissues) during surgery and into the post-operative period.

RESUSCITATION

Any child who is dehydrated or hypovolaemic should be resuscitated prior to surgery unless the nature of the illness and operation precludes this. In this case rapid correction of hypovolaemia should commence at, or as soon as possible after induction to maintain circulating volume and cerebral perfusion.

Hypovolaemia (losses from the intravascular space) should be corrected rapidly, initially with boluses of 10-20ml.kg⁻¹ isotonic crystalloid (0.9% saline, Hartmann's, Ringer's lactate, Plasma-Lyte). Blood should be considered if the haemoglobin or haematocrit are low (Hct less than 25%) or more than 40ml.kg⁻¹ of fluid is required.²

Summary

This article is a revision of one written for Update 10 years ago. Changes in the intervening years include greater awareness and more defined acknowledgement of the dangers of using hypotonic fluids, and increasing evidence that isotonic crystalloids (0.9% saline, Hartmann's, Ringer's lactate, Plasma-Lyte) are the fluid of choice for resuscitation and maintenance during surgery. New on the horizon is the Fluid Expansion as Supportive Therapy (FEAST) study for African children with severe infection; this will be considered at the end of this article.¹

Table 1. Fasting guidelines for elective surgery

Type of food/fluid	Minimum fasting time (hours)
Clear liquids	2
Breast milk	4
Infant formula	4 (<3 months old) 6 (> 3 months old)
Non-human milk	6
Light meal	6

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Dehydration (total body water loss) should be corrected more slowly, preferably by the oral route if tolerated and time allows, but otherwise intravenously. The rapid rehydration technique advocated by Assadi and Copelovitch describes an initial rapid (1-2 hours) infusion of isotonic saline up to 60-100ml.kg⁻¹ as required to correct hypovolaemia.³ This is followed by a slower correction of dehydration over 24-72 hours with 0.9%, 0.45% or 0.2% saline depending on measured plasma sodium. The plasma sodium should be measured at regular intervals (at least 6 hourly if outside the normal range). Too rapid correction of dehydration with hypotonic fluid will result in cerebral oedema secondary to hyponatraemia.

An otherwise healthy child starved pre-operatively will have a fluid deficit. The size of the deficit may be calculated by multiplying the hourly maintenance requirement (see table 2) by the number of hours starved. The majority of children do not require replacement of this deficit if the period of starvation for fluid is short. If the child has been starved for a longer period, for instance overnight, and has not had any fluid orally preoperatively, the deficit can be replaced during surgery using a bolus of 10-20ml.kg⁻¹ isotonic crystalloid. For children undergoing minor day case surgery this ensures the child is well hydrated and may possibly reduce postoperative nausea and vomiting.

MAINTENANCE FLUIDS – THE ‘4,2,1 RULE’

Maintenance fluid requirements have been calculated a number of ways including by estimation of expenditure of calories and body surface area. The simplest and most commonly used formula was devised by Holliday and Segar and modified by Oh, and is known as the ‘4,2,1’ rule.^{4,5} The formula relates energy (calorie) expenditure, and volume of fluid required to

the weight of the child in kg (see Table 2 below).

Electrolyte and glucose requirements were also estimated depending on the weight of the child (dietary sodium and potassium = 1.2mmol.kg⁻¹.day⁻¹). An “ideal” maintenance solution was proposed that contained the maintenance requirements for water and sodium in one bag, with dextrose added to make the fluid isotonic with the vessels (0.2% saline in 5% dextrose in the US, 0.18% saline in 4% dextrose in the UK, with added potassium chloride (KCl) 20mmol.l⁻¹ if required). This solution became the mainstay of IV maintenance fluid for many years, however its validity has recently been questioned and the use of isotonic fluid advocated instead, particularly in the perioperative period.^{7,8,9,10}

Neonates (up to 44 weeks post-conceptual age) have a slightly different fluid requirement. They are born physiologically “waterlogged” but then lose up to 10% of their body weight in the first week of life.

They must not be given too much water or sodium on the first few days of life, so much smaller maintenance volumes are prescribed initially, which then increase over the next few days. Premature or low birth weight babies have a greater surface area to weight ratio, lose more water by evaporation and consequently require more replacement fluid (Table 3). The fluid is usually given as 10% dextrose with saline added only after the postnatal diuresis has occurred (i.e around day 3 of life).¹¹

REPLACEMENT

Measured losses should be replaced with a similar fluid to that lost. This will usually be isotonic crystalloid or blood to replace haemorrhage and to avoid unacceptably low haemoglobin levels.

Table 1. Holliday and Segar formula and Oh modification

	Holliday and Segar	Oh
Body weight		
1-10kg	4ml.kg ⁻¹ .hour ⁻¹	4ml.kg ⁻¹ .hour ⁻¹
10-20kg	40ml.hour ⁻¹ + 2 ml.kg ⁻¹ .hour ⁻¹ for each kg above 10kg	20 + (2 x weight in kg) in ml.hour ⁻¹
>20kg	60ml.hour ⁻¹ + 1 ml.kg ⁻¹ .hour ⁻¹ for each kg above 20kg	40 + (weight in kg) in ml.hour ⁻¹

For example a child of:

9kg requires $4 \times 9 = 36 \text{ ml.hour}^{-1}$

18kg requires $40 + (2 \times 8) = 56 \text{ ml.hour}^{-1}$

or, using the Oh version of the formula:

$20 + (2 \times 18) = 56 \text{ ml.hour}^{-1}$

36kg requires $60 + 16 = 76 \text{ ml. hour}^{-1}$

or, using the Oh formula:

$40 + 36 = 76 \text{ ml. hour}^{-1}$

Table 3. Paediatric surgical unit fluid guidelines for neonates at Sheffield Children's Hospital, taking postnatal weight and age into account

Age	Fluid requirement (ml.kg ⁻¹ .day ⁻¹)			
	<1.0kg	1.0-1.5kg	1.5-2.0kg	>2.0kg
Day 1	100-200	80-100	60-80	40-60
Day 2	120-150	110-130	90-110	60-90
Day 3	150-170	140-160	120-140	80-100
Day 4	180-200	160-180	140-160	100-120
Day 5	180-200	170-200	150-180	120-150

Fluid evaporation from an open wound or 3rd space losses varies depending on the operation and may be up to 20 ml.kg⁻¹.hour⁻¹. Loss of fluid via the respiratory tract due to humidification of inspired gas may be reduced by using a circle system or HME (heat and moisture exchange filter) in the breathing circuit.

Neonates have a large ECF volume relative to adults so greater 3rd space losses. Replacement with colloids (specifically 4.5% albumin) is more common in neonatal practice than in older children.

Blood or other fluid loss is often difficult to measure especially when irrigation fluids are used. For this reason the child's clinical state should be monitored continuously looking at heart rate, capillary refill time and blood pressure. In longer or more complicated cases core-peripheral temperature gradient, urine output (volume and osmolarity), invasive blood pressure and central venous pressure should be measured. In a warm and otherwise stable child with good analgesia, a rise in heart rate and prolonged capillary refill time are reliable indicators of fluid loss; hypotension due to hypovolaemia occurs relatively late.

WHICH FLUIDS AND WHY?

Isotonic fluids

An isotonic fluid contains the same concentration of solutes as plasma, and therefore exerts an equal osmotic force. Dextrose is metabolised in blood, so although 5% dextrose solution is isosmolar to plasma, and isotonic in vitro, once given, the dextrose is metabolised and it effectively becomes water. Dextrose solutions, unless they contain electrolytes of an equivalent amount to plasma are therefore termed hypotonic fluids. Table 4 shows the electrolyte content of different IV fluids.

Hyponatraemic encephalopathy in children

Children given hypotonic fluid may become hyponatraemic.^{7,8,9,10} Ordinarily the kidneys will excrete a free water load rapidly, and homeostasis is maintained. When

the body is subjected to stress such as surgery, pain, nausea or hypovolaemia, antidiuretic hormone (ADH) levels rise. ADH blocks the renal excretion of water; water is conserved, and plasma sodium levels fall. Even the relatively mild hypovolaemia of pre-operative starvation causes a rise in ADH levels.¹² If the plasma sodium falls rapidly to a low level (acute hyponatraemia), water moves into the cells in compensation and causes swelling of the cells. The brain is particularly vulnerable to acute hyponatraemia; this can manifest as cerebral oedema, raised intracranial pressure, and can cause brain stem herniation, coning and death. Prepubertal children are particularly susceptible to brain damage associated with postoperative hyponatraemic encephalopathy. Retrospective analyses of patients with acute hyponatraemia have shown that more than 50% of children develop symptoms when the plasma sodium is less than 125mmol.l⁻¹, and that there is a mortality of 8.4% for severe acute hyponatraemia.^{7,8}

Acute hyponatraemic encephalopathy typically presents with non-specific features such as nausea, vomiting and headache; if untreated, this will progress to reduced level of consciousness, seizures, respiratory depression and death. Nausea, vomiting and drowsiness may be attributed to the side effects of anaesthesia, but unfortunately by the onset of seizures and respiratory depression it may be too late to salvage the situation. A high index of suspicion should be maintained in all children receiving IV fluids; hypotonic fluids should NEVER be given in the perioperative period (see below). If there are any concerns about hyponatraemia, plasma electrolytes should be measured urgently.

Acute symptomatic hyponatraemia presenting with seizures is a medical emergency. A typical case is as follows:

A healthy 9-year-old presented for routine elective surgery. He was given 4% dextrose 0.18% saline at maintenance rate during the operation, and the fluid was continued postoperatively. He was slow to get going after surgery, complaining of headache and nausea. IV fluids were continued. At 4.00am he suddenly developed a seizure. Electrolytes taken at this time showed a plasma sodium of 123mmol.l⁻¹.

Table 4. Electrolyte content of different IV fluids

Fluid	Na ⁺ mmol.l ⁻¹	K ⁺ mmol.l ⁻¹	Cl ⁻ mmol.l ⁻¹	HCO ₃ ⁻ mmol.l ⁻¹	Osmolality mOsm.l ⁻¹	Tonicity*	Other	pH
5% dextrose	0	0	0	0	252	Hypotonic	Dextrose 50g	4.0
4% dextrose 0.18% saline	30	0	30	0	284	Hypotonic	Dextrose 40g	4.0
5% dextrose 0.45% saline	75	0	75	0	432	Hypotonic	Dextrose 50g	4.0
5% dextrose 0.9% saline	150	0	150	0	586	Hypotonic	Dextrose 50g	4.0
0.9% saline	150	0	150	0	308	Isotonic	-	5.0
Ringer's lactate	130	4	109	28 (as lactate)	273	Isotonic	Ca ²⁺ 2 mmol.l ⁻¹	6.5
Hartmann's	131	5	111	29 (as lactate)	255	Isotonic	Ca ²⁺ 2 mmol.l ⁻¹	6.5
Plasma-Lyte 148®	140	5	98	27 (as acetate)	294	Isotonic	Mg ²⁺ 1.5 mmol.l ⁻¹ Gluconate 23mmol.l ⁻¹	4-6.5
4.5% albumin in saline	100-160	<2	150	0	275	Isotonic		7.4
Gelofusine	154	<0.4	120	0	274	Isotonic	Gelatin 40g	7.4
Haemaccel	145	5	145	0	293	Isotonic	Gelatin 35g	7.4

* with respect to plasma.

This child must be treated immediately with hypertonic saline (3% saline) to correct the plasma sodium, not an isotonic fluid (and definitely not a hypotonic fluid). Ideally, the child should be cared for in a PICU and 3% saline administered as follows:⁶

- Give 3% NaCl 2ml.kg⁻¹ over 10 minutes.
- Repeat as necessary 1-2 times.
- Recheck plasma Na⁺ after second bolus or 2 hours.
- Stop therapy when the patient is symptom free (awake, alert, responding to commands, resolution of nausea and headache); a rise of Na⁺ 5-6mmol is achieved; or there is an acute rise in Na⁺ of 10mmol.l⁻¹ in the first 5 hours.
- Do not exceed a correction of Na⁺ more than 15-20mmol.l⁻¹ in 48 hours.

What about dextrose?

Dextrose may be required to prevent hypoglycaemia while the child is starved, although this appears to be less of a problem

than was previously thought.

The diurnal variation in cortisol levels effects blood glucose levels. Cortisol levels are higher in the morning than the afternoon, so children starved overnight have a higher blood glucose than those starved during the day.¹³ The stress response to surgery may result in hyperglycaemia in children as young as two weeks of age, even if no dextrose-containing fluids are given.¹⁴ Although less catastrophic than severe hypoglycaemia, hyperglycaemia also has detrimental effects, and should be avoided. In the ischaemic or hypoxic brain hyperglycaemia may result in accumulation of lactate, cellular acidosis and compromised cellular function. Hyperglycaemia also causes an osmotic diuresis, which may lead to dehydration and electrolyte disturbance. Routine administration of dextrose-containing fluids during surgery should be reserved for those at risk of hypoglycaemia.

Recent studies have shown that hypoglycaemia during surgery is rare in most children. Exceptions to this are premature

infants, neonates less than 48 hours old, neonates in whom a preoperative glucose infusion is interrupted and children below the 3rd centile in weight.¹⁶ Children with an extensive regional block or surgery of greater than 3 hours duration may also be at risk of intraoperative hypoglycaemia and these groups of children should be maintained on dextrose infusions without prolonged interruption.¹⁶

However, if children are given dextrose free solutions postoperatively, they may become hypoglycaemic, or they may metabolise lipids and develop ketosis, particularly if they are less than 6 years of age.¹⁰

The majority of children can therefore be given dextrose free isotonic crystalloid solutions such as Hartmann's/Ringer's Lactate during surgery. After surgery, all children should receive maintenance fluids containing dextrose. Any child perceived to be at risk of hypoglycaemia should have their blood glucose monitored at regular intervals.

A low concentration of dextrose should be used (1%-2.5%) if dextrose is required for a child at risk of hypoglycaemia during surgery; 0.9% saline with 5% dextrose may result in moderate to marked hyperglycaemia. Isotonic crystalloid solutions containing 1% or 2.5% dextrose are commercially available in some countries. As an alternative, solutions can be made up as follows:

- Hartmann's 1% dextrose – add 10ml 50% dextrose to 500ml Hartmann's
- Hartmann's 2.5% dextrose – add 25ml 50% dextrose to 500ml Hartmann's

Choice of isotonic crystalloid during surgery; saline or balanced salt solution?

Saline has a greater chloride content than plasma, also than Hartmann's, Ringer's or Plasma-Lyte, and may cause hyperchloraemic acidosis (see Table 4).

Balanced salt solutions such as Hartmann's or Ringer's are slightly hypotonic and may result in a fall in serum sodium if given in large quantities. The clinical impact of hyperchloraemic acidosis is uncertain, but it is common practice to use a balanced salt solution such as Hartmann's, Ringer's or Plasma-

Lyte during surgery. Always check electrolytes with prolonged use of any IV fluids.

Crystalloid or colloid?

The use of crystalloids or colloids has been controversial for many years. Recently, concerns have been raised about the use of intravenous starch solutions in patients with sepsis and/or at risk for renal failure. There is little evidence in children (and even less evidence to support the use of intravenous gelatins in children). Colloid solutions also cause a greater fall in plasma haemoglobin than an equivalent volume of crystalloid solution, and may increase the requirement for blood transfusion. Colloids are more expensive than crystalloids, gelatins may be associated with increased risk of anaphylaxis, and the long term side-effects of starch solutions in children are not known. A pragmatic approach suggests that a balanced salt solution should be used in preference to colloids during surgery, with blood transfused when required.

Trigger for transfusion?

Most children undergoing surgery are healthy with normal cardiorespiratory function, and excellent tissue oxygen delivery, particularly during infancy when their cardiac output is relatively high, and they tolerate anaemia well. However, newborns have high levels of HbF and require a higher haemoglobin level, as HbF is less efficient at off-loading oxygen to the tissues; similarly, children with cyanotic heart disease have less effective tissue oxygen delivery. It is difficult to be prescriptive about the haemoglobin level that should be the 'trigger' for transfusion, but suggested levels are shown in Table 5. Ideally, the haemoglobin level should be measured regularly during surgery; blood should be transfused to minimise donor exposure, usually in a dose of 10-20 ml.kg⁻¹. A useful formula to predict the haemoglobin rise is as follows:

- Transfusion 8ml.kg⁻¹ whole blood raises the haemoglobin by 1g.dl⁻¹
- Transfusion 4ml.kg⁻¹ packed cells raises the haemoglobin by 1g.dl⁻¹

POSTOPERATIVE MAINTENANCE FLUIDS

The choice of maintenance fluids in the postoperative period

Table 5. Suggested trigger haemoglobin for blood transfusion in children

	Haemoglobin level that should trigger blood transfusion g.l ⁻¹
Healthy child	70
Newborn (untransfused) - high HbF	120
Cyanotic heart disease	100-120
Early severe sepsis	100
Chronic anaemia	To maintain normal baseline haemoglobin

remains controversial; the choice of both sodium content and dextrose needs to be considered. Hypotonic fluids containing 0.18% saline in 4% dextrose must NOT be used at any time in the perioperative period.¹

Children younger than 6 years require a source of dextrose postoperatively to avoid hypoglycaemia and to avoid lipolysis.¹⁰ Hypotonic fluids should not be administered if the plasma sodium is less than 140 mmol.l⁻¹, although measurement of electrolytes may not always be possible. If a hypotonic fluid containing 0.45% saline is given in the immediate postoperative period, the plasma sodium tends to fall due to the effect of raised ADH. If the plasma sodium is already low, it will remain low if 0.45% saline is used. When the plasma electrolytes are not known it is safer to give 0.9% saline to a patient with an elevated plasma sodium, than it is to give hypotonic fluids to a hyponatraemic patient. A pragmatic approach therefore suggests that isotonic solutions containing dextrose, such as 0.9% saline 5% dextrose, or Hartmann's/Ringer's/Plasma-Lyte 5% dextrose should be used for maintenance fluids in the immediate postoperative period.

It is essential that fluid balance and vital signs continue to be monitored in the post-operative period, ideally including urine output, and daily plasma electrolytes and the weight of the child. Abnormal losses such as naso-gastric tube or wound drain losses should be measured and replaced ml for ml with 0.9% saline + 10mmol KCl. KCl should not be added to maintenance fluids until urine output is established (usually day 2 postoperatively). Intravenous fluids should be stopped as soon as possible in the postoperative period; it is much better for the child to control their own fluid balance.

Suggested perioperative fluid regimen:

- During surgery; use Hartmann's/Ringer's/Plasma-Lyte and/or blood as clinically indicated. Give fluid boluses of 10-20ml.kg⁻¹ and assess clinical signs.
- Low dose dextrose may be required for neonates and those at risk of hypoglycaemia; check the blood sugar regularly.
- Postoperative maintenance fluids; give an isotonic fluid with 5% dextrose and calculate the fluid requirement using the Holliday and Segar '4:2:1' rule.
- Give additional fluids to correct deficits, measured or suspected ongoing losses using 0.9% saline, Hartmanns/Ringer's/Plasma-Lyte, colloid or blood as indicated.

THE FEAST STUDY

The FEAST study was a large randomised controlled study carried out in six hospitals in Africa (Kenya, Uganda and Tanzania), published in 2011.¹ Children aged 2 months to 12 years with a diagnosis of a severe febrile illness (impaired consciousness and/or respiratory distress with impaired

perfusion) were randomised to receive a fluid bolus of 20ml.kg⁻¹ 0.9% saline or 20ml.kg⁻¹ 5% albumin when they were admitted to hospital, or they were included in a control group and treated with routine maintenance fluids using the '4,2,1' rule. More than 3000 children were included in the trial, which is the largest study of its kind in fluid resuscitation in children. Children with gastroenteritis, burns or requiring surgery were NOT included in the study.

The results were surprising, and led to the trial being stopped early on safety grounds. Children who received a fluid bolus were 3.3% more likely to die in the first 48 hours after admission than the control group children who received routine maintenance fluids. The children in the study were all severely unwell (76% had impaired consciousness, 83% had respiratory distress); 57% had malaria and 32% had a haemoglobin <5g.dl⁻¹, but the adverse effect of a fluid bolus was still seen in those without malaria and those who did not have severe anaemia. The reason for the excess mortality in those receiving a fluid bolus is not clear; the children appeared to do well initially, but the clinical benefit was not maintained. There were no intensive care facilities in the study hospitals and the terminal event in most cases was cardiogenic shock. It may be that shock is an important adaptive response in this setting, and that administration of a fluid bolus overrides this adaptation; or perhaps there are subtle effects of a fluid bolus related to hyperchloraemic acidosis. More research is required.

The important conclusion from this study is that critically ill children with sepsis in Africa should NOT receive rapid fluid resuscitation with saline or albumin, but should receive IV fluids at normal maintenance rates whilst definitive treatment for sepsis is started (e.g. antibiotics or antimalarials). The authors state that children with acute gastroenteritis or burns or surgical conditions may still require resuscitation fluids, and the recommendations do not apply to these patients. The relevance of the FEAST study to children in high-income countries is still uncertain, but current treatment protocols must re-evaluated in the light of this important study.

CONCLUSIONS

Children should not be starved for prolonged periods before surgery, and oral fluids should be given wherever possible. Intravenous fluids should be prescribed carefully, as for any drug:

- The majority of healthy children undergoing minor surgery will re-establish oral intake in the early postoperative phase and will not need routine intravenous fluids.
- Hypovolaemia should be corrected by rapid infusion of isotonic fluid while dehydration is corrected more slowly over 14-72 hours as appropriate. Ongoing losses should be measured and replaced.

- During surgery the majority of children over 1 month of age will maintain a normal blood glucose if isotonic, non-dextrose containing fluids are given.
 - Hypotonic fluids should be used with care in the perioperative period, and must not be infused in large volumes or at greater than maintenance rates. 0.18% saline 4% dextrose must NOT be used.
 - Ideally, plasma electrolytes, glucose and haemoglobin (or haematocrit) should be measured regularly in any child receiving large volumes of IV fluid, or who remains on IV fluids for more than 24 hours.
 - Critically ill children with sepsis should receive IV fluids at normal maintenance rates whilst definitive treatment is started. They must not receive IV fluid boluses for resuscitation.
6. Moritz et al. New aspects in the pathogenesis, prevention and treatment of hyponatraemic encephalopathy in children. *Pediatr Nephrology* 2010; **25**: 1225-38.
 7. Arieff AI, Ayus JC, Fraser CL. Hyponatraemia and death or permanent brain damage in healthy children. *British Medical Journal* 1992; **304**: 1218-22.
 8. Halberthal M, Halperin ML, Bohn D. Acute hyponatraemia in children admitted to hospital: retrospective analysis of factors contributing to its development and resolution. *British Medical Journal* 2001; **322**: 780-82.
 9. Bailey et al. Perioperative crystalloid and colloid fluid management in children: where are we now and how did we get here? *Anesth Analg* 2010; **110**: 375-90.
 10. Neville et al. Prevention of hyponatraemia during maintenance intravenous administration; a prospective randomised study of fluid type versus fluid rate. *J Pediatr* 2010; **156**: 313-9.

REFERENCES

- Wilson CM. Perioperative fluids in children. Update in Anaesthesia 2005; 19: 36-8. Available from: http://www.wfsahq.org/components/com_virtual_library/media/07417f70e0b243fc795d737bbb8ebd18-57883b2855874aa26a469a64e36d77af-Perioperative-Fluids-in-Children--Update-19-2005-.pdf
1. Maitland K, Kiguli S, Opoka RO et al. Mortality after fluid bolus in African Children with severe infection. *NEJM* 2011; **364**: 2483-95.
 2. NPSA (2007) Reducing the risk of hyponatraemia when administering intravenous infusions to children. Alert no 22. National Patient safety Agency London <http://www.nhs.npsa.nhs.uk/resources/type/alerts>
 3. Assadi F, Copelovitch L. Simplified treatment strategies to fluid therapy in diarrhea. *Pediatric Nephrology* 2003; **18**: 1152-6.
 4. Holliday MA, Segar WE. The maintenance need for water in parenteral fluid therapy. *Paediatrics* 1957; **19**: 823-832.
 5. Oh TH. Formulas for calculating fluid maintenance requirements. *Anesthesiology* 1980; **53**: 351.
 11. O'Brian F, Walker IA. Fluid homeostasis in the neonate. *Paediatric Anaesthesia* 2014; **24**: 49-59.
 12. Judd BA, Haycock GB, Dalton RN, Chantler C. Antidiuretic hormone following surgery in children. *Acta Paediatrica Scandinavia* 1990; **79**: 461-6.
 13. Redfern N, Addison GM, Meakin G. Blood glucose in anaesthetised children. Comparison of blood glucose concentrations in children fasted for morning and afternoon surgery. *Anaesthesia* 1986; **41**: 272-5.
 14. Nilsson K, Larsson LE, Andreasson S, Ekstrom-Jodal B. Blood-glucose concentrations during anaesthesia in children. Effects of starvation and perioperative fluid therapy. *British Journal of Anaesthesia* 1984; **56**: 375-9.
 15. Larsson LE, Nilsson K, Niklasson A, Andreasson B, Ekstrom-Jodal B. Influence of fluid regimens on perioperative blood glucose concentrations in neonates. *British Journal of Anaesthesia* 1990; **64**: 419-24.
 16. Leelanukorum R, Cunliffe M. Intraoperative fluid and glucose management in children. *Paediatric Anaesthesia* 2000; **10**: 353-9.

Update in Anaesthesia

Paediatric caudal anaesthesia

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INDICATIONS FOR CAUDAL ANAESTHESIA

The indications for single shot CA are abdominal, urologic or orthopaedic surgical procedures located in the sub-umbilical abdominal, pelvic and genital areas, or the lower limbs, where postoperative pain does not require prolonged strong analgesia. Examples of appropriate surgery include inguinal or umbilical herniorrhaphy, orchidopexy, hypospadias and club foot surgery. CA is useful for day case surgery, but opioid additives to the local anesthetic agent should be avoided in this setting. When CA is used, requirement for mild or intermediate systemic analgesia must be anticipated to prevent pain resurgence at the end of caudal block. Catheter insertion can extend the indications to include surgical procedures located in the high abdominal or thoracic areas, and to those requiring prolonged effective analgesia.

CONTRAINDICATIONS

The usual contraindications to regional anaesthesia such as coagulation disorders, local or general infection, progressive neurological disorders and patient or parental refusal apply to CA. Furthermore, cutaneous anomalies (angioma, hair tuft, naevus or a dimple) near the puncture point require radiological examination (ultrasound, CT or MRI), in order to rule out underlying spinal cord malformation such as a tethered cord.²³ A Mongolian spot is not a contraindication to CA.

ANATOMY

Anatomical landmarks (Figure 1)

The sacrum is roughly the shape of an equilateral triangle, with its base identified by feeling the two posterosuperior iliac processes and a caudal summit corresponding to the sacral hiatus. The sacrum is concave anteriorly. The dorsal aspect of the sacrum consists of a median crest, corresponding to the fusion of sacral spinous processes. Moving laterally, intermediate and lateral crests correspond respectively to the fusion of articular and transverse processes.

The sacral hiatus is located at the caudal end of the median crest and is created by failure of the S5 laminae to fuse (Figure 1). The hiatus is surrounded by the sacral cornua, which represent remnants of the inferior

S5 articular processes and which face the coccygeal cornua. Palpation of the sacral cornua is fundamental to locating the sacral hiatus and to successful caudal block.

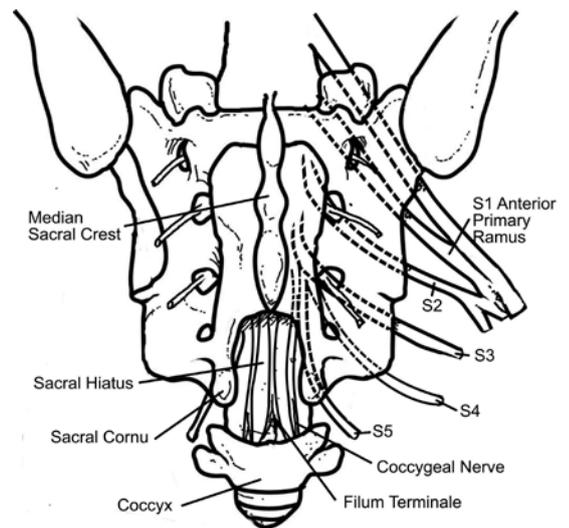


Figure 1. The posterior aspect of the sacrum and sacral hiatus

The sacral hiatus is the shape of an inverted U, and is covered by the sacro-coccygeal ligament, which is in continuity with the ligamentum flavum. It is large and easy to locate until 7-8 years of age. Later, progressive ossification of the sacrum (until 30 years old) and closing of the sacro-coccygeal angle make its identification more difficult. Note that anatomical anomalies of the sacral canal roof are observed in 5% of patients and this can lead to unplanned cranial or lateral puncture.

The sacral canal

The sacral canal is in continuity with the lumbar epidural space. It contains the nerve roots of the cauda equina, which leave it through anterior sacral foraminae. During CA, leakage of local anaesthetic agent (LA) through these foraminae explains the high quality of analgesia, attributable to diffusion of LA along the nerve roots. Spread of analgesia cannot be enhanced above T8-T9 by increasing injected LA volume.

Summary

Caudal anaesthesia (CA) is epidural anaesthesia of the cauda equina roots in the sacral canal, accessed through the sacral hiatus. CA is a common paediatric regional technique that is quick to learn and easy to perform, with high success and low complication rates.

CA provides high quality intraoperative and early postoperative analgesia for sub-umbilical surgery. In children, CA is most effectively used as adjunct to general anaesthesia and has an opioid-sparing effect, permitting faster and smoother emergence from anaesthesia.

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The dural sac (i.e. the subarachnoid space) ends at the level of S3 in infants and at S2 in adults and children. It is possible to puncture the dural sac accidentally during CA, leading to extensive spinal anaesthesia. Therefore the needle or cannula must be cautiously advanced into the sacral canal, after crossing the sacro-coccygeal ligament. The distance between the sacral hiatus and the dural sac is approximately 10mm in neonates. It increases progressively with age (>30mm at 18 years), but there is significant inter-individual variability in children.¹ The contents of sacral canal are similar to those of lumbar epidural space, predominantly fat and epidural veins. In children, epidural fatty tissue is looser and more fluid than in adults, favoring LA diffusion.

TECHNIQUE

Preparation

Obtain consent for the procedure either from the patient or, if appropriate, from the parents. After induction of general anaesthesia and airway control, the patient is positioned laterally (or ventrally), with their hips flexed to 90° (Figure 2). Skin disinfection should be performed carefully, because of the proximity to the anus. Aseptic technique should be maintained.



Figure 2. Preparation of patient - lateral position with the surgical site down

According to the child's size, needle diameter and length are respectively between 21G and 25G, and 25mm and 40mm. A short bevel improves the feeling of sacrococcygeal ligament penetration and decreases risk of vascular puncture or sacral perforation.² Use of a needle with a stylet avoids risk of cutaneous tissue coring, and the (theoretical) risk of epidural cutaneous cell graft. If a stylet needle is not available, a cutaneous 'pre-hole' can be made with a different needle prior to puncture with the caudal needle. Another solution is to puncture with an IV catheter, the hollow needle of which is removed before injection through the sheath.

Puncture (Figures 3, 4 and 5)

After defining the bony landmarks of the sacral triangle, the two sacral cornuae are identified by moving your fingertips from side to side. The gluteal cleft is not a reliable mark of the midline. The puncture is performed between the two sacral cornuae. The needle is oriented 60° in relation to back plane, 90° to skin surface. The needle bevel is oriented ventrally, or parallel to the fibers of the sacro-coccygeal ligament. The distance between the skin and sacro-coccygeal ligament

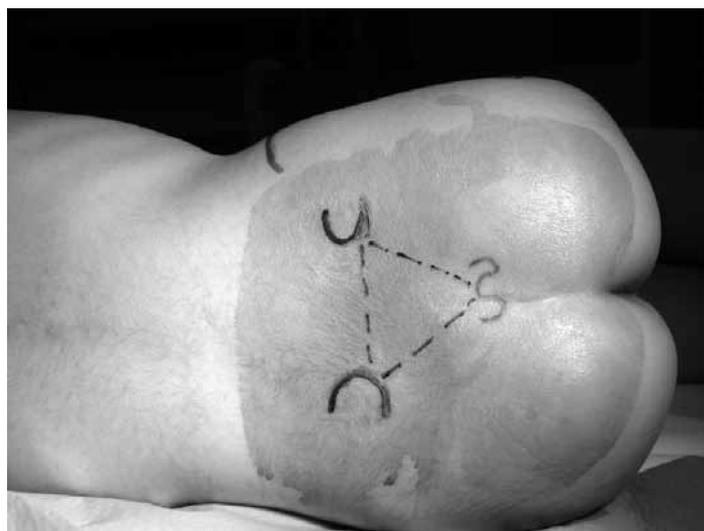


Figure 3. Bony landmarks

is between 5 and 15mm, depending on the child's size. The sacro-coccygeal ligament gives a perceptible 'pop' when crossed, analogous to the ligamentum flavum during lumbar epidural anaesthesia. After crossing the sacro-coccygeal ligament, the needle is redirected 30° to the skin surface, and then advanced a few millimeters into sacral canal. If in contact with the bony ventral wall of sacral canal, the needle must be moved back slightly.

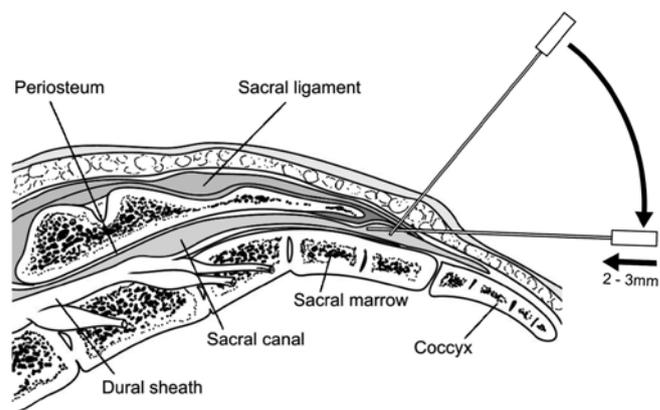


Figure 4. Puncture - orientation of the needle and reorientation after crossing the sacro-coccygeal ligament



Figure 5. Orientation of the needle during puncture

After verifying absence of spontaneous reflux of blood or cerebrospinal fluid (more sensitive than an aspiration test), injection of LA should be possible without resistance. Inject slowly (over about one minute). Where available this may be preceded with an epinephrine

test dose under ECG and blood pressure monitoring, in order to detect intravascular placement. Subcutaneous bulging at the injection site suggests needle misplacement. Blood reflux necessitates repeating the puncture, however in case of cerebrospinal fluid reflux caudal anaesthesia should be abandoned, in order to avoid the risk of extensive spinal anaesthesia. Aspiration tests should be repeated several times during injection.

In skilled hands, the success rate of CA is about 95%, however a variety of misplacements of the needle are possible (Figure 6). The moment of surgical incision is the true test of block success, but various techniques have been suggested to authenticate the puncture success, such as injection site auscultation (the 'swoosh test'), or searching for anal sphincter contraction in response to electrical nerve stimulation on the puncture needle. No clear benefit of these techniques against simple clinical assessment have been shown.^{3,4} More recently, ultrasound has been suggested to help sacro-coccygeal hiatus location and to visualize isotonic serum or LA injection into sacral epidural space (Figures 7 and 8).^{5,6} These authors have also outlined the interest in ultrasound control within the context of learning the technique, rather than for use in standard practice.^{5,7}

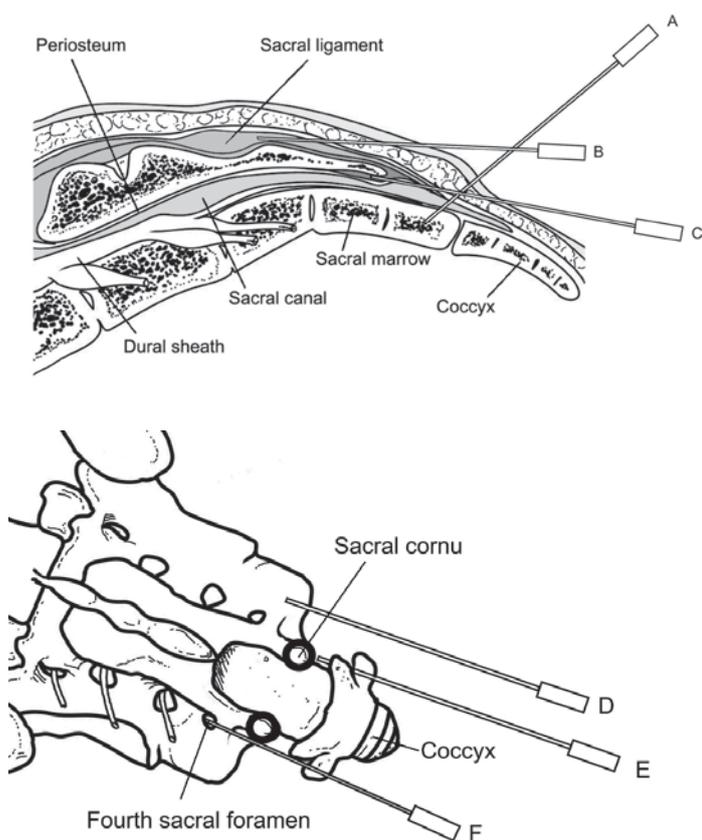


Figure 6A and B. Needle misplacement

- A marrow (resistance +++). Equivalent to IV injection)
- B posterior sacral ligament (subcutaneous bulge)
- C subperiosteal
- D "decoy" hiatus
- E intrapelvic (risk of damaging intrapelvic structures: rectum)
- F 4th sacral foramen (unilateral block).

Catheter insertion

Although CA was initially described as a single shot technique, some authors have described use of a caudal catheter to prolong analgesic administration in postoperative period. In addition advancement of the catheter in the epidural space up to lumbar or even thoracic levels can achieve analgesia of high abdominal or thoracic areas.⁸ However, two pitfalls restrict extension of this technique; a high risk of catheter bacterial colonization, particularly in infants and a high risk of catheter misplacement.^{9,10} Subcutaneous tunnelling at a distance from the anal orifice, or occlusive dressings decrease bacterial colonization.¹¹ Electrical nerve stimulation or ECG recording on the catheter, or its echographic visualization have been suggested to guide its advancement in epidural space.^{12,13} However, most anaesthetists presently prefer a direct epidural approach at the desired level that is appropriate to the surgical intervention.^{14,15}

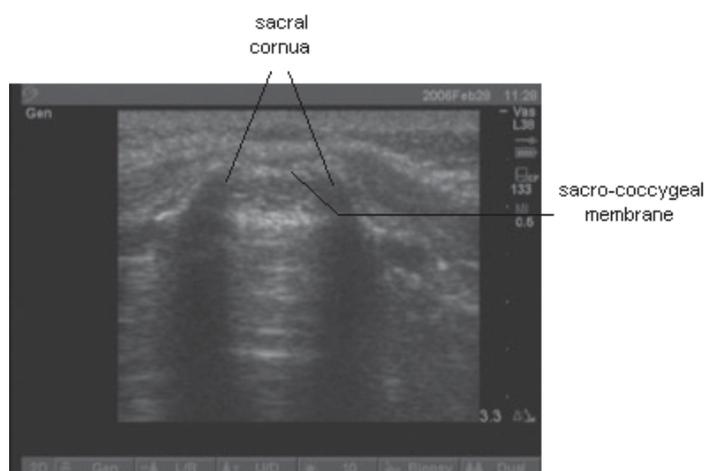
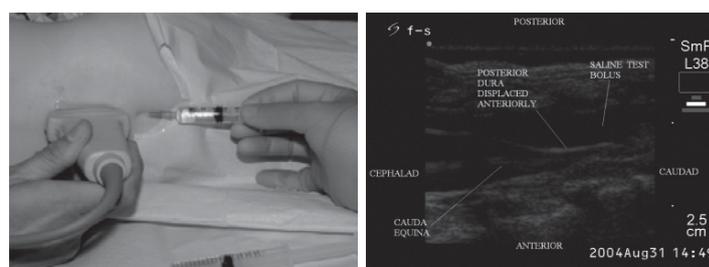


Figure 7.



Figures 8a and 8b.

LOCAL ANAESTHETIC AGENTS

Test dose

Early neurosensory warning symptoms of LA systemic toxicity are concealed by general anaesthesia. Halogenated anaesthetic agents worsen LA systemic toxicity and can also blunt the cardiovascular signs of an intravenous epinephrine test dose injection. Aspiration tests to elicit blood reflux are not very sensitive, particularly in infants. A test dose of epinephrine 0.5mcg.kg^{-1} (administered as 0.1ml.kg^{-1} lidocaine with epinephrine 1 in 200 000) allows detection of intravenous injection with sensitivity and specificity close to 100%, under halogenated anaesthesia. Warning symptoms are cardiac frequency modification (an increase or decrease by 10 beats per minute), increased in blood pressure (up to 15mmHg), or T-wave amplitude change in



Figure 9. T-wave amplitude change after intravascular injection of a local anaesthetic agent

the 60 to 90 second period after injection (Figure 9).^{16,17} Slow injection of the whole LA dose under haemodynamic and ECG monitoring remains essential for patient safety.

Full dose

The volume of caudally injected LA determines the spread of the block and this must be adapted to surgical procedure (Table 1). Analgesic spread will be two dermatomes higher on the down positioned side at the time of puncture. Injected volume must not exceed 1.25 ml.kg⁻¹ or 20 to 25ml, in order to avoid excessive cerebrospinal fluid pressure.

Table 1. Spread of block as a function of caudally injected local anaesthetic volume¹⁸

Volume (ml.kg ⁻¹)	Dermatome level	Indication
0.5	Sacral	Circumcision
0.75	Inguinal	Inguinal herniotomy
1	Lower thoracic (T10)	Umbilical herniorraphy, orchidopexy
1.25	Mid thoracic	

LA choice prioritizes long lasting effects with the weakest motor block possible, since motor block is poorly tolerated in awake children. Bupivacaine meets these criteria. More recently available, ropivacaine and L-bupivacaine have less cardiac toxicity than bupivacaine at equivalent analgesic effectiveness. They may also confer a more favorable differential block (less motor block for the same analgesic power) and the 2.5mg.ml⁻¹ (0.25%) concentration is optimal for these agents. Four to six hours analgesia is usually achieved with minimal motor block.^{19,20}

Maximal doses must not be exceeded (Table 2) but use of a more dilute mixture may allow the desired volume to be achieved within the recommended maximum dose. Hemodynamic effects of CA are weak or absent in children, so intravenous fluid preloading or vasoconstrictive drugs are unnecessary.

Table 2. Maximal allowable doses of local anaesthetic agents

	Plain local anaesthetic (mg.kg ⁻¹)	With epinephrine (mg.kg ⁻¹)	Neonates
Bupivacaine	2	2	
Lidocaine	3	7	↓ 20%
Ropivacaine	3	3	

Additives

Several drugs have been demonstrated to prolong duration of analgesia by a few hours after single shot caudal injection of LA. Among them, most popular are opioids (fentanyl 1 µg.kg⁻¹), clonidine (1-2 µg.kg⁻¹) and preservative free ketamine (not the IV form) (0.5 mg.kg⁻¹).²¹ Recent reports of spinal cord toxicity of intrathecal ketamine in neonatal rats leads us to discourage its use by caudal route in neonates and infants.²² Morphine and clonidine do not provoke such spinal cord toxicity in neonatal rats, but their dose requirements are decreased in younger pups.²³ Principal adverse effects are: pruritus and nausea and vomiting for opioids, light sedation for clonidine, and hallucinations for ketamine. Theoretical risk of respiratory depression with opioids mandates adequate postoperative monitoring. Some cases of respiratory depression have been reported with caudal clonidine in neonates.²⁴

COMPLICATIONS

Complications of CA are uncommon (0.7 per 1000 cases), are more likely if inadequate equipment is used and are more frequent in infants.¹⁶ If the technique fails it should be abandoned to avoid occurrence of potentially serious complications.

Significant complications, in order of decreasing frequency, are:

- **Dural tap.** This is more likely if the needle is advanced excessively in the sacral canal when subarachnoid injection of local anaesthetic agent may cause extensive spinal anaesthesia. Under general anaesthesia this should be suspected if non-reactive mydriasis (pupillary dilation) is observed.
- **Vascular or bone puncture** can lead to intravascular injection and consequently LA systemic toxicity. Preventative measures are use of a test dose, cessation of injection if resistance is felt and slow injection under hemodynamic and ECG monitoring. Sacral perforation can lead to pelvic organ damage (e.g. rectal puncture).
- **Exceeding the maximal allowed LA dose** risks overdose and related cardiovascular or neurological complications.
- **Delayed respiratory depression** secondary to caudally injected opioid.
- **Urinary retention** - spontaneous micturition must be observed before hospital discharge.
- **Sacral osteomyelitis** is rare (one case report).²⁵

CONCLUSION

This technique has an established role in paediatric regional anaesthesia practice since it is easy to learn and has a favorable risk/benefit ratio. Despite being more complex to learn, alternative peripheral regional anaesthesia techniques are gaining popularity and may begin to replace caudal anaesthesia as a popular choice.

REFERENCES

- Raux O, Dadure C, Carr J, Rochette A, Capdevila X. Paediatric caudal anaesthesia. Update in Anaesthesia (2010) 26:32-36. Available from: <http://www.wfsahq.org/documents/Update%20in%20Anaesthesia/update%20026%202010.pdf>
1. Adewale L, Dearlove O, Wilson B, Hindle K, Robinson DN. The caudal canal in children: a study using magnetic resonance imaging. *Paediatr Anaesth* 2000; **10**: 137-41.
 2. Dalens B, Hasnaoui A. Caudal anesthesia in pediatric surgery: success rate and adverse effects in 750 consecutive patients. *Anesth Analg* 1989; **68**: 83-9.
 3. Orme RM, Berg SJ. The "swoosh" test - an evaluation of a modified "whoosh" test in children. *Br J Anaesth* 2003; **90**: 62-5.
 4. Tsui BC, Tarkkila P, Gupta S, Kearney R. Confirmation of caudal needle placement using nerve stimulation. *Anesthesiology* 1999; **91**: 374-8.
 5. Raghunathan K, Schwartz D, Conelly NR. Determining the accuracy of caudal needle placement in children: a comparison of the swoosh test and ultrasonography. *Paediatr Anaesth* 2008; **18**: 606-12.
 6. Roberts SA, Guruswamy V, Galvez I. Caudal injectate can be reliably imaged using portable ultrasound - a preliminary result. *Paediatr anaesth* 2005; **15**: 948-52.
 7. Schwartz DA, Dunn SM, Conelly NR. Ultrasound and caudal blocks in children. *Paediatr Anaesth* 2006; **16**: 892-902 (correspondence).
 8. Tsui BC, Berde CB. Caudal analgesia and anesthesia techniques in children. *Curr Op Anesthesiol* 2005; **18**: 283-8.
 9. Kost-Byerly S, Tobin JR, Greenberg RS, Billett C, Zahurak M, Yaster M. Bacterial colonisation and infectious rate of continuous epidural catheters in children. *Anesth Analg* 1998; **86**: 712-6.
 10. Valairucha S, Seefelder D, Houck CS. Thoracic epidural catheters placed by the caudal route in infants: the importance of radiographic confirmation. *Paediatr Anaesth* 2002; **12**: 424-8.
 11. Bubeck J, Boss K, Krause H, Thies KC. Subcutaneous tunneling of caudal catheters reduces the rate of bacterial colonization to that of lumbar epidural catheters. *Anesth Analg* 2004; **99**: 689-93.
 12. Tsui BC, Wagner A, Cave D, Kearny R. Thoracic and lumbar epidural analgesia via the caudal approach using electrical stimulation guidance in pediatric patients: a review of 289 patients. *Anesthesiology* 2004; **100**: 683-9.
 13. Chawathe MS, Jones RM, Gildersleve CD, Harrison SK, Morris SJ, Eickmann C. Detection of epidural catheters with ultrasound in children. *Paediatr Anaesth* 2003; **13**: 681-4.
 14. Bösenberg AT. Epidural analgesia for major neonatal surgery. *Paediatr Anaesth* 1998; **8**: 479-83.
 15. Giauffré E, Dalens B, Gombert A. Epidemiology and morbidity of regional anesthesia in children: a one-year prospective survey of the French-language society of pediatric anesthesiologists. *Anesth Analg* 1996; **83**: 904-12.
 16. Kozek-Langenecker SA, Marhofer P, Jonas K, Macik T, Urak G, Semsroth M. Cardiovascular criteria for epidural test dosing in sevoflurane- and halothane-anesthetized children. *Anesth Analg* 2000; **90**: 579-83.
 17. Tobias JD. Caudal epidural block: a review of test dosing and recognition injection in children. *Anesth Analg* 2001; **93**: 1156-61.
 18. Armitage EN. Local anaesthetic techniques for prevention of postoperative pain. *Br J Anaesth* 1986; **58**: 790-800.
 19. Bösenberg AT, Thomas J, Lopez T, Huledal G, Jeppsson L, Larsson LE. Plasma concentrations of ropivacaine following a single-shot caudal block of 1, 2 or 3 mg/kg in children. *Acta Anaesthesiol Scand* 2001; **10**: 1276-80.
 20. Breschan C, Jost R, Krumpholz R, Schaumberger F, Stettner H, Marhofer P, Likar R. A prospective study comparing the analgesic efficacy of levobupivacaine, ropivacaine and bupivacaine in pediatric patients undergoing caudal blockade. *Paediatr Anaesth* 2005; **15**: 301-6.
 21. Menzies R, Congreve K, Herodes V, Berg S, Mason DG. A survey of pediatric caudal extradural anesthesia practice. *Paediatr Anaesth* 2009; **19**: 829-36.
 22. Walker SM, Westin BD, Deumens R, Grafe M, Yaksh TL. Effects of intrathecal ketamine in the neonatal rat. Evaluation of apoptosis and long-term functional outcome. *Anesthesiology* 2010; **113**: 147-59.
 23. Walker SM, Yaksh TL. Neuraxial analgesia in neonates and infants: a review of clinical and preclinical strategies for the development of safety and efficacy data. *Anesth Analg* 2012; **115**: 638-62.
 24. Fellmann C, Gerber AC, Weiss M. Apnoea in a former preterm infant after caudal bupivacaine with clonidine for inguinal herniorrhaphy. *Paediatr Anaesth* 2002; **12**: 637-40.
 25. Cohen IT. Caudal block complication in a patient with trisomy 13. *Paediatr Anaesth* 2006; **16**: 213-5.

Abdominal wall blocks

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INTRODUCTION

Regional anaesthesia is an essential component of paediatric anaesthetic practice. Regional blocks allow for a lighter plane of anaesthesia during surgery, and provide excellent pain control after surgery.¹ The aim of this review is to describe how to perform the three most common abdominal wall blocks in children: ilioinguinal/iliohypogastric, rectus sheath and transversus abdominis plane.

We will describe landmark techniques as well as ultrasound-guided techniques. Ultrasound guided blocks are increasingly considered the gold standard as it is possible to identify the anatomy more accurately, which increases the reliability of the block and allows a smaller dose of local anaesthetic to be used.

Regional anaesthetic blocks are simple to do, but should be taught by an appropriately skilled mentor. All local anaesthetic blocks should be performed using an aseptic technique; clean the skin with an alcohol-based cleaning solution and wear gloves. Ideally use a short-bevelled block needle for abdominal wall blocks, but a 23G or 21G hypodermic needle may also be used; many advocate 'blunting' the tip of the needle on the inside of the cap of the needle to better appreciate the facial planes. All the blocks described should be performed after induction of general anaesthesia.

ILIOINGUINAL/ILIOHYPOGASTRIC NERVE BLOCK (ILNB)

The ilioinguinal/iliohypogastric nerve block (ILNB) provides excellent analgesia after inguinal hernia repair, hydrocele repair and orchidopexy. It does not abolish visceral pain due to peritoneal traction or manipulation of the spermatic cord during inguinal hernia repair or orchidopexy.

Bilateral blocks can be used, but it is important to keep the dose of local anaesthetic within safe limits.

Perform ILNBs after induction of anaesthesia, before the start of surgery; it is important to make sure that the child is adequately anaesthetised when the cord structures are mobilised, and that additional local infiltration/analgesia is used if a scrotal incision is made.

There is much anatomical variation of nerve position between the abdominal wall muscles. The effectiveness of this block can be improved greatly when performed with ultrasound, and lower amounts of local anaesthetic can be used.²

Anatomy (see Figure 1):

- The iliohypogastric (T12, L1) and ilioinguinal (L1) nerves are terminal branches of the lumbar plexus. They lie deep to the internal oblique.
- The iliohypogastric nerve supplies the gluteal region and the skin over the pubic symphysis.
- The ilioinguinal nerve supplies the area of the skin beneath that supplied by the iliohypogastric nerve and the anterior scrotum.
- The nerves emerge at the lateral border of psoas major and pass anterior to quadratus lumborum. They pierce the lumbar fascia at the lateral border of quadratus lumborum and run in the plane between the internal oblique muscle and transversus abdominis muscles.
- The iliohypogastric nerve pierces (again) the internal oblique and runs under the external oblique superior to the inguinal canal

SUMMARY

Regional anaesthesia is an essential component of paediatric anaesthetic practice. This review describes how to perform the three most common abdominal wall blocks in children: ilioinguinal/iliohypogastric, rectus sheath and transversus abdominis plane, using either landmark or [if available] ultrasound-guided techniques.

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- The ilioinguinal nerve continues in the inguinal canal.
- In infants the average nerve-peritoneum distance is only 3.3mm³.
- The fascial plane between the transversus abdominis muscle and the transversalis fascia is in continuity with the space around the femoral nerve.

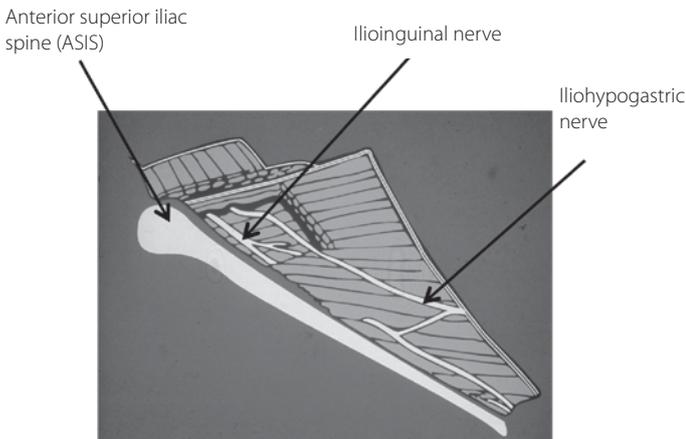


Figure 1. Anatomy of the ilioinguinal/iliohypogastric nerve block

Dose

Use a volume of up to 0.5ml.kg⁻¹ 0.25% bupivacaine for the landmark technique. In expert hands as little as 0.075 ml.kg⁻¹ 0.25% bupivacaine can be effective using ultrasound-guidance; ⁴ we recommend 0.1-0.2ml.kg⁻¹.

Complications

The most common complication is block failure (more common using the landmark technique). Transient femoral nerve palsy with transient quadriceps paresis may be seen if the injection is too deep. Visceral perforation (colon puncture, small bowel puncture, pelvic retroperitoneal haematoma, bowel haematoma) is associated with poor technique, particularly an injection that is too medial.

Techniques

Landmark technique

Place the patient supine. Clean the skin over the lower quadrant of the abdominal wall, including the skin over the anterior superior iliac spine (ASIS). Draw up the appropriate dose of local anaesthetic.

The needle insertion point is close to the ASIS, approximately 2 - 5mm medial to the ASIS on a line drawn between the ASIS and the umbilicus.⁵ Some suggest using the child's finger as an appropriate guide for the distance from the ASIS to the injection point (NOT the operator's finger! - see Figure 2). It is important to keep the injection point high, away from the skin crease in the groin where the surgeon will make the incision; otherwise the operating field will be obscured.



Figure 2. The injection point for the ILNB should be equal to the child's finger breadth medial to the ASIS, not the operator's finger..

Insert the needle just through the skin into the subcutaneous tissues; advance the needle slowly until a fascial 'click' or loss of resistance is felt. The click is felt as the aponeurosis of the external oblique is pierced. Aspirate and then inject the local anaesthetic in this position; there is no need to 'fan' the injection, and this may increase the incidence of complications.

Ultrasound guided technique

Position the patient supine and clean the skin. Place a high frequency linear probe on the anterior abdominal wall along the line joining the anterior superior iliac spine (ASIS) and the umbilicus (a small footprint probe is useful for infants). (See Figure 3).



Figure 3. Ultrasound probe position for ilioinguinal/iliohypogastric nerve block

The ASIS is the most easily recognizable landmark for this block, it appears as a dark echo-lucent shadow beneath a hyperechoic peak and should be kept at the lateral part of the screen for orientation. Identify (always from the inside out) the peritoneum (hyperechoic line, underneath it you may see peristalsis), transversus abdominis muscle, and internal oblique muscle. The external oblique muscle may not be visible as a distinct muscle layer at this level as it may have become an aponeurosis.

Slide the probe up over the iliac crest, whilst maintaining the same orientation of the probe, to bring all three muscles into view as three distinct layers. This may be useful if there is any doubt about the anatomy and the relevant planes.

The ilioinguinal and iliohypogastric nerves are seen in close proximity to each another as two small round hypoechoic structures with a hyperechoic border. They lie in the plane between the internal oblique muscle and the transversus abdominis muscle close to the ASIS. In children the average distance from the ilioinguinal nerve to the ASIS is 7mm.⁴

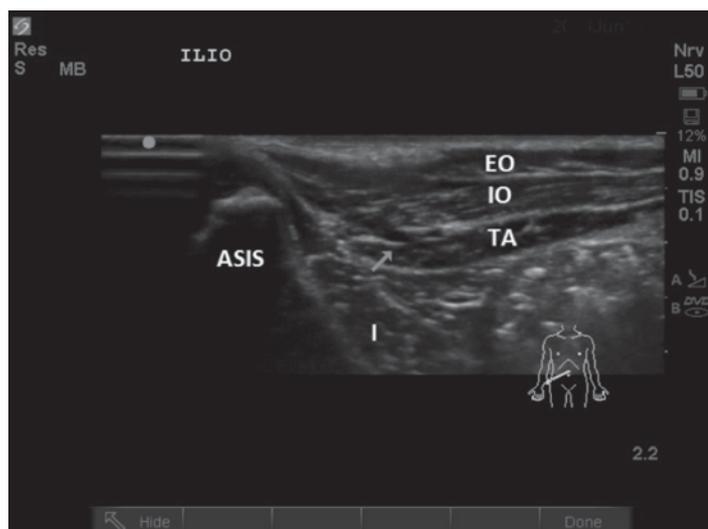


Figure 4. Ultrasound landmarks for iliinguinal/iliohypogastric nerve block. ASIS=anterior superior iliac spine; EO=external oblique muscle; IO=internal oblique muscle; TA=transversus abdominus muscle; I=ilium. Arrow shows location of the nerves.

Insert the block needle in plane from medial to lateral and ensure that the needle tip is visible at all times as it is advanced. Deposit local anaesthetic around the nerves in the transversus abdominis plane. Note that it may not always be easy to visualise the nerves, in this cause you could place the local anaesthetic in the transversus abdominis plane. If you have any concerns regarding the proximity of your injection to the nerve use a higher volume of local anaesthetic (e.g. 0.2ml.kg⁻¹ 0.25% bupivacaine).

RECTUS SHEATH NERVE BLOCK

A rectus sheath block provides intraoperative and

postoperative analgesia for mid-line abdominal incisions, for instance, periumbilical surgery (e.g. umbilical hernia repair,⁶ paraumbilical hernia repair, epigastric hernia repair), pyloromyotomy, laparoscopic surgery and excision of urachal remnants.

Anatomy

- The rectus sheath encloses the rectus abdominis muscle and is formed by the aponeuroses of the three flat abdominal muscles. These aponeuroses join in the lateral border of the rectus muscle in the point called linea semilunaris.
- Medial to the semilunaris, the aponeuroses split with some fibres passing anterior to the rectus muscle and some posterior:
 - The external oblique aponeurosis and the anterior layer of the internal oblique aponeurosis form the anterior wall of the rectus sheath.
 - The transversus abdominis aponeurosis and the posterior layer of the internal oblique aponeurosis form the posterior wall of the sheath.
 - In the midline the aponeuroses from both sides join to form the linea alba.
- The anterior cutaneous branch of the ventral rami of the inferior six thoracic nerves (T7-T12) run anteriorly through the posterior of the rectus muscle to give off sensory branches to the paraumbilical skin.
- The anterior layer of the rectus sheath is firmly attached to the rectus abdominis muscle at three tendinous intersections (at the level of the xiphoid process, the umbilicus and half-way in between). These tendinous intersections are what separate the muscle into the well-known '6-pack', but it is actually an 8-pack.
- The rectus sheath is loosely attached posteriorly, forming a potential space. Local anaesthetic can spread caudad and cephalad in the plane between the rectus muscle and the posterior rectus sheath.
- There is little correlation between the depth of the posterior rectus sheath with age, weight, height or surface area.
- This block is not advisable in neonates without ultrasound guidance as the muscle is so thin and the viscera (especially liver and spleen) are in close proximity.

Dose

Using a landmark technique, use a volume of 0.4ml.kg⁻¹ of 0.25% bupivacaine per side. Using an ultrasound-guided technique a dose of 0.1-0.2ml.kg⁻¹ of 0.25% bupivacaine per side is sufficient.

Complications

Intraperitoneal injection, visceral damage, vascular puncture (it is possible to identify the inferior epigastric vessel in larger children with Doppler).

Techniques

Landmark technique

Use an aseptic technique and draw up the appropriate doses of local anaesthetic. The injection point is just above the umbilicus at the apex of the bulge of the rectus muscle, at 11 o'clock and 1 o'clock to the umbilicus (thinking of the umbilicus as the centre of a clock) (See Figure 5).



Figure 5. Injection point for rectus sheath block for repair of umbilical hernia



Figure 6. Ultrasound probe position for rectus sheath block

Introduce a short-bevelled needle perpendicularly through the skin. Advance the needle medially at an angle of 60° towards the umbilicus. Identify the anterior sheath by moving the needle back and forth until a scratching sensation is felt; a pop is felt as the needle passes through the anterior sheath. Advance the needle through the muscle with continued movement of the needle until a scratching is again felt (this indicates the

posterior sheath). If there is resistance to injection, it is not sited correctly. Repeat the technique for the opposite side.

In children under 10 years of age the rectus muscle is rarely greater than 1cm in thick, therefore when performing this technique the needle should not be inserted any further than this. The depth of the posterior rectus sheath in children is unpredictable, and many advocate using ultrasound for this reason.⁷

Ultrasound technique

Position the patient supine. Select the screen depth (in neonates this will usually be 2cm, infants 3cm, thereafter 4cm). A high frequency linear probe is placed transverse on the abdomen, midline above the umbilicus (see Figure 6).

The initial image will have the linea alba in the midline, with a rectus abdominis muscle either side. Posterior to the rectus muscle there are two hyperechoic lines (“train track”): the more superficial one is the posterior part of the rectus sheath and the deeper one is the peritoneum. Use the Doppler to identify the epigastric vessels, although this is not easy in small children (See Figure 7).

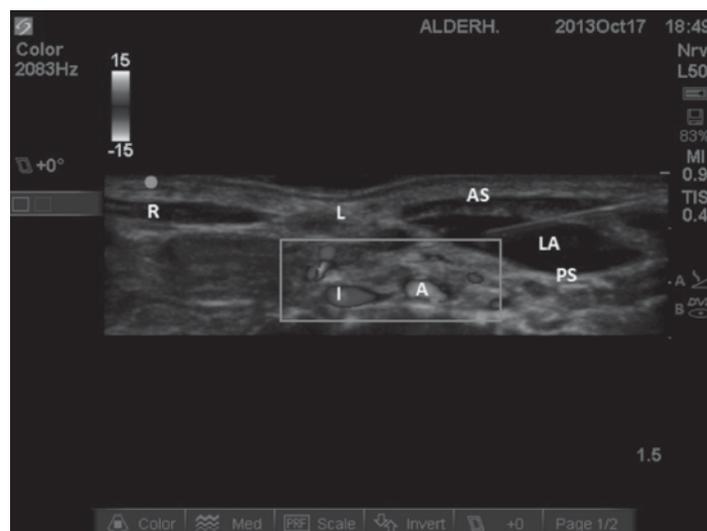


Figure 7. Ultrasound landmarks for rectus sheath block: L=linea alba; R=rectus abdominis muscle; LA=local anaesthetic; AS=anterior rectus sheath; PS=posterior rectus sheath; A=epigastric artery (blood flow seen with colour Doppler function). Note needle visible over its whole length

Insert the block needle in-plane from lateral to medial. Note it can be difficult to puncture the skin with a block needle (either lift the skin and push the needle through or make a nick in the skin using a sharp bevelled needle). The needle is advanced from lateral to medial, in a shallow trajectory; aim to position the tip of the needle between the rectus muscle and the posterior rectus sheath.

Stop the needle tip just superficial to the first white line (the posterior sheath); often a small ‘give’ is felt as you come out

of the muscle into the potential posterior space. The needle is positioned correctly if the rectus sheath peels away from the muscle during injection of the local anaesthetic. Deposit local anaesthetic in this potential space between the rectus abdominis muscle and the posterior rectus sheath.

In neonates you could use saline to identify the correct plane, avoiding the waste of the limited local anaesthetic. Note that in small children it is possible to use a large footprint probe in the midline and block both sides without adjusting the probe.

Spread of the local anaesthetic after injection can be assessed by turning the probe into a paramedian longitudinal plane.

TRANSVERSUS ABDOMINIS PLANE BLOCK (TAP) BLOCK

The standard transversus abdominis plane (TAP) block provides intraoperative and postoperative analgesia for lower abdominal incisions. A subcostal TAP block can be provide analgesia for abdominal surgery above the umbilicus. TAP block can be performed unilaterally (e.g. inguinal hernia repair), or bilaterally (e.g. for laparoscopic surgery). It may be used as an alternative to an epidural, but it does not provide visceral analgesia; it should be performed after induction of anaesthesia, and adequate anaesthesia should be provided during visceral manipulation. A comprehensive review of the transversus abdominis plane (TAP) block can be found in WFSA ATOTW 239.⁸



Figure 8. Ultrasound probe position for the transversus abdominis plane (TAP) block

A TAP block is ideally used for inguinal hernia repair, colostomy formation, appendectomy (laparoscopic and open techniques), iliac crest bone graft harvesting, laparoscopic cholecystectomy, and laparoscopic nephrectomy or to provide analgesia after renal transplant.

Anatomy

- The skin of the anterior abdominal wall is supplied by the ventral rami of the inferior six thoracic spinal nerves (T7 to T12).
- In the lateral part of the anterior abdominal wall there are 3 muscle layers, from deep to superficial they are: the transversus abdominis muscle (TA, it is the most internal of the 3 muscle layers), the internal oblique muscle (IO, it is positioned in-between TA and EO) and the external oblique muscle (EO, it is the largest and most superficial of the 3 muscles).
- The ventral rami of T 7 to T12 run in the plane between the transversus abdominis muscle and the internal oblique muscle: the transversus abdominis plane (TAP).

Dose

Use a volume of 0.3 – 0.5ml.kg⁻¹ of 0.25% bupivacaine per side.

Techniques

The landmark technique is not recommended in children due to the danger of visceral damage and is therefore not described here. If ultrasound is not available, in terms of risk-benefit, other techniques such as simple infiltration with local anaesthetic are preferable.

Ultrasound-guided mid-axillary TAP block

Position the patient supine. Place a high frequency linear ultrasound probe in a transverse plane between the iliac crest and the costal margin. Start scanning from the linea alba in the midline, then move laterally until the probe is between the iliac crest and the costal margin (See Figure 8).

Identify the peritoneum and then the abdominal muscles: transversus abdominis, internal oblique, and external oblique. In obese children fascial planes may be present within the adipose tissue, this can lead to misidentification of the muscle layers; therefore always identify the muscle layers from deep to superficial.

The target is the fascial plane between the transversus abdominis and the internal oblique muscles. Advance the needle in an antero-posterior direction (See Figure 8). Aim to puncture the fascia on the deep aspect of the internal oblique muscle layer (a slight give or pop is often felt) (See Figure 9).

In neonates the muscles are less developed and therefore differentiation between the layers can be difficult to the inexperienced practitioner. The injection is seen as a neat lens-shaped deposit of local anaesthetic forming between the transversus abdominis muscle and the fascia separating the internal oblique muscle and transversus abdominis. If you are concerned that the local anaesthetic dose may be wasted whilst

identifying the correct plane, use saline to identify the correct plane before injecting the local anaesthetic dose.

In older children it may prove difficult to enter the TAP. Guide the needle very carefully into the transversus abdominis muscle, then slowly withdraw the needle as an assistant injects; as the needle enters the correct position, local can be seen spreading through the plane.

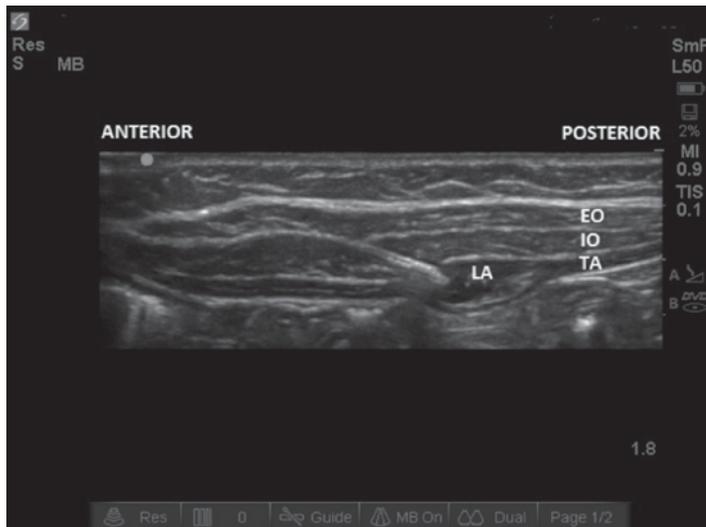


Figure 9. Ultrasound landmarks for TAP block: EO=external oblique muscle; IO=internal oblique muscle; TA=transversus abdominis muscle; LA=local anaesthetic; needle seen slanting from anterior.

Ultrasound guided subcostal approach

Position the patient supine. Place high frequency linear ultrasound probe in an oblique transverse plane, parallel to the subcostal margin (lateral to the rectus sheath). Identify the medial border of the external oblique, internal oblique and transversus abdominis muscles. Subsequently identify the TAP in between these inner two muscles. Insert the needle at the lateral edge of rectus abdominis muscle. Advance the needle in-plane away from the midline and parallel to the costal margin; use the local anaesthetic to hydrodissect the plane, carefully advancing the needle into the space created. Repeat this process until the anterior 2/3 of the subcostal margin has been covered.

Complications

Peritoneal perforation, organ perforation (in neonates the liver

and spleen are particularly prominent). The TAP block has a higher rate of complications than other blocks in children.⁹

FURTHER READING

<http://www.euroespa.com/science-education/specialized-sections/us-regional-anaesthesia/>

REFERENCES

1. Boretzky KR. Regional anesthesia in pediatrics: marching forward. *Curr Opin Anaesthesiol.* 2014; **27**: 556-560.
2. van Schoor AN, Boon JM, Bosenberg AT, Abrahams PH, Meiring JH. Anatomical considerations of the pediatric ilioinguinal/iliohypogastric nerve block. *Paediatr Anaesth.* 2005; **15**: 371-377.
3. Willschke H, Marhofer P, Bosenberg A et al. Ultrasonography for ilioinguinal/iliohypogastric nerve blocks in children. *Br J Anaesth.* 2005; **95**: 226-230.
4. Willschke H, Bosenberg A, Marhofer P et al. Ultrasonographic-guided ilioinguinal/iliohypogastric nerve block in pediatric anesthesia: what is the optimal volume? *Anesth Analg.* 2006; **102**: 1680-1684.
5. Weintraud M, Marhofer P, Bosenberg A et al. Ilioinguinal/iliohypogastric blocks in children: where do we administer the local anesthetic without direct visualization? *Anesth Analg.* 2008; **106**: 89-93.
6. Flack SH, Martin LD, Walker BJ et al. Ultrasound-guided rectus sheath block or wound infiltration in children: a randomized blinded study of analgesia and bupivacaine absorption. *Paediatr Anaesth.* 2014; **24**: 968-973.
7. Willschke H, Bosenberg A, Marhofer P et al. Ultrasonography-guided rectus sheath block in paediatric anaesthesia--a new approach to an old technique. *Br J Anaesth.* 2006; **97**: 244-249.
8. Russon K et al. Transversus abdominis plane block. WFSA Anaesthesia Tutorial of the Week 2011 No. 239. http://www.wfsahq.org/components/com_virtual_library/media/ea51ff0934644a9e41bcf82f65a96a58-474f4fcc0e20052dd9ed683ca9995db2-239-Transversus-Abdominis-Plane-Block.pdf
9. Long JB, Birmingham PK, De Oliveira GSJ, Schaldenbrand KM, Suresh S. Transversus abdominis plane block in children: a multicenter safety analysis of 1994 cases from the PRAN (Pediatric Regional Anesthesia Network) database. *Anesth Analg.* 2014; **119**: 395-99.

Upper and lower limb blocks in children

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INTRODUCTION

Regional anaesthesia is commonly used as an adjunct to general anaesthesia and plays a key role in the multimodal approach to perioperative pain management in children.^{1,2} Peripheral nerve blockade has increased with the development of age-appropriate equipment, safer, long-acting local anaesthetic agents, increasing experience and low complication rates.

The safety of peripheral nerve blocks in children has been established in large-scale prospective studies, although caudal epidural remains the most popular and frequently used block in infants and small children.³⁻⁵ The experience of the operator, pathology, the site and extent of surgery, the child's body habitus and the presence of contractures will dictate the final choice of technique.

This article outlines regional anaesthetic techniques and methods used to identify and block individual nerves to provide analgesia for surgical procedures of the upper and lower limbs. The differences between adults and children are highlighted together with techniques to improve the success of blocks.

METHODS TO IMPROVE ACCURACY OF PERIPHERAL NERVE BLOCKS

Peripheral nerve blocks, particularly in young children, can be challenging because anatomical landmarks are poorly defined and vary with age. Successful peripheral nerve blocks require an awareness of these differences, knowledge of developmental anatomy and an understanding of the equipment used.

Most children are sedated or under general anaesthesia when nerve blocks are performed. Ultrasound guidance is preferable when available. A peripheral nerve stimulator and

insulated needles are the next best option although successful blocks can be achieved with non-insulated needles.⁶

Nerve stimulator

Many peripheral nerve stimulators are available. Familiarise yourself with a particular device and stick with it. Read the manufacturer's instructions and understand how the nerve stimulator works before you use it.⁷

The following basic principles should be followed to locate a peripheral nerve or plexus accurately:

- Muscle relaxants must be withheld until after completion of the block.
- Attach the Negative electrode to the Needle and the Positive electrode to the Patient using a standard ECG electrode.
- Set the initial peripheral nerve stimulator output to 1-1.5 mA at 1-2 Hz for 0.1msec. Advance the needle through the skin and underlying tissue planes until you elicit nerve-specific muscle contractions distally.
- Decrease the current output and adjust the needle location until you see maximum motor response with least current i.e. at approximately 0.3-0.5mA.
- Aspirate and inject local anaesthetic (provided no blood returns when you aspirate as this implies the needle tip is within a blood vessel) - muscle twitches will stop immediately indicating a successful block is likely. If this does not happen you must reposition the needle and repeat the process.
- Do NOT inject local anaesthetic if you find vigorous muscle contraction at <0.2mA or resistance to injection. Both suggest intraneural injection, and nerve damage could result.

SUMMARY

This article outlines regional anaesthetic techniques and methods used to identify and block individual nerves to provide analgesia for surgical procedures of the upper and lower limbs. Expertly performed, peripheral nerve blocks can provide long lasting anaesthesia and analgesia for surgery or after injury to the upper or lower limbs in children.

Specific knowledge is required for the indication, technique and dose of local anaesthetic for each block. A safe dose of local anaesthetic must be used at all times, and care taken to avoid inadvertent intravascular injection of local anaesthetic. The differences between adults and children are highlighted together with techniques to improve the success of blocks.

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Surface nerve mapping

This is a modification of the standard nerve stimulator technique.⁸ The path of a superficial motor nerve (or plexus) can be traced by stimulating the motor component of the nerve transcutaneously. The nerve stimulator output is set at 2-5mA at 1-2Hz and the negative electrode used as the “mapping electrode”. The current required varies and is dependent on the depth of the nerve and the skin moistness. Mark the point at which maximal motor response is elicited. This serves as the surface landmark for that block. Excess pressure applied over the nerve may inhibit the response. Direct muscle stimulation is finer and more localized.

The ‘nerve mapping technique’ may be used for:

- The brachial plexus (supraclavicular, axillary)
- Musculocutaneous, ulnar, median and radial nerve blocks of the upper limb
- Femoral, sciatic and popliteal nerve blocks in the lower limb.

Surface nerve mapping is particularly useful where anatomical landmarks are difficult e.g. children with arthrogryposis or congenital limb defects.

Ultrasound guidance

This has become an important adjunct in regional anaesthesia; although anaesthesia departments in low- and middle-income countries (LMIC) may not be able to make this expensive item a priority.⁹⁻¹³ Using real time ultrasound imaging, correct needle and local anaesthetic placement around the nerve can be verified and thus the risk of intraneural or intravascular injection reduced. Detailed descriptions of ultrasound-guided blocks can be obtained from recent review articles.¹⁰⁻¹³

UPPER LIMB BLOCKS

The motor and sensory innervation of the whole upper extremity is supplied by the brachial plexus, with the exception of part of the shoulder (innervated by the cervical plexus), and the sensory innervation to the medial aspect of the upper arm (supplied by intercostobrachial nerve, a branch of the 2nd intercostal nerve).

Anatomy of the brachial plexus (see Figure 1)

The anterior primary rami of C5-8 and the bulk of T1 form the brachial plexus. These five roots emerge from the intervertebral foramina to lie between the scalenus anterior and scalenus medius muscles (which attach to the anterior and posterior tubercles of the transverse process of the cervical vertebrae respectively).

The fascia of these muscles encloses the plexus in a sheath that extends laterally into the axilla. A single injection of

local anaesthetic within this sheath produces complete plexus blockade by blocking the trunks (supraclavicular approaches) or the cords (infraclavicular approaches). As the spinal roots pass between the scalenus muscles they unite to form three trunks - upper C5-C6; middle C7; lower C8-T1. Emerging from the interscalene groove, the three trunks pass downward and laterally to lie postero-lateral to the subclavian artery as it crosses the upper surface of the first rib. The subclavian artery is not easily palpable above the clavicle in children. At the lateral border of the first rib, each trunk divides into anterior and posterior divisions, which then join to form the lateral, medial and posterior cords, named according to their relationship to the axillary artery. These cords then divide into the nerves of the brachial plexus – musculocutaneous, ulnar, median and radial.

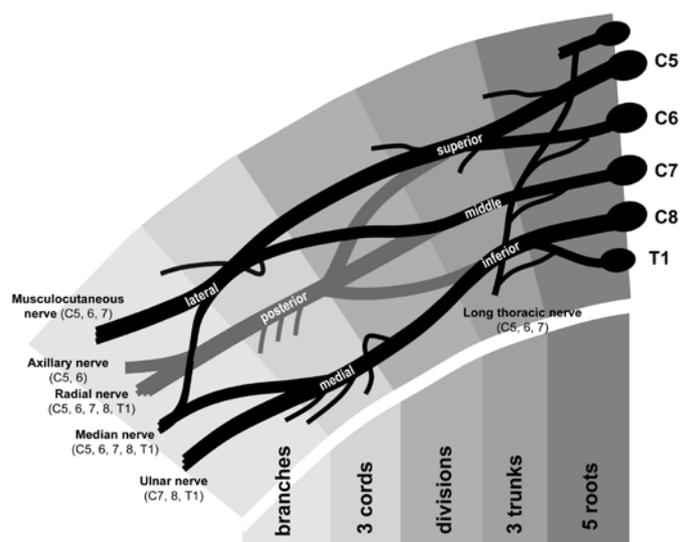


Figure 1. Diagrammatic representation of brachial plexus anatomy (reproduced with permission from: Harclerode Z, Michael S. Axillary brachial plexus block landmark techniques. *Tutorial of the Week 165 (January 2010)*)

Many anatomical landmarks used in adults maybe difficult to feel in anaesthetised children, particularly infants⁸. The scalenus muscles are poorly developed making the interscalene groove difficult to delineate. The subclavian artery is seldom palpable above the clavicle in infants and preadolescent children.

The brachial plexus can be blocked at various levels, the choice depending on the planned surgical procedure, the experience of the provider and anatomical variants (See Table 1).^{12,13}

Interscalene approach

Although the interscalene approach has been used for shoulder and elbow surgery in children, this approach must be used with caution.¹⁴ Potential complications include intravascular injection, intrathecal injection, pneumothorax (reported incidence in children is low), Horner’s syndrome and temporary phrenic nerve palsy.

Table 1. Approaches to the brachial plexus

Neck
Interscalene
Parascalene
Supraclavicular
Infraclavicular
Axillary
Perivascular approach
Transarterial approach

Supraclavicular approach

This is indicated for all upper extremity surgery, particularly if the shoulder is involved. Position: Supine, pillow under shoulders, arm extended along side the body, head turned to opposite side.

Landmarks: Clavicle-mid point, transverse process of C6 (Chassaignac's tubercle), posterior border sternocleidomastoid, cricoid cartilage, brachial plexus.

Dose: 0.2-0.3ml.kg⁻¹ 0.25-0.5% bupivacaine or 1-2% lignocaine. Lower concentrations reduce the degree of motor block.

Technique: With the patient correctly positioned, the components of the brachial plexus become more superficial and are easily palpable in most children. The site of puncture is at the junction of the middle and lower third of a line joining Chassaignac's tubercle to the midpoint of the clavicle (if Chassaignac's tubercle cannot be palpated extending a line from the cricoid cartilage to posterior border of sternocleidomastoid should suffice).

Alternatively, insert an insulated needle perpendicular to the skin at the site where maximal distal muscle twitches (usually flexion or extension at the elbow) is 'mapped' or simply over the point where the brachial plexus can be palpated.

Complications: Success rate is high. Complications caused by faulty technique include pneumothorax, vascular puncture, Horner's syndrome and phrenic nerve palsy.¹⁴ Nerve damage is possible with injudicious injection against resistance but the possibility of surgical damage should always be excluded.

Axillary approach

The axillary block is the most popular brachial plexus block in children.¹⁴⁻¹⁷ It is relatively safe and provides good analgesia for surgery of the forearm and hand. The risk of complications is

low. The main limitation is incomplete block of the shoulder and lateral aspect of the forearm onto the thenar eminence (musculocutaneous nerve sensory distribution). Axillary block may be used for a variety of procedures on the hand and forearm (particularly on the medial aspect), such as open reduction with internal fixation of a forearm fracture, closed reduction of forearm fractures, congenital hand anomalies (syndactyly repair), treatment of vascular insufficiency or finger re-implantation.^{16,17}

Perivascular approach

Position: Supine, arm abducted 90°, elbow flexed, hand behind head.

Landmarks: Pectoralis major, the coracobrachialis muscle, axillary artery.

Dose: 0.2-0.5ml.kg⁻¹ 0.25-0.5% bupivacaine or 1-2% lignocaine. Lower concentrations reduce the degree of motor block.

Technique: Palpate the axillary artery as high as possible in the axilla in the tissues overlying the humerus at the junction of the lower border of pectoralis major and coracobrachialis muscles (see Figure 2). If using a nerve stimulator technique, introduce an insulated needle immediately superior to the pulsation at a 45-60° angle to the skin and direct parallel to the artery towards the midpoint of the clavicle with the nerve stimulator set at 1mA.¹⁸ Distal muscle twitches are elicited, usually in the median or radial nerve distributions, as the sheath is penetrated. Gradually reduce the output of the nerve stimulator to approximately 0.3-0.4mA while the muscle twitch is maintained, adjust the position of the needle as needed. Local anaesthetic solution can then be injected.

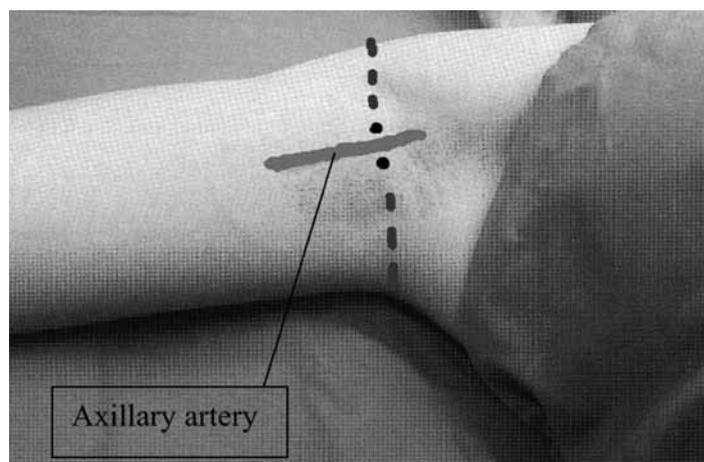


Figure 2. Axillary approach to brachial plexus. Palpate the axillary artery as high as possible in the axilla at the junction of the lower border of pectoralis major and coracobrachialis muscle (reproduced with permission from: Harclerode Z, Michael S. Axillary brachial plexus block landmark techniques. *Tutorial of the Week* 165 (January 2010))

Alternatively, use a nerve stimulator set at 3-5mA to map the median nerve, radial and ulnar or musculocutaneous nerves transcutaneously in the axilla on either side of the arterial pulsation. Insert an insulated needle at the point of maximum muscle twitch.

A cannula-over-needle technique can be used to cannulate the brachial plexus sheath in the axilla using a landmark technique. The axillary artery is located as described above, and a 22G cannula is inserted at 30° to the skin, anterior to the artery and in a line parallel to the pulsation of the axillary artery.¹⁷ The cannula is passed over the needle after feeling a 'click' when the cannula pierces the axillary sheath. Check for negative aspiration of blood and inject 0.5ml.kg⁻¹ 0.25% plain bupivacaine.

Whichever technique is used, distal pressure applied during and immediately following injection will facilitate proximal spread and blockade of the musculocutaneous nerve.

The musculocutaneous nerve may be blocked separately to provide analgesia for procedures involving the lateral forearm. Advance a needle, introduced perpendicularly to the skin just above the axillary pulsation, into the coracobrachialis muscle

until forearm flexion is elicited using a nerve stimulator. Inject 0.5-1ml local anaesthetic just deep to the fascia.

Complications are rare but include hematoma from accidental vascular puncture; in this event apply pressure for at least 5 minutes.

Infraclavicular approach

Position: Supine, pillow under shoulders, head turned to opposite side, upper arm adducted alongside body, elbow flexed to 90° with forearm placed on abdomen.

Landmarks: Clavicle-lower border, coracoid process of scapula, axillary artery as it emerges beneath clavicle.

Dose: 0.2-0.3ml.kg⁻¹ 0.25-0.5% bupivacaine, 1-2% lignocaine. Lower concentrations reduce the degree of motor block.

Technique: A number of approaches have been described.¹⁸⁻²⁰ With the child positioned correctly, divide the clavicle into three parts. Make a mark at the point where the pulse is felt as it emerges below the clavicle, or where any distal flexion or pronation is "mapped". Insert a needle infra-clavicularly at the junction of the middle and lateral third of the clavicle and

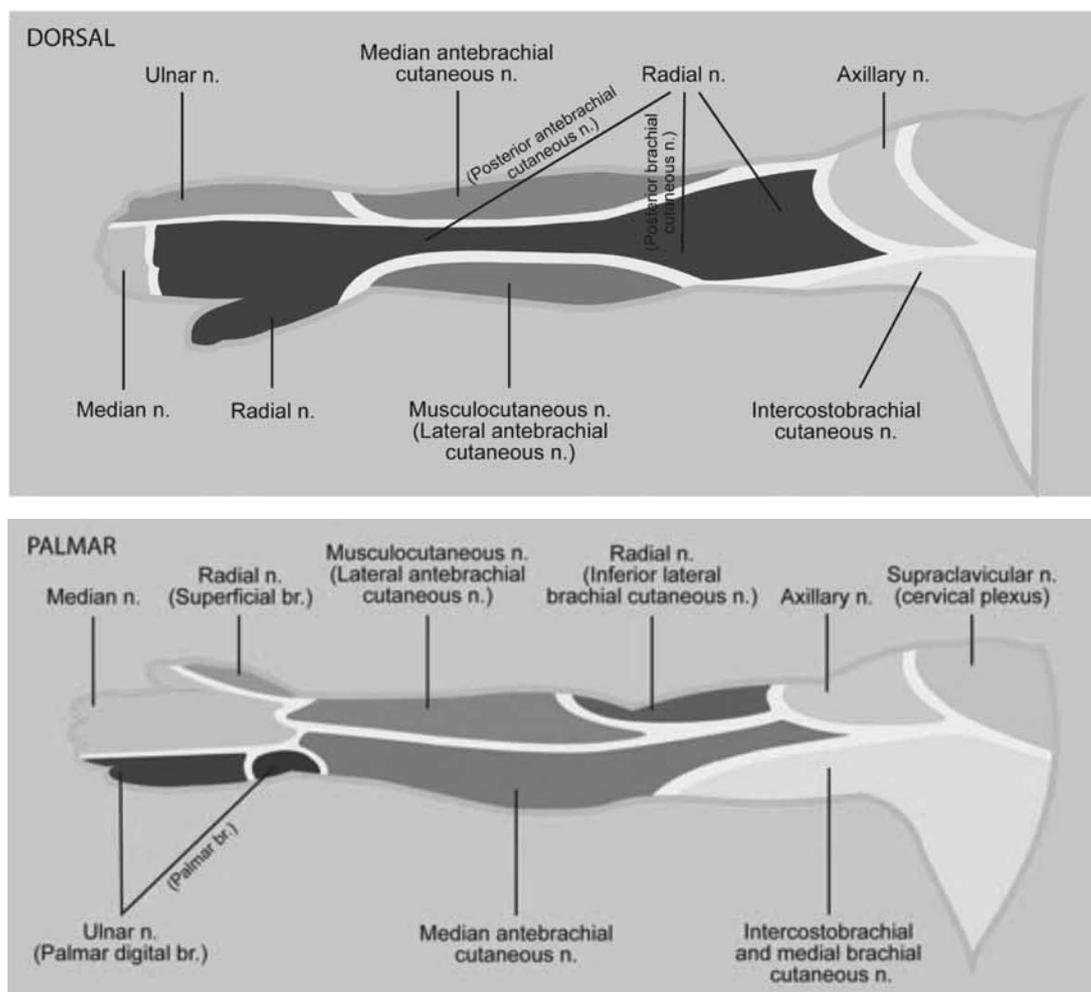


Figure 3. Cutaneous nerve supply of the upper limb. (reproduced with permission from: Harclerode Z, Michael S. Axillary brachial plexus block landmark techniques. Tutorial of the Week 165 (January 2010)

directed towards this mark. The needle passes lateral to the cupola of the lung and is unlikely to encounter the lung along its course. Seek pronation or flexion at the elbow. Once the nerve is located, reduce the voltage on the nerve stimulator to 0.2 to 0.3mA.

Alternatively, insert the needle at the midpoint of the lower border of the clavicle at 45-60° angle and direct towards the axilla in the same manner until distal muscle twitches are elicited.

A vertical infra-clavicular approach using the coracoid process as a landmark has also been described in children.¹⁸⁻²⁰ The site of puncture is 1-2cm caudad and 0.5-1cm lateral to the coracoid process in the lower part of the delto-pectoral groove. Insert the needle perpendicular to the skin until distal muscle twitches are elicited.

Location of the brachial plexus may be difficult in some patients and ultrasound guided approach has been recommended.^{12,21} Proponents of this technique claim more effective sensory and motor block than with the axillary approach.^{20,21}

ELBOW BLOCKS

Median nerve

Indication: Surgery on volar aspect of the forearm and the palmar portion of the hand.

Position: Supine, arm extended, elbow slightly flexed to accentuate the tendons of the biceps and the brachioradialis.

Landmarks: Cubital fossa, brachial artery, biceps tendon.

Dose: 0.1-0.2ml.kg⁻¹ 0.25-0.5% bupivacaine or 1-2% lignocaine.

Anatomy: The median nerve in the cubital fossa is located medial to the brachial artery and the biceps tendon beneath the deep fascia.

Technique: After 'surface mapping' the median nerve, insert an insulated needle medial to the pulsation of the brachial artery. Pronation of the arm with opposition of the fingers is noted when the median nerve is stimulated.

Ulnar nerve

Indication: For surgery in the ulnar distribution of the hand including the medial aspect of the hand and fingers

Position: Supine, elbow flexed 90°, arm on chest with hand on opposite shoulder.

Landmarks: Olecranon groove.

Dose: 1-2ml 0.25-0.5% bupivacaine, or 1-2% lignocaine.

Anatomy: The ulnar nerve lies in the groove posterior to the medial condyle of the humerus midway between the olecranon

and the medial epicondyle.

Technique: The ulnar nerve can be blocked at the olecranon groove. A small volume of 0.25% bupivacaine is injected into the area.

Complications: Nerve injury, compression of the nerve.

Radial nerve

Position: Supine, elbow slightly flexed.

Landmarks: Biceps tendon, lateral condyle of humerus.

Dose: 1-2ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

Technique: Identify lateral condyle of humerus and tendon of the biceps muscle. The radial nerve lies adjacent to the condyle, lateral to the biceps tendon. With the arm slightly flexed at the elbow, the radial nerve can be stimulated with a mapping probe in this position. Movement of the thumb confirms the location and small volume of local anaesthetic can be injected at that point.

Musculocutaneous nerve in the forearm

Position: Arm extended or arm on abdomen.

Landmarks: Lateral condyle of humerus.

Dose: 1-2ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

The musculocutaneous nerve courses superficially along the lateral aspect of the forearm where it can be mapped along most of its course. A superficial ring of local anaesthetic solution (0.1ml.kg⁻¹) injected at the distal end of the lateral condyle of the humerus along the pronator teres muscle should block the musculocutaneous nerve.

Wrist blocks

An appropriate nerve block at the wrist may be used to provide analgesia for children undergoing minor surgical procedures on the hand or fingers. The nerves that can be blocked at this level are the median, ulnar and radial nerves. Small volumes of local anaesthetic (1-2ml) can provide good analgesia for a number of hours.

Median nerve

The median nerve is the major nerve supplying the hand and therefore most surgical procedures of the hand will require a median nerve block at least.

Position: Hand pronated.

Landmarks: Palmaris longus tendon. Volar aspect wrist

Dose: 0.5-1ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

Anatomy: The median nerve lies within a fascial sheath between palmaris longus tendon and flexor carpi radialis. Surface nerve mapping at this point will elicit opposition of the thumb. A

sheath that communicates with the neurovascular bundle is located at the ulnar aspect of the palmaris longus tendon.

Complications: Rare - carpal tunnel syndrome or injury to the median nerve can be avoided by restricting the volume of local anaesthetic used.

Radial nerve

Position: Hand supinated.

Landmarks: Anatomical 'snuff box', styloid process-radius, radial artery.

Doses: 1-2ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

Anatomy: The radial nerve at the wrist is purely sensory and thus nerve mapping is not possible. Just above the styloid process, the radial nerve divides into two branches, one supplying dorsum of the hand and another supplying the thenar eminence and 1.5 fingers.

Technique: The nerve lies superficial proximal to the 'anatomical snuff box'. Inject a wheal subcutaneously starting lateral to the radial artery on the lateral aspect of the wrist using a fine needle.

Ulnar nerve

Position: Hand supinated.

Landmarks: Flexor carpi ulnaris tendon, ulnar artery.

Dose: 1-2ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

Anatomy: The ulnar nerve is located in the palmar sheath immediately lateral to the flexor carpi ulnaris tendon but medial to the ulnar artery. "Mapping" at this point will elicit flexion of the little finger. Inject into this area to provide analgesia for surgery on the ulnar aspect of the hand and the medial 1.5 fingers.

Bier's block or intravenous regional anaesthesia

Bier's block or intravenous regional anaesthesia (IVRA) is performed by introducing local anaesthetic into a limb isolated by means of tourniquet. IVRA is suitable for short (<30min) surgical procedures on the distal arm or leg or reduction of fractures in older children. Operating time is limited by tourniquet pain (30-40 min).

Equipment: Single or double cuff tourniquet cuff, IV cannula, monitors.

Drugs: 10-40ml 0.5% lignocaine or prilocaine depending on limb and size of patient. Maximum safe dose: lignocaine 3mg.kg⁻¹; prilocaine 6mg.kg⁻¹

Technique: Set up monitors (ECG, BP, SpO₂) and insert an IV cannula as distal as possible in the limb to be operated upon.

Insert a second cannula into the opposite arm for IV access in case of emergency. Elevate the limb for several minutes to exsanguinate. Inflate the tourniquet 50-100mmHg above systolic BP and ensure the cuff does not leak. Inject the local anaesthetic slowly via the IV cannula. Surgery may proceed after about 5 minutes. The tourniquet must remain inflated for a minimum of 20 minutes from the time of local anaesthetic injection. At the end of the procedure the cuff can be deflated - closely observe the patient for at least 10min for signs of toxicity. IVRA is generally safe provided the tourniquet cuff does not leak or accidentally deflate. A purpose made 'double cuff' may be used to increase the safety of this technique and to manage pain from the tourniquet (see <http://www.nysora.com/techniques/3071-bier-block.html>).

LOWER LIMB BLOCKS

Most lower extremity procedures can be performed under a caudal block (see article p88). Peripheral nerve blocks are more specific and confined to the site of surgery, the duration of analgesia is longer and the potential side effects of neuraxial blockade can be avoided (bilateral motor weakness, urinary retention).²³⁻²⁵

Anatomy of the lumbar and sacral plexus

Motor and sensory innervation of the lower extremity is supplied by the lumbar and sacral plexus. The lumbar plexus is

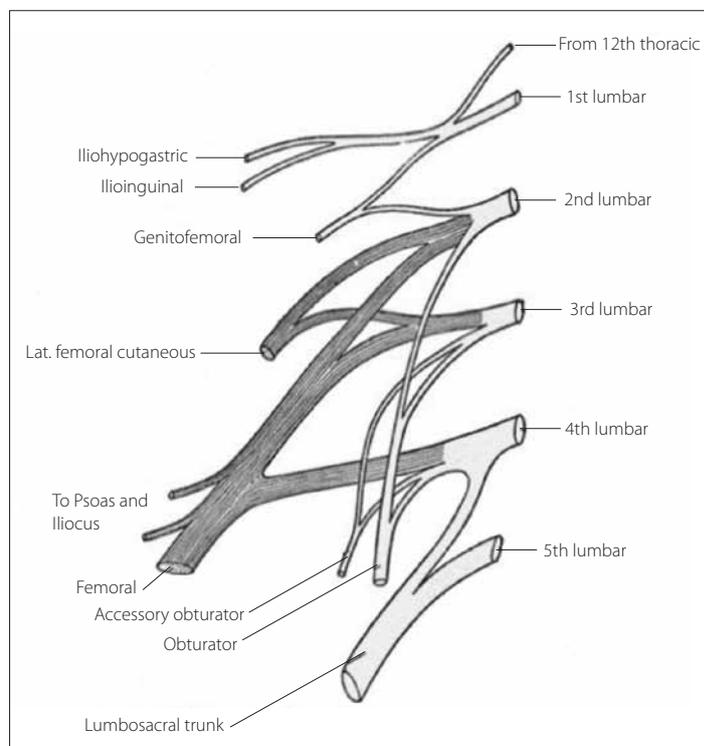


Figure 4. Anatomy of the lumbar and sacral plexus reproduced with permission from McEwen A. Femoral nerve block: landmark approach. TOTW 249 (February 2012)

Table 2. Lower extremity nerve blocks

Lumbar plexus block

Femoral nerve

Lateral cutaneous nerve of thigh

Sciatic nerve

Posterior approach

Infragluteal approach

Popliteal fossa approach

Ankle block

derived from the anterior primary rami of lumbar nerves L1-L4 and a variable contribution from T12 and L5. The lumbar plexus is located anterior to the transverse processes of the lumbar vertebrae within the psoas major muscle. The psoas compartment is bordered posteriorly by quadratus lumborum and anteriorly by psoas major. The femoral nerve, lateral cutaneous nerve of thigh, and obturator nerve are branches of the lumbar plexus and supply the majority of the upper leg including the thigh and its lateral aspect. The saphenous nerve provides sensory innervation below the knee to the medial aspect of the lower leg and foot. (See Table 2 and Figure 4)

The sacral plexus is derived from the anterior primary rami of L5, S1-S3 with contributions from L4 and S4. The plexus lies anterior to the piriformis muscle behind the pelvic fascia on the posterior wall of the pelvic cavity. The sciatic nerve is derived from the sacral plexus to supply the knee, the leg and most of the foot except for the medial aspect supplied by the saphenous nerve. A proximal branch of the sciatic nerve, the posterior cutaneous nerve of the thigh, supplies the posterior aspect of the thigh and the hamstring muscles.

Lumbar plexus block

The lumbar plexus can be blocked within the psoas muscle at the level of the transverse process of L4. In children the transverse process is not fully developed and using the transverse process as a guide usually places the needle too medial and increases the risk of complications, i.e. spinal anaesthesia secondary to puncture of the dural cuff on the spinal roots, or retrograde epidural spread to opposite side.

Indications: Unilateral block of the hip (congenital dislocation of hip), thigh (open reduction and internal fixation of femoral fractures) or knee surgery.

Position: Lateral position, hips and knees flexed.

Landmarks: Posterior superior iliac spine, intercrystal line, spinous process L4.

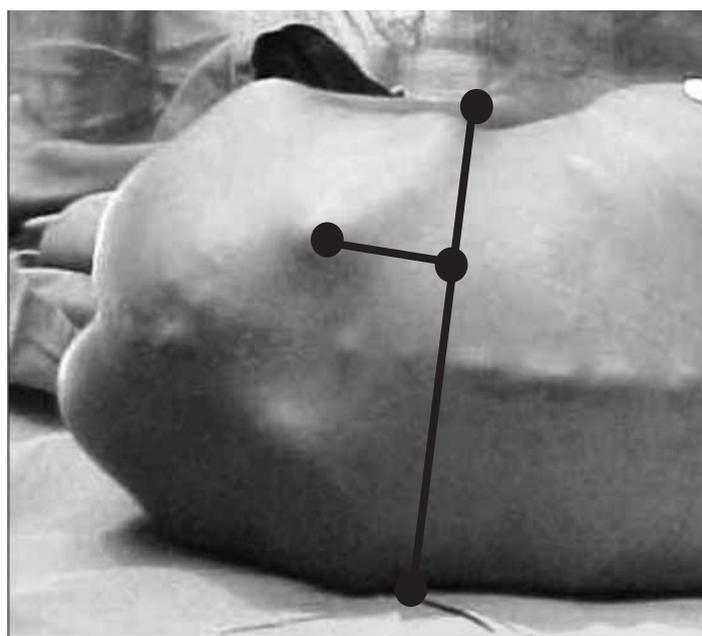


Figure 5. Psoas compartment block. The needle is inserted at the intersection of a perpendicular line drawn from the posterior superior iliac spine to the intercrystal line.

Dose: 0.5ml.kg^{-1} 0.25-0.5% bupivacaine or 1-2% lignocaine.

Technique: With the child in position, insert an insulated needle perpendicular to the skin where a line drawn from the posterior superior iliac spine (PSIS) parallel to the spinous processes of the vertebrae intersects the intercrystal (Touffier's) line (see Figure 5). Advance the needle slowly through the posterior lumbar fascia, paraspinous muscles, anterior lumbar fascia, quadratus lumborum and into the psoas muscle. Passage through these fascial layers may be detected by distinct "pops" when using a short bevelled needle. Using a nerve stimulator quadriceps muscle twitches in the ipsilateral thigh are sought. If hamstring contractions are observed, direct the needle laterally. If hamstring and quadriceps contractions are observed simultaneously, direct the needle more cephalad to isolate the lumbar plexus rather than sacral plexus.²⁷

The depth from the skin to the lumbar plexus is approximately the same distance as the posterior superior iliac spine is to the intercrystal line.²⁷ The depth of the needle is emphasized because complications include renal haematoma, vascular puncture (retroperitoneal haematoma) or even bowel puncture.

Femoral nerve block

Position: Supine, foot rotated outward.

Landmarks: Femoral pulse, inguinal ligament.

Dose: $0.2\text{-}0.3\text{ml.kg}^{-1}$ 0.25-0.5% bupivacaine or 1-2% lignocaine.

Femoral nerve blocks are most useful in cases of fractured femur, and allow painless transport, radiographic examination

and application of splints.^{23,28,29} Most lower limb surgery can be performed when femoral nerve block is used in combination with sciatic nerve or lateral cutaneous nerve of thigh block. For surgery on the knee it is best used in combination with a sciatic nerve block.^{23,30}

Technique: The femoral nerve may be blocked as it emerges from below the inguinal ligament in the femoral canal lateral to the femoral artery in the femoral triangle. Although a femoral nerve block can be performed without a nerve stimulator (e.g. for unsedated children with femoral fractures to avoid pain caused by muscle contractions) greater success can be achieved using one.

Insert the needle approximately 0.5-1cm lateral to the femoral pulsation and approximately 0.5-1cm below the inguinal ligament. It is useful to map the course of the femoral nerve prior to inserting the needle.⁸

The nerve lies beneath the fascia lata and fascia iliaca and often two distinct 'pops' are felt as needle traverses these layers. Quadriceps contraction confirms femoral nerve stimulation but should not be confused with direct stimulation of the sartorius muscle. Local anaesthetic should be easy to inject into the femoral canal. Resistance to injection suggests intraneural injection.

"3 in 1" block

A "3 in 1" block is essentially a femoral nerve block that attempts to anaesthetize the femoral, lateral cutaneous nerve of thigh and obturator nerves with one injection by causing retrograde spread of local within the femoral sheath up to the lumbar plexus by applying digital pressure distal to the injection site. It is only 20% effective in blocking all three nerves.³¹ All three nerves are more reliably blocked with a fascia iliaca or a lumbar plexus block.

Lateral femoral cutaneous nerve of thigh

Indications: To provide analgesia for plating of the femur, plate removal, drainage of femoral osteitis, harvesting skin grafts or muscle biopsies.

Position: Supine.

Landmarks: Anterior superior iliac spine, inguinal ligament.

Dose: 1-3ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

Anatomy: The lateral cutaneous nerve of thigh is derived from the lumbar plexus (L2-L3) and is purely sensory supplying the anterolateral aspect of the thigh. The nerve descends over the iliacus muscle just below the pelvic rim in an aponeurotic canal formed in the fascia lata and enters the thigh close to the anterior superior iliac spine (ASIS) behind the inguinal ligament. In the thigh it crosses or passes through the tendinous origin of the sartorius muscle. It divides into anterior and

posterior branches.

Technique: Insert a needle 1-2 cm below and medial to the origin of the inguinal ligament at the anterior superior iliac spine until a 'pop' is felt as the fascia lata is penetrated and a second 'pop' when the fascia iliaca is entered. If bone contact is made, the needle should be withdrawn and redirected. Correct placement of the needle can also be determined with a loss of resistance technique noted as the fascia iliaca compartment is entered.

Fascia iliaca compartment block

Indications: This block is particularly useful for any surgery performed on the lower extremity above the knee and for femoral shaft fractures. The fascia iliaca compartment block was originally described in children by Dalens and is more effective in blocking the femoral, lateral cutaneous nerve of thigh and obturator nerves than the "3-in-1" block.³¹

Position: Supine.

Landmarks: ASIS, inguinal ligament, pubic tubercle.

Dose: 0.5-1ml.kg⁻¹ 0.25-0.5% bupivacaine, or 1-2% lignocaine.

Technique: With the child in the supine position and the thigh slightly abducted and externally rotated, draw a line from the pubic tubercle to the ASIS along the inguinal ligament. Insert a needle perpendicular to the skin 0.5-1cm below the junction of lateral and middle third of this line. Two "pops" are felt as the needle pierces the fascia lata and then the fascia iliaca. Larger volumes are required because the aim is to block all three nerves with one injection. Massaging the area in an upward direction may facilitate the upward spread of local anaesthetic. No significant complications are seen provided that the needle remains (below) inferior to the inguinal ligament and away from the femoral vessels.

Sciatic nerve block

The sciatic nerve leaves the posterior pelvis via the greater sciatic foramen through the piriformis muscle into the buttock and descends in the midline of the leg posteriorly to the apex of the popliteal fossa. The sciatic nerve lies midway between the greater trochanter and the ischial tuberosity at the gluteal cleft where it is palpable in most young children.⁶ The sciatic nerve divides into the common peroneal and the tibial nerve within the popliteal fossa in the majority of children.³²

The sciatic nerve can be blocked using several different approaches at the hip or in the popliteal fossa.³²⁻³⁵ The approach chosen ultimately determines the distribution of motor and sensory blockade. Sciatic nerve block at the gluteal level provides anaesthesia for the posterior aspect of the thigh and leg below the knee but excludes the medial aspect of the

lower half of the leg, the medial malleolus and the medial aspect of the foot.

Indications: All surgical procedures involving the posterior aspect of the leg especially below the knee, for example lengthening of the Achilles tendon, uni- or bilateral club foot repair as well as for major foot arthrodesis.⁶ It may need to be supplemented with other blocks depending on the type of surgery, for example knee surgery or tibial osteotomies.

The different approaches to the sciatic nerve in children are described below.

Posterior approach to the sciatic nerve

Position: Lateral recumbent, hip flexed, knee flexed, the side to be blocked uppermost.

Landmarks: Coccyx, greater trochanter, ischial tuberosity.

Dose: 0.2-0.3ml.kg⁻¹ 0.25-0.5% bupivacaine or 1-2% lignocaine.

Technique: With the child in position, draw a line from the tip of the coccyx to the greater trochanter. Insert a needle at the midpoint of this line and direct towards the ischial tuberosity.

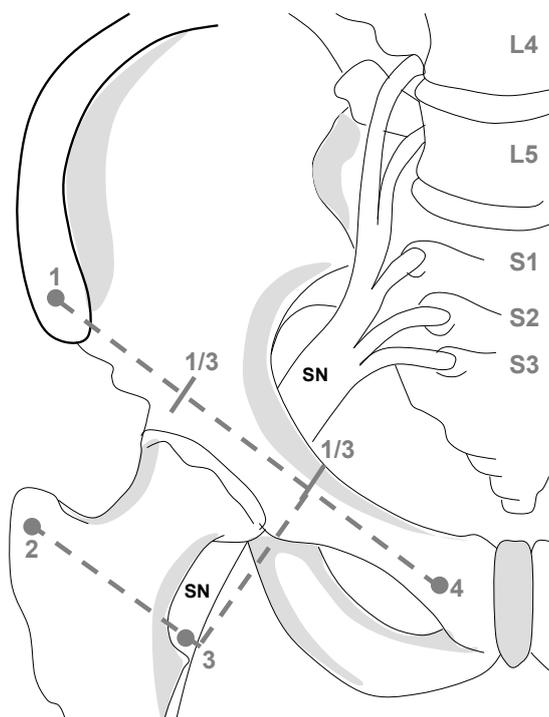


Figure 6. Anterior approach to the sciatic nerve: anatomy. Morphett S. Nerve blocks for anaesthesia and analgesia of the lower limb A practical guide: Femoral, Lumbar plexus, Sciatic. Update in Anaesthesia 11(2000):56-66

A nerve stimulator, set at 0.3 to 0.4mA, will elicit distal motor responses in the leg or foot confirming stimulation of the sciatic nerve (which supplies all the muscles below the knee).

Infragluteal approach to the sciatic nerve

Position: Supine or lateral decubitus, hip flexed, knee extended.

Landmarks: Greater trochanter, ischial tuberosity, gluteal crease, biceps femoris muscle.

Dose: 0.2-0.3ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

Technique: A useful technique in children has been described by Raj.^{6,35} Insert a needle posteriorly perpendicular to the skin at a point midway between the ischial tuberosity and greater trochanter on the gluteal crease. With exaggerated hip flexion the glutei may be flattened and the sciatic nerve becomes relatively superficial even in some obese children. The nerve is often palpable in the groove lateral to biceps femoris in young children. Plantar flexion, inversion or dorsi-flexion at 0.3-0.4 mA confirms sciatic nerve stimulation. The posterior cutaneous nerve of the thigh may be missed since the nerve may separate more proximally in the thigh.

Anterior approach to the sciatic nerve

Position: Supine, hip and knee extended

Landmarks: pubic tubercle, anterior superior iliac spine, greater trochanter

Dose: 0.2-0.3ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

Technique: (see Figure 6). A line drawn from the pubic tubercle to the anterior superior iliac spine (inguinal ligament) is divided into thirds. A perpendicular is then dropped from the junction of the inner and middle thirds onto a line drawn parallel to the inguinal ligament through the greater trochanter. This point corresponds approximately with the lesser trochanter below.

Posterior mid-thigh approach to the sciatic nerve

Position: Supine, hip flexed, knee flexed or extended.

Landmarks: Ischial tuberosity, head of fibula.

Dose: 0.2-0.3ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

Technique: Insert a needle perpendicular to the midpoint of a line drawn from the head of fibula to the ischial tuberosity in the posterior thigh. The nerve is surrounded by the hamstring muscles at this point and this compartment can be located using a loss of resistance technique or by nerve stimulation that produces a motor response in the ankle, foot or big toe.³⁶ The posterior cutaneous nerve of thigh is missed at this level.

Sciatic nerve in the popliteal fossa

Position: Prone, lateral or supine.

Landmarks: Apex-popliteal fossa.

Dose: 1-2ml 0.25-0.5% bupivacaine or 1-2% lignocaine.

The sciatic nerve may be blocked as it courses through the popliteal fossa behind the knee for procedures of the distal

lower extremity.³⁶⁻³⁸ The popliteal fossa is formed by the semimembranosus and semitendinosus tendons medially, the biceps femoris tendon laterally, and the popliteal crease between the femoral condyles inferiorly.

The sciatic nerve divides in the vicinity of popliteal fossa into two branches - the common peroneal and the posterior tibial nerves. The exact location of this division is variable but in the majority of children it is within the popliteal fossa.^{32,39} The common peroneal nerve courses laterally medial to the biceps femoris tendon before passing over the lateral head of gastrocnemius and around the head of the fibula. The posterior tibial nerve courses down the midline of the lower leg posteriorly in close proximity, but superficial to the popliteal artery within the popliteal fossa.

Although the nerves branch, there is a common epineurial sheath that envelops both the posterior tibial and the common peroneal nerve.³⁸ For this reason a high rate of success can be achieved even when the motor response of only one branch is elicited. Stimulation of the common peroneal will cause dorsiflexion and eversion of the foot while stimulation of the posterior tibial nerve will elicit inversion and plantar flexion. (Internal nerve, i.e. posterior tibial nerve - Inversion; External nerve i.e. common peroneal nerve - Eversion). The nerves can be "mapped" individually particularly in young infants.

Various landmarks have been described for the insertion of the needle. For each 10kg body weight the needle insertion moves 1cm further above the popliteal crease just lateral to the midline.³² Alternatively, insert a needle at the apex of the popliteal fossa to block both the posterior tibial and common peroneal nerve.

Complications are rare but take care to avoid intravascular injection.

Lateral approach to the sciatic nerve at the knee

Position: Supine.

Landmarks: Biceps femoris tendon.

Dose: 0.1ml.kg⁻¹ 0.25-0.5% bupivacaine or 1-2% lignocaine.

A lateral approach to the popliteal fossa has recently been described in children.⁴⁰ Identify the biceps femoris tendon on the postero-lateral aspect of the knee approximately 4-6 cm above the popliteal crease. Insert a needle anterior to the biceps femoris tendon until the needle contacts the shaft of the femur. At this point gently walk the needle off the femur posteriorly and advance until foot dorsiflexion or plantar flexion along with eversion is elicited.

Saphenous nerve block

Indication: The main indication for blocking this nerve is to

compliment a sciatic nerve block for surgery on the medial aspect of the lower limb or foot.

Position: Supine.

Landmarks: Femoral artery, sartorius, inguinal ligament.

Dose: 0.1-0.2ml.kg⁻¹ 0.25-0.5% bupivacaine or 1-2% lignocaine.

The saphenous nerve runs along the medial aspect of the thigh just lateral to and within the same fascial sheath as the motor nerve supplying the vastus medialis.

The nerve to vastus medialis can be located using a nerve stimulator. Insert an insulated needle perpendicular to the skin 0.5cm lateral to the point where the femoral artery crosses the medial border of the sartorius muscle in the anterior thigh. Muscle twitches in the sartorius muscle confirm the close proximity to the saphenous nerve at this level. The distance from the inguinal ligament (3-5cm) varies with age, as does the depth of the nerve (0.5-3cm).

An advantage of this block over a femoral block is that motor activity in the remainder of the quadriceps is spared.

The saphenous nerve is a purely sensory nerve at the knee. At the level of the tibial plateau the saphenous nerve perforates the fascia lata between the sartorius and gracilis where it lies subcutaneously in close proximity to the long saphenous vein.

Make a deep linear subcutaneous infiltration below and behind the insertion of the sartorius tendon (medial surface of tibia) where the nerve lies in a shallow gutter immediately in front of the upper part of the medial head of gastrocnemius. Intermittent aspiration will reduce the risk of injection into the long saphenous vein.

Ankle block

Position: Supine or prone.

Landmarks: Medial and lateral malleolus, extensor hallucis longus tendon, Achilles tendon, dorsalis pedis pulse.

Dose: 0.1ml.kg⁻¹ 0.25-0.5% bupivacaine or 1-2% lignocaine.

Ankle blocks are used for procedures confined to the foot including distal phalangeal amputations, foreign body removal and simple reconstructive surgery. The five peripheral nerves blocked at this level are the terminal branches of the sciatic (posterior tibial, superficial peroneal, deep peroneal and sural nerves) and femoral (saphenous) nerves.

An ankle block is relatively easy to perform by injecting a subcutaneous ring at the ankle. Avoid local anaesthetics containing adrenaline since it may compromise end-arteries in the foot. Block each nerve separately for best results (see Figure 7).

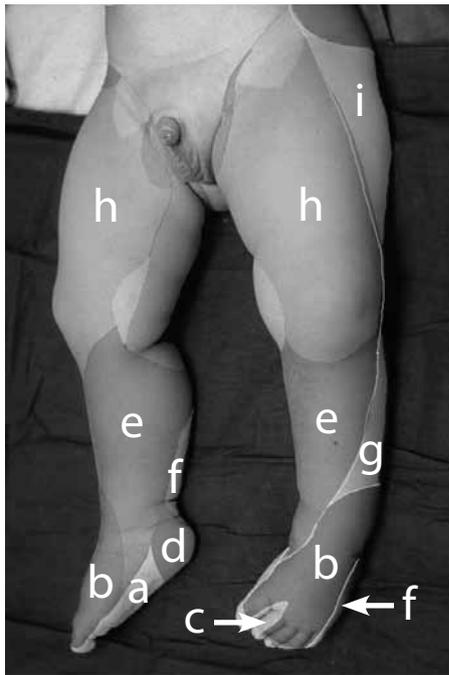


Figure 7. Cutaneous nerve supply of the leg and foot, demonstrating the nerves involved in an ankle block: a) medial and lateral plantar nerves; b) superficial peroneal nerve; c) deep peroneal nerve; d) medial calcaneal nerve (branch of the tibial nerve); e) saphenous nerve; f) sural nerve; g) lateral cutaneous nerve; h) femoral nerve; i) lateral femoral cutaneous nerve

- (a) The tibial nerve innervates the sole of the foot and medial aspect of the heel and lies between the medial malleolus and the calcaneum deep to the flexor retinaculum immediately posterior to the posterior tibial artery. Local anaesthetic injected through a needle inserted postero-medially to the Achilles tendon and directed towards the arterial pulsation should block the tibial nerve.
- (b) The superficial peroneal nerve supplies the dorsum of the foot and is blocked by a subcutaneous injection across the dorsum of the foot between the lateral malleolus and the extensor hallucis longus tendon.
- (c) The deep peroneal nerve innervates the first web space between the 1st and 2nd toe. Insert a needle medial to extensor hallucis longus tendon along anterior tibial arterial pulsation until it contacts the tibia. Withdraw the needle a few millimetres and inject the local anaesthetic solution.
- (d) The sural nerve innervates the lateral aspect of the foot. Introduce a needle postero-lateral to the Achilles tendon between the lateral malleolus and the calcaneum until bony contact is made. Withdraw the needle a few millimetres and inject the local anaesthetic solution.
- (e) The saphenous nerve innervates the medial aspect of the ankle and foot and lies in close proximity to the saphenous

vein. It is located on the medial side of the dorsum of the foot anterior to the medial malleolus. A subcutaneous injection from the medial malleolus along the anterior aspect of the ankle towards the saphenous vein will block the nerve.

SAFE CONDUCT OF PERIPHERAL NERVE BLOCKS

Patients should be carefully monitored whilst nerve blocks are performed. This should include ECG, BP and pulse oximetry. Resuscitation equipment (oxygen, anticonvulsants, Ambu bag and Intralipid) should be available in the event of local anaesthetic toxicity or anaphylaxis. (See Table 3)

Table 3. Management of local anaesthetic toxicity

Management of systemic toxicity⁴²

Airway - Maintain the patient's airway, administer 100% oxygen

Breathing - Start ventilation: if breathing inadequate intubate

Circulation - Pulse check: if cardiac arrest, start CPR.

Convulsions - IV diazepam, 1-3mg.kg⁻¹ thiopentone, muscle relaxation prn

Hypotension - Ephedrine 3-6mg increments, elevate legs, IV fluid bolus

Intralipid 20% - 1-1.5ml.kg⁻¹ followed by infusion 0.25ml.kg⁻¹.min⁻¹

CONCLUSION

Expertly performed, peripheral nerve blocks can provide long lasting anaesthesia and analgesia for surgery or after injury to the upper or lower limbs in children. Specific knowledge is required for the indication, technique and dose of local anaesthetic for each block. Accuracy is improved if a nerve stimulator or ultrasound guidance is used. A safe dose of local anaesthetic must be used at all times, and care taken to avoid inadvertent intravascular injection of local anaesthetic. Additional information and video descriptions of peripheral nerve blocks can be obtained from the Internet, for instance from the New York School of Anaesthesia website: <http://www.nysora.com/>

REFERENCES

1. Suresh S, Wheeler M. Practical pediatric regional anesthesia. *Anesthesiol. Clin. North America* 2002; **20**: 83-113.
2. Ross AK, Eck JB, Tobias JD. Pediatric regional anesthesia: beyond the caudal. *Anesth Analg* 2000; **91**: 16.
3. Giaufre E, Dalens B, Gombert A. Epidemiology and morbidity of regional anesthesia in children: A one-year prospective survey of the French-language society of pediatric anesthesiologists. *Anesth Analg* 1996; **83**: 904-12.

4. Polaner DM, Taenzer AH, Walker BJ et al. Pediatric Regional Anaesthesia Network (PRAN): A Multi-Institutional Study of the Use and Incidence of Complications of Pediatric Regional Anaesthesia. *Anesth Analg* 2012; **22**: 115-9.
5. Ecoffey C, Lacroix F, Giaufre E et al. Epidemiology and morbidity of regional anaesthesia in children: a follow-up one-year prospective survey of the French- Language Society of Paediatric Anaesthesiologists (ADARPEF). *Pediatr Anesth* 2010; **20**: 1061-9.
6. Bosenberg AT. Lower limb blocks in children using unsheathed needles and a nerve stimulator. *Anaesthesia* 1995; **50**: 206-10.
7. Hadzic A. Peripheral nerve stimulators: cracking the code – one at a time. *Reg Anesth Pain Med* 2004; **29**: 185-8.
8. Bosenberg AT, Raw R, Boezaart AP: Surface mapping of peripheral nerves in children with a nerve stimulator. *Paediatr. Anaesth* 2002; **12**: 398-403.
9. Chan V: Advances in regional anaesthesia and pain management. *Can.J Anaesth.* 1998; 45: R49-63
10. Marhofer P, Greher M, Kapral S. Ultrasound guidance in regional anaesthesia. *Br J Anaesth* 2005; **94**: 7-17.
11. Tsui B, Suresh S. Ultrasound imaging for regional anaesthesia in infants, children, and adolescents: a review of current literature and its application in the practice of extremity and trunk blocks. *Anesthesiology* 2010; **112**: 473-92.
12. Marhofer P, Willschke H, Kettner S. Ultrasound-guided upper extremity blocks-tips and tricks to improve the clinical practice. *Pediatr Anesth* 2012; **22**: 65-71.
13. Sites BD, Chan VW, Neal JM et al. The American Society of Regional Anaesthesia and Pain Medicine and the European Society of Regional Anaesthesia and Pain Therapy joint committee recommendations for education and training in ultrasound-guided regional anaesthesia. *Reg Anesth Pain Med* 2010; **35**: S74-80.
14. Tobias JD. Brachial plexus anaesthesia in children. *Paed Anaesth* 2001; **11**: 265.
15. Carre P, Joly A, Field BC, et al. Axillary block in children: single or multiple injection? *Paed Anaesth* 2000; **10**: 35-9.
16. Cramer KE, Glasson S, Mencia G, Green NE: Reduction of forearm fractures in children using axillary block anesthesia. *J.Orthop. Trauma* 1995; **9**: 407-10.
17. Fisher WJ, Bingham RM, Hall R. Axillary brachial plexus blocks for perioperative analgesia in 250 children. *Paediatr Anaesth* 1999; **9**: 435-8.
18. Kilka HG, Geiger P, Mehrkens HH: Infraclavicular vertical brachial plexus blockade. A new method for anesthesia of the upper extremity. An anatomical and clinical study *Anaesthesist* 1995; **44**: 339-44.
19. Dadure C, Raux O, Troncin R, Rochette A, Capdevila X: Continuous infraclavicular brachial plexus block for acute pain management in children. *Anesth Analg.* 2003; **97**: 691-3.
20. Fleischman E, Marhofer P, Greher M et al. Brachial plexus anaesthesia in children: lateral infraclavicular vs axillary approach. *Paediatr Anaesth* 2003; **13**:103-8.
21. Marhofer P, Sitzwohl C, Greher M et al. Ultrasound guidance for infraclavicular brachial plexus anaesthesia in children. *Anaesthesia* 2004; **59**: 642-46.
22. Dalens B, Vanneuville G, Tanguy A. A new parascalene approach to the brachial plexus in children: comparison with the supraclavicular approach. *Anesth Analg* 1987; **66**: 1264-71
23. Tobias JD. Regional anaesthesia of the lower extremity in infants and children. *Paediatr Anaesth* 2003; **13**:152-63.
24. McLeod DH, Wong DH, Claridge RJ, Merrick PM: Lateral popliteal sciatic nerve block compared with subcutaneous infiltration for analgesia following foot surgery. *Can.J.Anaesth.* 1994; **41**: 673-6.
25. Tobias JD, Mencia GA. Popliteal fossa block for postoperative analgesia after foot surgery in infants and children. *J Pediatr Ortho* 1999; **19**: 511-4.
26. Dalens B, Tanguy A, Vanneuville G. Lumbar plexus block in children: a comparison of two procedures in 50 patients. *Anesth Analg* 1988; **67**: 750-8.
27. Walker BJ, Flack SH, Bosenberg AT. Predicting lumbar plexus depth in children and adolescents. *Anesth Analg.* 2011; **112**: 661-5.
28. Ronchi L, Rosenbaum D, Athouel A, et al. Femoral nerve blockade in children using bupivacaine. *Anesthesiology* 1989; **70**: 622-4.
29. Tobias JD: Continuous femoral nerve block to provide analgesia following femur fracture in a paediatric ICU population. *Anaesth Intensive Care* 1994; **22**: 616-8.
30. Reuben SS, Sklar J: Pain management in patients who undergo outpatient arthroscopic surgery of the knee. *J Bone Joint Surg Am.* 2000; 82-A: 1754-66.
31. Dalens B, Vanneuville G, Tanguy A. Comparison of the fascia iliaca compartment block with the 3-in-1 block in children. *Anesth Analg* 1989; **69**: 705-13.
32. Konrad C, Johr M. Blockade of the sciatic nerve in the popliteal fossa: a system for standardization in children. *Anesth Analg* 1998; **87**: 1256-8.
33. Dalens B, Tanguy A, Vanneuville G. Sciatic nerve blocks in children: comparison of the posterior, anterior, and lateral approaches in 180 pediatric patients. *Anesth Analg* 1990; **70**: 131-7.
34. Sukhani R, Candido KD, Doty R, et al. Infragluteal-parabiceps sciatic nerve block: an evaluation of a novel approach using a single-injection technique. *Anesth Analg.* 2003; **96**: 868-73.
35. Raj PP, Parks RI, Watson TD, et al. A new single-position supine approach to sciatic-femoral nerve block. *Anesth Analg* 1975; **54**: 489-94.

36. Kempthorne PM, Brown TCK. Nerve blocks around the knee in children. *Anaesth Intens Care* 1984; **12**: 14-17.
37. Tobias JD, Mencia GA. Popliteal fossa block for postoperative analgesia after foot surgery in infants and children. *J Pediatr Ortho* 1999; **19**: 511.
38. Vloka JD, Hadzik A, Lesser JB, et al. A common epineural sheath for the nerves in the popliteal fossa and its possible implications for sciatic nerve block. *Anesth Analg* 1997; **84**: 387-90.
39. Schwemmer U, Markus CK, Greim CA, Brederlau J, Trautner H, Roewer N: Sonographic imaging of the sciatic nerve and its division in the popliteal fossa in children. *Pediatr. Anesth* 2004; **14**:1005-8.
40. terRahe CT, Suresh S: Popliteal fossa block: Lateral approach to the sciatic nerve. *Tech in Reg Anesth Pain Management* 2002; **6**:141-3.
41. Neal JM, Bernards CM, Butterworth JF et al ASRA Practice Advisory on Local Anesthetic Systemic Toxicity. *Reg Anesth Pain Med* 2010; **35**: 152-61.
42. Harclerode Z, Michael S. Axillary brachial plexus block landmark techniques. *Tutorial of the Week* **165** (January 2010)

Update in Anaesthesia

Paediatric spinal anaesthesia

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EDITORIAL COMMENT

This article correctly suggests that the resurgence of interest in spinal anaesthetic techniques in neonates and young infants was driven by the reduced risk of postoperative apnoea. More recently, concerns have arisen about the long term effects on neurodevelopment after exposure to general anaesthesia due to evidence from a broad base of literature in animals. Although the current human evidence is not conclusive, a large paediatric trial is underway and will begin to report in 2014 as this issue goes to press. The GAS (General Anaesthesia vs Spinal 2015) study will look at the differential rates of postoperative apnoea and examine neurodevelopment up to age 5 years, the final reports are due in 2018. Spinal anaesthesia is currently the mainstay of practice for young infants in many low resource environments. It could be that this pattern of practice may be adopted throughout many more healthcare systems in years to come.

INTRODUCTION

Spinal anaesthesia consists of inserting a spinal needle into the subarachnoid space and, when a free flow of cerebrospinal fluid (CSF) is obtained, injection of a solution of local anaesthetic directly into the CSF.

Spinal anaesthesia (SA) was first described in children in 1909¹ but did not become part of routine practice until the 1980's when regional anaesthesia increased in popularity. The particular advantage suggested for SA in children was the avoidance of general anaesthesia (GA) in those at risk of postoperative apnoea. Several studies demonstrated that SA had a particular role in high-risk former preterm neonates undergoing inguinal herniorrhaphy.²

APPLICATIONS OF SPINAL ANAESTHESIA

SA remains popular for ex-premature infants, specifically those undergoing inguinal herniorrhaphy. These patients often have a history of apnoea of prematurity, bronchopulmonary dysplasia and chronic lung disease. The incidence of postoperative apnoeas correlates with gestational age at birth, the post-conceptual age at surgery, weight, anaemia and

a history of apnoeas. General anaesthesia increases the risk of apnoea and bradycardia, and ex-premature infants remain at risk until after 60 weeks post-conception.^{3,4}

Outside the neonatal period, SA has been used for general surgery (rectal biopsy, incision of rectal abscess), urological surgery (orchidopexy, circumcision), lower limb orthopaedic surgery,⁵ and may be of particular use in developing countries as an alternative to general anaesthesia.

SA has also been suggested for patients in whom GA may pose a significant risk such as those with facial dysmorphism and difficult intubation, muscular dystrophy, family history of malignant hyperthermia or a full stomach with aspiration risk.⁵

SA has also been described in combination with GA in children undergoing complex surgery. For instance, preoperative morphine SA combined with GA for scoliosis surgery is associated with reduced blood loss and better pain control.^{6,7} SA has been used with GA during cardiopulmonary bypass in neonates to blunt the stress response, protect hemodynamic status and reduce perioperative morbidity and mortality,^{4,9} although its use in this situation is not common. SA has also been described for use in chronic pain management.^{4,8}

CONTRAINDICATIONS TO SA

There are a number of specific contraindications to SA in children that are listed below:

- Coagulation abnormalities
- Systemic sepsis or local infection at the puncture point
- Uncorrected hypovolaemia
- Parental refusal or an uncooperative child
- Neurological abnormalities such as spina bifida, increased intracranial pressure
- Procedures lasting more than 90 minutes.

ANATOMICAL CONSIDERATIONS

A line connecting the top of the iliac crests crosses the

Summary

Spinal anaesthesia provides a good alternative to general anaesthesia in newborns with increased anaesthesia-related risk, and for infants undergoing lower abdominal or lower extremity surgery during the first 6 months of life.

It is most successful as a single shot technique, limited to surgery lasting less than ninety minutes.

Spinal anaesthesia in children requires the technical skills of experienced anaesthesia providers.

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spinal axis at the L5-S1 level in neonates and infants up to one year of age and at the L4-L5 level in older children.⁵ The spinal cord ends approximately at L3 level at birth and at L1-L2 level in children over one year old.

The distance between the skin and the subarachnoid space is influenced by age – from 10 to 15mm in newborns.¹⁰ The distance between skin and subarachnoid space can be related to height or weight using the formulae:

$$\text{Distance from skin to subarachnoid space (cm)} = 0.03 \times \text{height (cm)}$$

$$\text{Distance from skin to subarachnoid space (mm)} = [2 \times \text{weight (kg)}] + 7(\text{mm})^{11}$$

The subarachnoid space in newborns is very narrow (6 to 8mm) and successful lumbar puncture in this population requires great precision and avoidance of lateral deviation.

Cerebrospinal fluid is a clear body fluid that occupies the subarachnoid space and the ventricular system of the brain and spinal cord. Cerebrospinal fluid volume at different periods of life is shown in Table 1.

Table 1. CSF volume in children

	Cerebrospinal fluid volume (ml.kg ⁻¹)
Neonates	10
Infants less than 15kg	4
Young children	3
Adolescent /Adult	1.5 - 2

The volume of distribution of drugs injected into the subarachnoid space is higher in infants and neonates than in adults and consequently the injected dose is relatively greater in infants and neonates.

PHYSIOLOGICAL EFFECTS OF SPINAL ANESTHESIA

Hemodynamic consequences of SA

Cardiovascular changes related to the SA are less common in children than in adults. In children under 5 years of age, minimal changes in heart rate and blood pressure have been reported.^{5,12,13} In older patients (>8 years old), the sympathetic block can induce bradycardia or hypotension. A few studies of SA in newborns have noted hypotension ten minutes after injection of the local anaesthetic. Cardiovascular changes due to spinal block are generally short lasting and respond to a bolus of intravenous fluid (10ml.kg⁻¹).¹⁴ Cardiovascular stability in infants undergoing SA is probably related to smaller venous capacitance in the lower limbs leading to less blood pooling, and to relative immaturity of the sympathetic nervous system resulting in less dependence on vasomotor tone to maintain blood pressure.

Respiratory effects of SA

Respiratory effects of SA are generally seen in association with high motor block above T6.⁵ Children with severe chronic lung disease should receive supplemental oxygen or Continuous Positive Airway Pressure (CPAP) during SA.

TECHNIQUE OF SA IN CHILDREN

Preoperative preparation

The technique should be explained fully to the parents (and child if appropriate), with a description of risks and benefits. Informed consent should be obtained.

A full blood count including platelet count and coagulation screen (prothrombin time, PT; activated partial thromboplastin time, APTT) may be performed preoperatively where clinically indicated. Blood tests are not usually required for a routine herniotomy, the most common indication for infant spinal anaesthesia.

The child should be fasted as for GA (4 to 6 hours for milk and 2 hours for clear liquid). If possible, topical anaesthesia is used by application of EMLA (Eutectic Mixture of Local Anaesthetics) or Ametop on the lumbar area, 60 to 90 minutes prior to SA. Premedication with oral or rectal atropine (20mcg.kg⁻¹) will vary with institutional preferences, but you should consider using it in all ex-premature neonates.

Operative management

In the operating room, routine monitoring and standard intravenous infusion are started. Some anaesthesiologists have suggested placing the intravenous cannula in an anaesthetized lower extremity after performing the subarachnoid block. We advise placing it prior to SA puncture. Although cardiopulmonary complications are unlikely following SA, they are possible and the presence of a cannula will allow more rapid intervention, particularly in environments where trained anaesthetic assistants are not universally available

There should be an assistant for the anaesthetist to help with preparation of the equipment, positioning and holding the child during insertion of the SA. All drugs and equipment should be prepared and checked prior to starting. Full barrier aseptic technique

Table 2. Dose of local anaesthetic for SA in children

Weight	< 5kg	5 to 15kg	> 15kg
Isobaric or hyperbaric bupivacaine 0.5%	1mg.kg ⁻¹ (0.2ml.kg ⁻¹)	0.4mg.kg ⁻¹ (0.08ml.kg ⁻¹)	0.3mg.kg ⁻¹ (0.06ml.kg ⁻¹)
Isobaric or hyperbaric tetracaine 0.5 %		0.4mg.kg ⁻¹ (0.08ml.kg ⁻¹)	0.3mg.kg ⁻¹ (0.06ml.kg ⁻¹)



Figure 1. Lateral position to perform SA in 4kg newborn

should be used, with a sterile work surface for equipment. The operator should use sterile gloves, gown and mask and the patient's skin should be cleaned with an alcoholic solution such as 0.5% or 2% chlorhexidine (+/- iodine). The skin should be allowed to dry and a sterile sheet should be placed over the child with a hole to reveal the field. The dose of local anaesthetic solution is calculated according to the weight of the child and is shown in Table 2;⁵ the drugs should be drawn into a 1-2ml syringe as appropriate and placed on the sterile work surface in preparation for use.

Both the sitting or lateral decubitus position have been described for lumbar puncture.^{4,5} We have great experience of the lateral position for awake neonates or infants but careful attention must be directed at maintaining patency of the airway which may be compromised with overzealous positioning (Figure 1). The lateral position may be easier than the sitting position for older patients for whom intravenous sedation with a benzodiazepine such as midazolam may be indicated.

Lumbar puncture is performed at L3-L4 or L4-L5 level. Various sizes and lengths of needles are available depending on the child's age. We use a 25G or 26G needle with stylet for neonates and infants (Figure 2). Using a needle without a stylet is not recommended since epithelial tissue can be deposited in the intrathecal space and may cause dermoid tumours of the neural axis.

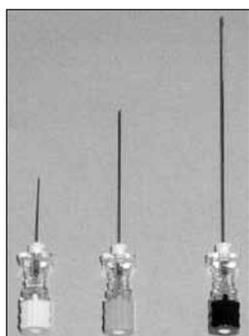


Figure 2. Different types of SA needles

A free flow of cerebrospinal fluid should be obtained when the spinal needle is advanced into the intrathecal space. The local anaesthetic syringe is attached and the anaesthetic solution is injected over 30 seconds (Figure 3). The legs should not be lifted after the spinal injection has been administered, otherwise an excessively high block will develop.



Figure 3. Lumbar puncture and LA injection with 1ml syringe

SA may produce a degree of sedation in newborns and infants so additional intravenous sedation is not required.¹⁵ Intravenous sedation should be avoided if at all possible in infants at risk of apnoea. We find that a dummy dipped in sucrose or honey will help to settle these infants.

Postoperative care

In our hospital, children are discharged from the post anaesthesia care unit when the block disappears, i.e. free lower limb movement returns. Children are allowed to feed on demand, provided there are no surgical restrictions. All infants younger than 60 weeks post conception are monitored on the ward for 24 hours after SA.

COMPLICATIONS OF SA

There are a number of potential complications of SA that are listed below:

- Potential traumatic puncture with spinal damage. Careful technique with the appropriate equipment and a trained assistant is essential
- Respiratory (+/-cardiovascular) insufficiency due to high SA or secondary to intravenous sedation. Resuscitation measures must be taken (ABC) - tracheal intubation and volume resuscitation may be required.
- Convulsions due to overdose of local anaesthetic. All doses should be calculated carefully and checked with another practitioner.
- Post dural puncture headache. This has been reported in children >8 years old, but the incidence in younger children is unknown, in part since headaches in infants and young children are difficult to assess.^{5,16}
- Infectious complications such as meningitis. The incidence of meningitis is very low – careful aseptic technique must be used at all times and multidose ampoules of local anaesthetic must never be used. We suggest repeating lumbar puncture in patients who develop fever after SA.^{4,5}
- Neurological injury due to injection of incorrect solutions. Great care must be taken at all times in preparation and checking of drugs.

CONCLUSION

In our experience, the incidence of serious complications associated with SA is very low, even in small premature infants. We think that this technique provides a good alternative to general anaesthesia in newborns with increased anaesthesia-related risk and for infants undergoing lower abdominal or lower extremity surgery during the first 6 months of life. SA may be used to avoid GA in patients outside the neonatal period, if needed combined with intravenous sedation. SA is most successful as a single shot technique, limited to surgery lasting less than 90 minutes. SA in children requires the technical skills of experienced anaesthesia providers. Neonates and infants are at high risk of complications during surgery, irrespective of the type of anaesthesia, and the presence of clinician trained in paediatric anaesthesiology is mandated.

REFERENCES

1. Troncin R, Dadure C. Paediatric spinal anaesthesia. Update in Anaesthesia (2009);25(1):22-24 Available from: http://www.wfsahq.org/components/com_virtual_library/media/b11bcbdd8ca5b2fcdd6762e26503838b0-bd58f3feb0a0db474a5ea3c41f4f0431-Paediatric-Spinal-Anaesthesia.pdf
2. Tyrell-Gray H. A study of spinal anaesthesia in children and infants. *Lancet* 1909; **2**: 913-7.
3. Abajian JC, Mellish RW, Browne AF, Perkins FM, Lambert DH, Mazuzan JE. Spinal anesthesia for surgery in the high risk infant. *Anesth Analg* 1984; **63**: 359-62.
4. Welborn LG, Rice LJ, Hannallah RS, Broadman LM, Ruttimann VE, Fink R. Post-operative apnea in former infants: prospective comparison of spinal and general anesthesia. *Anesthesiology* 1990; **72**: 838-42.
5. Tobias JD. Spinal anaesthesia in infants and children. *Paediatric Anaesth* 2000; **10**: 5-16.

5. Dalens B, Veyckemans F. *Traité d'Anesthésie Loco-Régionale, de la naissance à l'âge adulte*. Montpellier: *Sauramps Médical* 2008; 463-512.
6. Dalens B, Tanguy A. Intrathecal morphine for spinal fusion in children. *Spine* 1988; **13**: 494-7.
7. Gall O, Aubineau JV, Bernière J, Desjeux L, Murat I. Analgesic effect of low dose intrathecal morphine after spinal infusion in children. *Anesthesiology* 2001; **94**: 447-52.
8. Berde C, Sethna N, Conrad LS, Hershenson MB. Subarachnoid bupivacaine analgesia for seven months for a patient with a spinal cord tumour. *Anesthesiology* 1990; **72**: 1094-6.
9. Kowalewski R, MacAdams C, Froelich J, Neil S, Maitland A. Anesthesia supplemented with subarachnoid bupivacaine and morphine for coronary artery bypass surgery in a child with Kawasaki disease. *J cardiothor Vasc Anesth* 1996; **10**: 243-6.
10. Blaise GA, Roy WL. Spinal anaesthesia for minor paediatric surgery. *Canadian Journal of Anaesthesia* 1986; **32**: 227-30.
11. Arthurs, Murray M, Zubier M, Tooley J, Kelsall W. Ultrasonographic determination of neonatal spinal canal depth. *Archives of Disease in Childhood - Fetal and Neonatal* Edition 2008; **93**: 451-4.
12. Dohi S, Naito H, Takahashi T. Age-related changes in blood pressure and duration of motor blockade in spinal anesthesia. *Anesthesiology* 1979; **50**: 319-23.
13. Oberlander TF, Berde CB, Lam KH, Rappaport LA, Saul JP. Infants tolerate spinal anesthesia with minimal overall autonomic changes: analysis of heart rate variability in former preterm infants undergoing hernia repair. *Anesth Analg* 1995; **80**: 20-7.
14. Mahe V, Ecoffey C. Spinal anesthesia with isobaric bupivacaine in infants. *Anesthesiology* 1988; **68**: 601-3.
15. Disma N, Tuo P, Astuto M, Davidson AJ. Depth of sedation using cerebral index in infants undergoing spinal anaesthesia. *Pediatric Anesthesia* 2009; **19**: 133-7.
16. Wee LH, Lam F, Cranston AJ. The incidence of post dural puncture headache in children. *Anaesthesia* 1996; **51**: 1164-6.

Update in Anaesthesia

Paediatric difficult airway management

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Summary

Unexpected difficult airways in paediatric practice are rare. Many problems can be prevented by routine pre-operative airway assessment, pre-oxygenation, and preparation of equipment. A simple step-wise approach to management improves outcome. Anaesthetists have a responsibility to be familiar with airway algorithms and make pragmatic modifications to account for available resources.

INTRODUCTION

Airway management in children is generally straightforward in experienced hands. Problems are more common for the non-paediatric anaesthetist, and are a major cause of anaesthesia-related morbidity and mortality. Genuine 'difficult airways' are rare in children compared to adults and many are predictable. However, differences in adult and paediatric physiology mean irreversible hypoxic damage occurs more quickly in children if there is an airway problem. Simple step-wise strategies are essential. Many guidelines exist for the management of difficult airways in adults, but there are few specifically designed for use in children.

The aim of this article is to outline the basic principles of paediatric airway assessment and to discuss the management of unexpected and expected difficult paediatric airways.

Evidence to support best practice is difficult to obtain for unpredictable events such as management of the paediatric difficult airway, and there is a lack of high quality data. Many new devices and techniques are available, but most are evaluated in healthy children or simulated 'difficult' situations. Due to this lack of evidence, guidelines are often based on a consensus of expert opinion, which may have a bias against newer devices and techniques, or indeed bias towards the latest technique that has gained popularity. This review takes a pragmatic and cautious approach in applying existing guidelines to settings where experts and a range of technology are not always available.

BACKGROUND

Management of the difficult airway can be divided into three critical areas:

1. Difficult mask ventilation
2. Difficult tracheal intubation
3. Can't intubate and can't ventilate (CICV).

The incidence of difficult airways in children is unknown. The incidence of impossible mask ventilation is reported as 0.15%, and is more frequently encountered by inexperienced paediatric anaesthetists. Difficult intubation ranges from 0.05%, rising to 0.57% in children less than one year of age.¹ Difficult

intubation is more common in children with cleft lip and palate (4.7%) and cardiac abnormalities (1.25%), most likely related to associated syndromes or limited cardiac reserve.

An audit of difficult airway management in the UK in 2001 (the 4th National Audit Project, NAP 4), prospectively measured major airway complications in almost 115,000 patients undergoing anaesthesia.² Children comprised a small proportion of the total population, and complications were rare (only 7-8% of total complications).

Common contributing factors to bad outcomes were:

- Poor airway assessment
- Poor planning
- 'Failure to plan for failure'
- Repeated attempts at intubations
- Lack of monitoring (oxygen saturation and capnography)
- Slow response to hypoxia resulting in bradycardia leading to cardiac arrest
- Failure to use devices such as the laryngeal mask airway (LMA) when faced with a difficult intubation.

One of the key findings in NAP4 was the 'failure to plan for failure'. Airway management plans should always include a back-up plan to use if the first plan fails. Whenever unexpected difficulties occur, seek experienced help immediately. Another key finding of NAP4 was that repeated attempts at intubation can cause severe airway oedema in children and worsen the situation, hence their recommendation, 'a change of approach is required, not repeated use of a technique that has already failed'.

Many countries have adult guidelines for management each of difficult airways, but few have child specific guidelines. The Association of Paediatric Anaesthetists of Great Britain and Ireland (APA) published paediatric guidelines in 2012, which are shown in Figures 1-3 and form the basis for management of the unexpected difficult airway discussed here.³

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AIRWAY ASSESSMENT

Proper airway assessment, proper planning for airway management, and the use of monitoring are essential basic principles for safe anaesthesia in children.

Airway assessment may be considered in two parts:

Will facemask ventilation be difficult?

- A tumour or abnormal face shape may prevent the facemask from sealing easily over the face.
- Syndromes associated with midface hypoplasia
- Children with severe obstructive sleep apnoea (e.g. tonsillar hypertrophy).

Will intubation be difficult?

Factors that may predict difficult intubation in children include:

- Mandibular hypoplasia e.g. syndromes such as Pierre Robin
- Poor mouth opening
- Obstructive sleep apnoea
- Stridor
- Syndromes associated with facial asymmetry. Note ear abnormalities are often associated (e.g. Goldenhar syndrome).

Various tests and scoring systems have been suggested for use in adults. Many have a very poor sensitivity and/or specificity and are not validated in children. However, assessment of the following is essential. The anaesthetist may need some ingenuity to achieve these assessments in a small uncooperative child!

- Mouth opening
- Range of neck movement
- Mandibular hypoplasia - micrognathia makes intubation difficult. Assess the airway by observing the child in side view rather than from the front.
- Mandibular hyperplasia - ameloblastoma may cause jaw protrusion and can make laryngoscopy and intubation difficult
- Inspection of the oral cavity (e.g. for intraoral masses).

The Mallampati score can be used for older children who are cooperative. Even though there is no validated scoring system in infants and young children, the anaesthetist must still make a risk assessment, and decide on the anticipated difficulty of intubation. This airway assessment must be documented in the anaesthetic record.⁴

PLAN FOR AIRWAY MANAGEMENT

After airway assessment, a structured plan for airway management is required before induction of anaesthesia. The plan must consider:

- Choice of airway e.g. facemask, supraglottic airway device or tracheal tube
- Mode of ventilation e.g. spontaneous ventilation or positive pressure
- Monitoring e.g. pulse oximeter (minimum); end tidal carbon dioxide.

Due to a lower functional residual capacity (FRC) and higher metabolic rate, oxygen saturation falls much faster in infants and young children than adults. Preoxygenation before induction of anaesthesia establishes a reservoir of oxygen in the lungs by displacing nitrogen. This means a patient can remain oxygenated for longer than otherwise expected, which gives more time to address unexpected airway problems. Therefore preoxygenation is an important part of the airway management plan and should form part of normal anaesthetic practice wherever possible, even in children.

THE UNEXPECTED DIFFICULT AIRWAY

Problems with airway management may be due:

1. Difficult mask ventilation
2. Difficult tracheal intubation
3. Can't intubate and can't ventilate (CICV).

The first step is to administer 100% oxygen and call for help. Another pair of hands is always useful.

The next step is to consider – is this a problem with the equipment or the patient? All equipment should be checked prior to induction of anaesthesia to minimise the chance of equipment failure.

1. Difficult mask ventilation

A simple algorithm for the management of difficult mask ventilation is given in Figure 1: Difficult mask ventilation algorithm. <http://www.apagbi.org.uk/sites/default/files/images/APA1-DiffMaskVent-FINAL.pdf>

Difficult mask ventilation – equipment problems

Equipment failure should be excluded quickly – check the mask, circuit, and oxygen supply. Always have a self-inflating bag available in case of equipment problems.

Difficult mask ventilation – patient factors

These can be divided into anatomical or functional problems.

Anatomical problems associated with difficult mask ventilation may be due to poor head positioning, large adenoids/tonsils, or due to airway obstruction from cricoid pressure (if used).

Functional problems may arise in the upper or lower airways. Upper airway obstruction may be due to inadequate depth of anaesthesia and laryngospasm; lower airway problems include inflation of the stomach (very common in infants), bronchospasm or chest wall rigidity (rare).

Management of difficult mask ventilation:

- Adjust the head position – does the child need a head roll (or should the head roll be removed)
- Use simple airway opening manoeuvres (chin lift, jaw thrust)
- Apply positive end expiratory pressure (PEEP)
- Adjust cricoid pressure if it has been used
- Insert an oropharyngeal airway (if the patient is deep enough)
- Increase depth of anaesthesia
- Ventilate using a two-person technique (one holding the mask with two hands, the other ventilating by squeezing the bag)

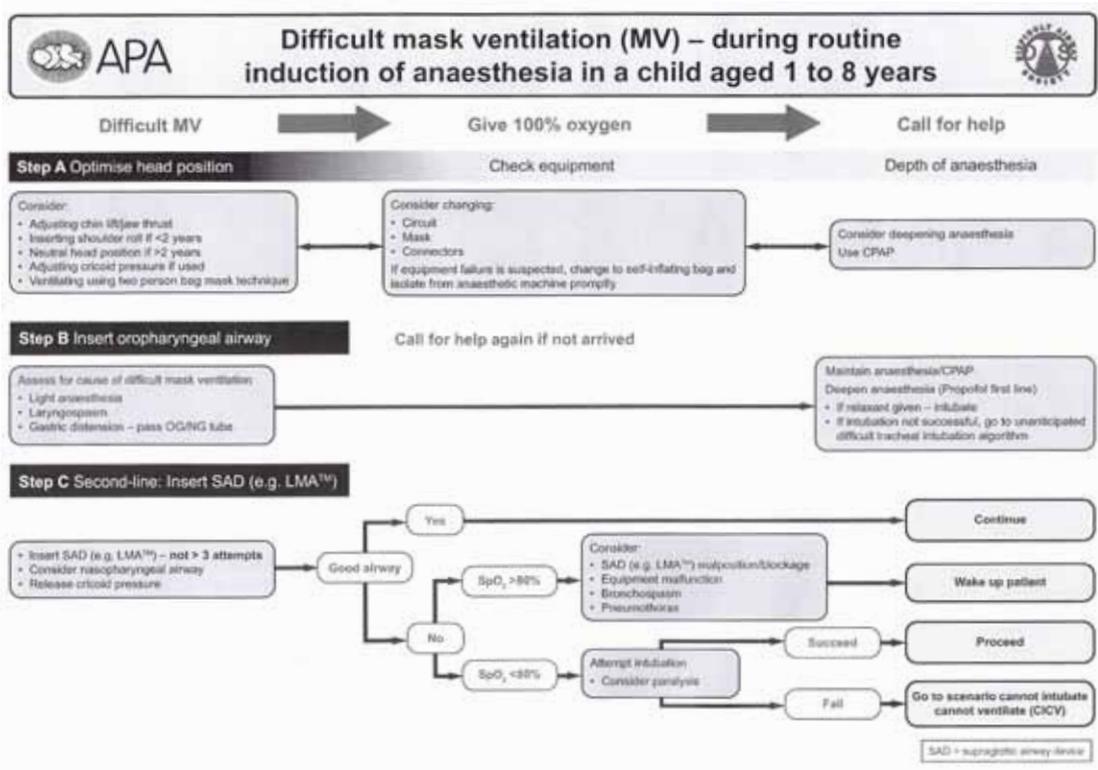


Figure 1: Difficult mask ventilation algorithm. Reproduced with kind permission of Association of Paediatric Anaesthetists from: <http://www.apagbi.org.uk/sites/default/files/images/APA1-DiffMaskVent-FINAL.pdf>

- Pass a nasogastric tube to deflate the stomach.

If mask ventilation is impossible despite all the above measures or the child's oxygen saturation begins to fall:

EITHER insert an LMA (if available),

OR deepen anaesthesia, attempt to visualise the vocal cords and intubate the trachea.

There is no randomised controlled trial to assess which is the best response, but insertion of an LMA is recommended first, and then intubation (Figure 1).

If oxygenation and ventilation is satisfactory through the LMA or tracheal tube then it is safe to proceed with surgery.

If in doubt, wake the child up.

2. Unexpected difficult tracheal intubation

A simple algorithm for the management of unexpected difficult tracheal intubation is given in Figure 2: Difficult tracheal intubation algorithm. <http://www.apagbi.org.uk/sites/default/files/images/APA2-UnantDiffTracInt-FINAL.pdf>

The key point is, if tracheal intubation fails, DO NOT simply repeat what has just failed. Multiple attempts at intubation may traumatise the airway and will cause airway oedema, which may make the child impossible to intubate. Intubation attempts must be limited to a maximum of three or four (Figure 2).

If the first intubation attempt fails, it is essential to make changes that improve the chance of successful intubation. These may include:

- Change of personnel (a more senior anaesthetist);
- Change of position
- Change of equipment.

Visualisation of the larynx and successful tracheal intubation are improved by:

- Proper positioning of the child,
- External laryngeal manipulation
- Adequate depth of anaesthesia and adequate muscle paralysis (if this has been used).

Simple aids such as a bougie or stylet may make intubation straightforward even when the view of the larynx is poor. An alternate laryngoscope may also be used if available and if the operator is familiar with its use.

Straight bladed laryngoscopes are traditionally used in children under one year old, but may be useful in older children, or in patients with relative macroglossia. They can be used with a paraglossal or retromolar technique. McCoy levering laryngoscopes are also available for paediatric use, based on a Seward blade (sizes 1 and 2) and may improve the view of the larynx, particularly if the view is obstructed by a large epiglottis.

In addition to straight bladed and McCoy laryngoscopes, new alternate laryngoscopes have been developed recently (see table 1). High quality evidence supporting efficacy is largely absent in the life-threatening scenario of unexpected failed intubation. Firm recommendations cannot be made so many algorithms suggest alternate laryngoscopes/techniques 'should be considered'.

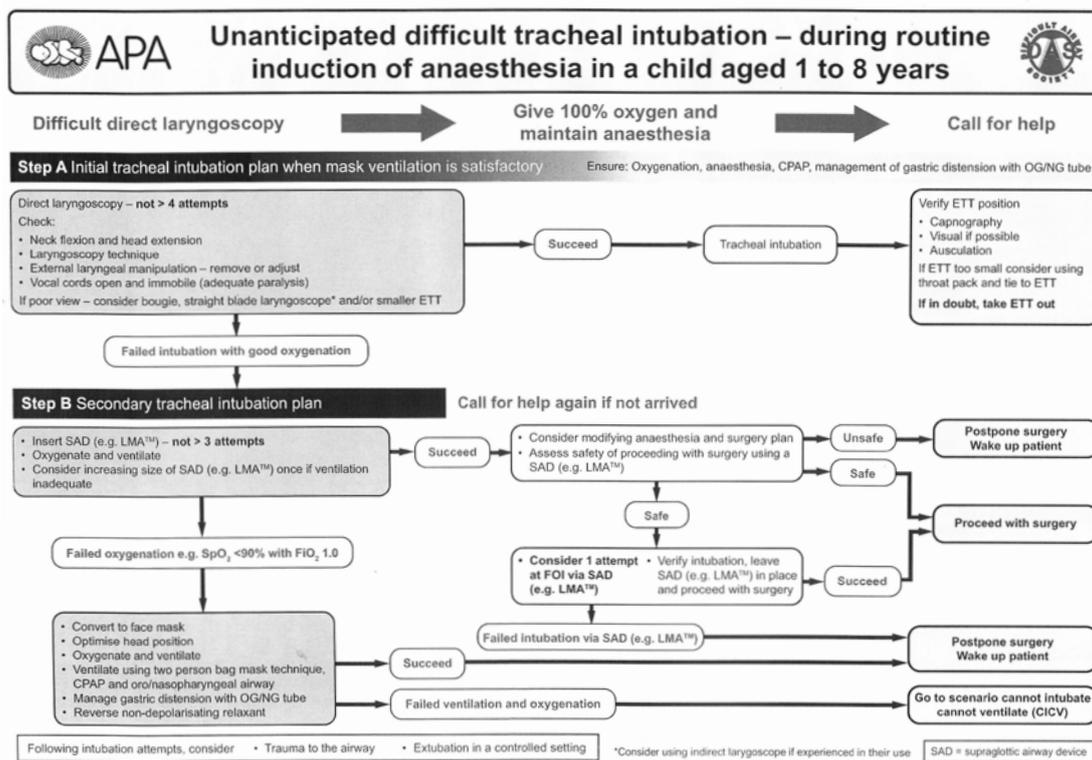


Figure 2: Difficult tracheal intubation algorithm. Reproduced with kind permission of Association of Paediatric Anaesthetists from: <http://www.apagbi.org.uk/sites/default/files/images/APA2-UnantDiffTraclnt-FINAL.pdf>

Table 1. Classification of rigid indirect laryngoscopes

Classification	Description of technique	Examples
'Non-guided' devices	Provide an indirect view of larynx but require direction of tracheal tube towards larynx.	Bullard laryngoscope Glidescope Storz DCI Videolaryngoscope Storz C-MAC laryngoscope
'Guided' devices	Provide indirect view and act as a conduit for passage of tracheal tube.	Airtraq
Optical stylets	Provide indirect view via rigid or semi-rigid stylet, with a 'loaded' tracheal tube for railroading.	Bonfils and Brambrink Shikani Lightwand

Alternate techniques

Traditional laryngoscopes (curved, straight or McCoy levering blades) give a direct view of the larynx. Alternate techniques use an indirect approach with flexible or rigid equipment.

- Flexible indirect laryngoscopy, in the form of fiberoptic intubation, is the established 'gold standard' for the management of the predicted difficult airway in adults (see below).
- New rigid indirect laryngoscopes are available, including in paediatric sizes. Rigid indirect laryngoscopy has a place in the unexpected difficult tracheal intubation algorithm. The choice of device depends on local availability and expertise.

If visualising the larynx is impossible, then an LMA should be inserted. LMAs provide a clear airway in the vast majority of children. This allows delivery of ventilation, oxygenation and anaesthetic gases with a lower risk of gastric insufflation. The LMA may also be used as a

conduit for fibreoptic intubation (FOI) where necessary, or in older children, the specifically designed intubating LMA, may be used.

If LMA insertion fails, then oxygenation and ventilation must be provided by mask ventilation.

3. Cannot intubate, cannot ventilate (CICV) - 'rescue techniques'

A simple algorithm for the management of 'cannot intubate, cannot ventilate' is given in Figure 3 : Can't intubate can't ventilate algorithm. <http://www.apagbi.org.uk/sites/default/files/images/APA3-CICV-FINAL.pdf>

Rescue techniques for the CICV situation have been extensively researched in the adult literature, but their use in paediatric emergencies is more anecdotal than evidence-based. The choice is between a surgical or needle (cannula) technique for cricothyroidotomy. The technique for cannula cricothyroidotomy is shown in Figure 3, and for surgical

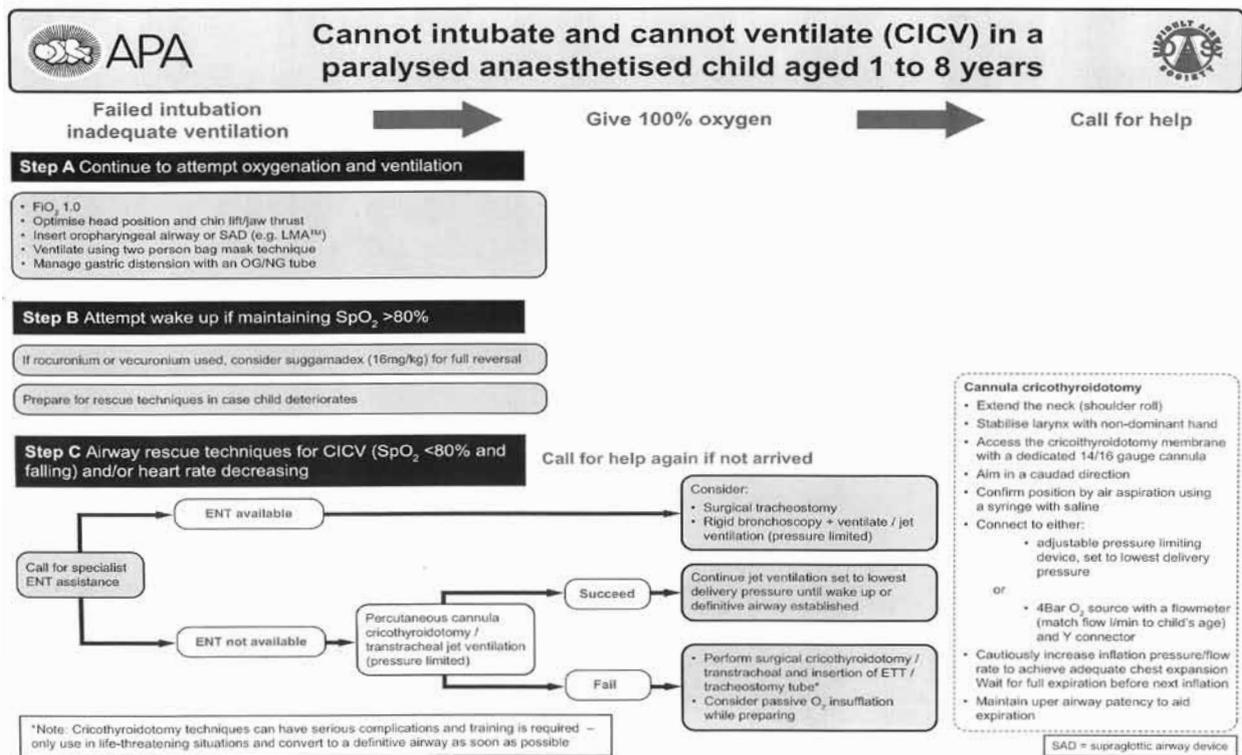


Figure 3: Can't intubate can't ventilate algorithm. Reproduced with kind permission of Association of Paediatric Anaesthetists from: <http://www.apagbi.org.uk/sites/default/files/images/APA3-CICV-FINAL.pdf>

Table 2. Technique for surgical cricothyroidotomy

<ol style="list-style-type: none"> 1. Position the patient so that the neck is fully extended so that the trachea and larynx are pushed forward 2. Locate the cricothyroid membrane and stabilise the trachea 3. With a scalpel blade make a stab incision through the skin and cricothyroid membrane* 4. Insert a tracheal hook or retractor at the lower edge of the incision 5. Pass an appropriately sized tracheal or tracheostomy tube 6. Ventilate patient and assess effectiveness 7. Secure the tube <p><i>Arterial forceps, the scalpel blade and tracheal dilators may be used to dilate the orifice.</i></p>

cricothyroidotomy, in Table 2.

Since there is no randomised controlled trial of one technique versus another, the choice should be determined by local experience and availability of equipment. This includes utilising the surgeon who may be more experienced than the anaesthetist. Adult evidence suggests surgical cricothyroidotomy is preferable, so this is recommended in older children.

The important factor is that at least one technique is actually attempted by someone in the CICV situation when the oxygen saturations are less than 80% and falling and/or the heart rate is decreasing.

The CICV situation is a particular challenge in infants and small children, due to important anatomical differences:

- The trachea is small, elastic, flaccid and mobile, and so prone to collapse during insertion of a transtracheal device.

- The cricothyroid membrane is much smaller, with an average size of only 2.6 x 3mm, smaller than the smallest tracheal tubes.
- It is more difficult to locate the cricothyroid membrane than in adults due to a differing orientation of the hyoid bone and the cricoid and thyroid cartilage. This orientation also increases the chance of laryngeal trauma during cricothyroidotomy.
- It is easier to locate the space between the tracheal rings rather than the cricothyroid membrane.

Together these factors mean it may be more appropriate in infants and small children to perform a surgical tracheostomy.

All 'rescue techniques' have significant potential for complications so should only be performed in life threatening situations. Clearly, all the steps for difficult facemask ventilation should be tried first. If muscle relaxants have been used and can be reversed, wake the child up.

THE EXPECTED DIFFICULT AIRWAY

If the preoperative airway assessment alerts the anaesthetist to expected difficulties in airway management then there are three key questions:

1. Does the anaesthetist have the necessary paediatric airway experience?
2. Does the hospital have the necessary paediatric equipment?
3. Does the relative benefit of the planned surgery outweigh the possible risks of anaesthesia?

If there is any doubt, full discussion should take place with the parents (or carers), child, surgeons and anaesthetists. It may be appropriate for the child to be referred to specialist hospital or wait for a more experienced paediatric anaesthetist to attend.

The anaesthesia plan must be carefully considered, including what to do if tracheal intubation is unsuccessful; will the child be woken up, or will a tracheostomy be necessary. The anaesthesia plan should be communicated clearly to the whole theatre team including surgeons and nursing staff. Difficult airway equipment must be checked and prepared.

The primary plan for management of the expected difficult paediatric airway will likely be one of the following:

1. Laryngoscopy anticipated to be difficult but may be possible: Attempt laryngoscopy and intubation. If fails, consider repositioning and try alternate laryngoscopes if available, or insert LMA and perform fiberoptic intubation (FOI) via LMA.
2. Laryngoscopy predicted to be impossible: Perform nasal FOI or insert LMA and perform FOI via LMA.
3. Laryngoscopy and LMA insertion known to be impossible: perform nasal FOI.
4. Laryngoscopy, LMA insertion and nasal FOI not available or known to be impossible: perform tracheostomy either using inhalational anaesthesia via face mask or intravenous ketamine especially if face mask anaesthesia impossible.

Blind intubation through an LMA is NOT recommended in children due to risk of airway trauma. Attempts at FOI should be limited to two and if unsuccessful, consider waking child, or continue with surgical procedure on an LMA. In situations where LMAs are unavailable, ventilation by face mask is the alternative. If neither LMAs nor FOI are available, the surgeon and anaesthetist need to discuss whether the benefits of surgery outweigh the risk of attempting anaesthesia in a child with a known difficult airway with insufficient equipment to provide safe management. This is a very difficult decision and will depend on the individual merits of each case.

Premedication

The use of sedative premedication in a child with a potential airway problem is controversial. A frightened, screaming child producing lots of secretions and in whom it is difficult to place monitoring, intravenous cannula, and even approach to do an inhalational induction, is also a risk.

Therefore, a small dose of sedative premedication, such as midazolam 0.3-0.5mg.kg⁻¹ is often appropriate. Atropine is useful as an antisialagogue (30-40 micrograms.kg⁻¹ PO or 20 micrograms.kg⁻¹ IM). Peak effect of

atropine is 90 minutes if given PO, 25 minutes if given IM.

Anaesthetic technique

The most important principle in managing the difficult airway in children is to maintain spontaneous ventilation until the airway is secure.

'Awake' techniques require good patient co-operation, which is rarely possible in children. Therefore, the child must be anaesthetised so the choice is between an inhalational or intravenous technique. The variety of airway problems encountered in children means the anaesthetic must be tailored to the individual situation:

- Large extraoral tumours may mean a face mask will not fit the child's face, so an inhalational induction is impossible and IV induction/sedation must be used instead.
- Large intraoral tumours prevent laryngoscopy and the use of an LMA - nasal fiberoptic intubation (FOI) should be used.
- Conditions such as noma (cancrum oris) often cause severe limitation of mouth opening - nasal FOI is likely to be required.
- Other problems such as partial mouth opening, severe retrorathia or bony abnormalities (ameloblastoma) often make laryngoscopy difficult but do permit the insertion of an LMA if laryngoscopy proves impossible.
- Burns contractures causing fixed flexion of the neck may be released prior to intubation using ketamine anaesthesia and with local infiltration.

The variety of clinical conditions mean a one-size-fits all approach is impossible. The best technique will depend on the equipment and expertise available, as well as the nature of the difficult airway.

Inhalational induction, using halothane or sevoflurane in 100% oxygen, is generally recommended. Intravenous access may be established either before or after induction but must occur before airway instrumentation. The general technique is to deepen anaesthesia until laryngoscopy is tolerated or LMA inserted or FOI performed depending on the airway management plan.

If inhalational induction is impossible, small doses of IV induction agent should be given to induce loss of consciousness but still preserving spontaneous ventilation. Propofol 0.5-1mg.kg⁻¹ or ketamine 0.5-1mg.kg⁻¹ should be given and titrated to effect.

If inhalational induction is not possible due to pain, for instance, from an infected facial mass/tumour (rather than because of a large extraoral tumour meaning a face mask will not fit), give a small dose of ketamine, then apply the face mask and deepen anaesthesia by spontaneous inhalation with sevoflurane or halothane. In our experience, this combination provides better conditions for laryngoscopy than when using intravenous ketamine alone.

Nasal fiberoptic intubation – general

- Maintain anaesthesia either with incremental doses of ketamine or inhalational anaesthesia either via a nasal airway in the other nostril connected to the breathing circuit or using a specially designed facemask with a port for insertion of the fiberoptic bronchoscope.

Table 3. Size compatibility of tracheal tubes, bronchoscopes and LMAs

Tracheal tube size	2.5	3.0	3.5	4	4.5	5	6
Will fit through classic LMA size:	1	1	1.5	2	2	2.5	3
Will fit over bronchoscope Outer Diameter:	2.0mm	2.5mm	2.8mm	3.5mm	3.5mm	4.1mm	5mm
Will fit over AEC size:	7F	8F	8F	11F	11F	11F	14F

[Note: different brands of LMA vary in their internal diameter. It is important to determine the compatibility of equipment within your own department.]

- Use a topical vasoconstrictor to prevent bleeding from the nose during FOI, as otherwise this may make intubation impossible. Pseudoephedrine, ephedrine, phenylephrine, oxymetazoline, or nasal packs soaked in 1:10,000 adrenaline may be used, depending on local availability.
- Apply topical lidocaine to the nose and oropharynx. Larger fiberoptic laryngoscopes often have a channel through which local anaesthesia can be injected. Alternatively an epidural catheter can be passed through the suction port (if present) and local anaesthetic injected through this. Be careful not to exceed the maximum dose of lidocaine (3mg.kg⁻¹ i.e. 0.3ml.kg⁻¹ of a 1% solution).

The correct size of tracheal tube is critical to success. Too large a tube will fail and require the bronchoscope to be withdrawn and the procedure repeated. Too small may make subsequent positive pressure ventilation difficult. It is sensible to use a small cuffed tube if available, rather than repeated bronchoscopy.

Fiberoptic intubation through an LMA

There are three main techniques available:

1. Railroad the tracheal tube over the fiberoptic bronchoscope into the trachea
2. Railroad an airway exchange catheter (AEC) over the bronchoscope into the trachea.
3. Pass a soft tip wire through the suction channel of the bronchoscope into the trachea, then pass an AEC or similar over the wire as a guide for the tracheal tube.

The choice of technique depends upon size of the child, the size of the LMA, and the diameter of available bronchoscope (Table 3). Removal of the LMA once the tracheal tube is in situ may be challenging. Options include:

- Leave the LMA in situ
- Use a long tracheal tube (croup tube)
- Fix two tracheal tubes together over the FOB; the LMA may be withdrawn over the tracheal tubes.
- Use an AEC.

Tracheostomy

A tracheostomy should be performed by an experienced practitioner, normally an ENT surgeon. Inhalational anaesthesia or small incremental doses of ketamine (as above) may be given to supplement local infiltration anaesthesia. The child should breathe 100% oxygen by facemask.

DIFFICULT AIRWAY CART

The equipment available in different institutions will vary considerably. It is good practice to organise airway equipment in such a way that

it is readily accessible in an emergency. Many hospitals use a 'difficult airway cart' to do this. This is simply a trolley or cart where all the useful equipment for managing difficult airways is stored according to the step-wise approach to managing a difficult airway.

For example, using the algorithms presented in this review, the difficult airway cart could consist of a series of drawers or boxes containing:

Drawer 1: simple laryngoscopes and airway adjuncts.

Drawer 2: alternative laryngoscopes and LMAs.

Drawer 3: equipment for fiberoptic intubation

Drawer 4: equipment for CICV situations.

Whatever the availability and variety of equipment, the difficult airway cart (or boxes) should always be stored in the same place, close to the operating rooms, and the contents regularly checked. The cart should be physically present in the operating room for any child with an anticipated difficult airway; and can be quickly fetched when faced with an unexpected problem.

CONCLUSION

Unexpected difficult airways in paediatric practice are rare. Many problems can be prevented by routine pre-operative airway assessment, pre-oxygenation, and preparation of equipment. A simple step-wise approach to management improves outcome. Anaesthetists have a responsibility to be familiar with airway algorithms and make pragmatic modifications to account for available resources.

REFERENCES

1. Weiss M, Engelhardt T. Proposal for the management of the unexpected difficult pediatric airway. *Pediatr Anesth* 2010; **20**: 454-64. <http://onlinelibrary.wiley.com/doi/10.1111/j.1460-9592.2010.03284.x/pdf>
2. The 4th National Audit Project of the Royal College of Anaesthetists and the Difficult Airway Society: Major Complications of Airway Management in the United Kingdom. March 2011. <http://www.rcoa.ac.uk/nap4/>
3. The Association of Paediatric Anaesthetists (APA) Guidelines for difficult airway management in children. 2012. <http://www.apagbi.org.uk/publications/apa-guidelines>
4. World Health Organisation. Guidelines for Safe Surgery 2009. http://whqlibdoc.who.int/publications/2009/9789241598552_eng.pdf and <http://www.who.int/patientsafety/safesurgery/en/>
5. Kheterpal S, Martin L, Shanks AM, Tremper KK. Prediction and outcomes of impossible mask ventilation: a review of 50,000 anesthetics. *Anesthesiol* 2009; **110**: 891-7.
6. Holm-Knudsen R. The difficult pediatric airway - a review of new devices for indirect laryngoscopy in children younger than two years of age. *Pediatr Anesth* 2011; **21**: 98-103
7. Doherty JS, Froom SR, Gildersleeve CD. Pediatric laryngoscopes and intubation aids old and new. *Pediatr Anesth* 2009; **19** (Suppl. 1): 30-37 <http://onlinelibrary.wiley.com/doi/10.1111/j.1460-9592.2009.03001.x/pdf>
8. Walker RWM and Ellwood J. The management of difficult intubation in children. *Pediatr Anesth* 2009; **19** (Suppl. 1): 77-87 <http://onlinelibrary.wiley.com/doi/10.1111/j.1460-9592.2009.03014.x/pdf>

Neonatal anaesthesia

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INTRODUCTION

Neonates present a challenge to the anaesthetist. They have unique physiology as they transition from intrauterine to extrauterine life, limited physiological reserve and immature drug handling. The goals of anaesthesia are to provide stable conditions for surgery, minimise physiological disturbance, reduce pain, and support the neonate during the postoperative period. This article will describe general considerations for anaesthesia in term and preterm neonates, and anaesthesia for some specific neonatal conditions.

PREOPERATIVE ASSESSMENT OF THE NEONATE

As for any child undergoing anaesthesia, it is important to take a detailed history and examination, together with relevant investigations to assess the current physiological status and the impact of any associated congenital abnormalities, which may or may not be related to the surgical condition. This helps to plan when best to proceed with the surgery, and the level of postoperative support required.

History

The history should include the gestational age, birth history, current age and weight, and significant peri-natal events such as low APGAR scores, respiratory distress requiring respiratory support, hypoglycaemic episodes, NICU admissions, evidence of sepsis or any antenatal concerns such as maternal illness. The anaesthetist should check whether intramuscular vitamin K has been given to prevent haemorrhagic disease of the newborn. The fasting status should be established if the child is receiving feeds - ideally 2 hours for clear fluids, 4 hours for breast milk, 6 hours for formula feed.

Examination

Examine the child carefully. In particular, it is important to look for signs of respiratory distress (respiratory rate, nasal flare, subcostal recession), and cardiovascular compromise (check heart rate, blood pressure, peripheral perfusion and capillary refill). Check the oxygen saturation – low oxygen saturation may be associated with respiratory disease, or in some cases with cyanotic congenital heart disease.

Investigations

Relevant investigations will be guided by the clinical findings and the underlying condition, although resources may limit investigations that can be performed. They may include the following:

Laboratory investigations:

- Full blood count and haemtocrit
- Blood glucose
- Urea and electrolytes
- Coagulation studies
- Liver function tests and bilirubin
- Capillary blood gas.

Radiological investigations:

- CXR, AXR
- Echocardiogram
- Cranial/spinal/renal ultrasound.

Finally, the anaesthetic plan, including risks, should be discussed with the parent(s) or guardian(s), and consent taken for anaesthesia including regional anaesthesia and blood transfusion if indicated.

DEFINITIONS

- Neonate is aged up to 28 days
- Term neonate is born between 37 to 40 weeks post conception
- Preterm neonate is born at <37 weeks post conception
- Extreme preterm neonate is born <28 weeks post conception
- Low birthweight <2.5kg
- Very low birthweight <1.5kg

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GENERAL PRINCIPLES OF ANAESTHESIA IN NEONATES

It is important to prepare and check all equipment that may be required, prior to the start of anaesthesia (see Figure 1).



Figure 1. Airway and monitoring equipment

Monitoring

Standard monitoring must be applied prior to induction of anaesthesia. This includes oxygen saturation, ideally pre-ductal (right hand) and post-ductal (other limbs). A low post-ductal oxygen saturation may be a sign of low pulmonary blood flow, for instance due to significant pulmonary hypertension in a septic neonate (see transitional circulation below).

ECG and non-invasive blood pressure measurement should be used. The lower limit of mean arterial blood pressure can be estimated to be equivalent to the gestational age in weeks; by about 6 weeks of age, the normal mean arterial pressure is 50-60 mmHg. Basic intra-operative monitoring should ideally also include a precordial or oesophageal stethoscope and, if available, capnography must be used.

Airway equipment

Intubation and ventilation will be required unless it is an extremely short procedure. The size of the tracheal tube will depend on the weight of the neonate; most term babies require

a size 3.5 tracheal tube (see Table 1). Make sure that strapping is available. Precut the tape to fix the tracheal tube firmly in place immediately after intubation. An appropriately sized oral airway (preterm 000 – 00 and term neonate 0) and face mask should be available. Dead space within the apparatus is kept to a minimum with the appropriate sized breathing circuit and filter.

Warming

Neonates are extremely vulnerable to heat loss and hypothermia. Hypothermia (core temperature $<36^{\circ}\text{C}$) is associated with postoperative apnoeas, coagulopathy and poor wound healing, and worsens outcomes. The theatre environment should be warmed (or air conditioning turned down) to at least $20\text{-}23^{\circ}\text{C}$ and the baby kept covered as much as possible. A forced air warmer and a radiant heater should be used if available. Warmed packs should be considered if other sources of warming are not available; take care not to place warmed packs directly in contact with the skin. Fluids and blood products should be warmed. The temperature of the baby should be measured unless the procedure is very quick.

Preparation of drugs

The first thing to be drawn up is a saline flush so that the IV line can be flushed immediately after a drug is given. Calculate the correct dose of analgesics, muscle relaxants and antibiotics and draw these up. Double check dose calculations – it is easy to make 10-fold errors in neonatal practice.

Emergency drugs should be drawn up in the appropriate doses. These include atropine (20mcg.kg^{-1}), suxamethonium ($1\text{-}2\text{mg.kg}^{-1}$) and adrenaline (10mcg.kg^{-1} , i.e. 0.1ml.kg^{-1} 1:10,000 adrenaline).

Induction of anaesthesia

Inhalational induction is ideally with sevoflurane although halothane can also be used. The MAC of volatile agents is lower in neonates than in older children, and the onset of anaesthesia is relatively fast due to the rapid respiratory rate and high cardiac output. However, the neonatal myocardium is extremely sensitive to the negative inotropic effects of volatile

Table 1. Uncuffed tracheal tube sizes and lengths in neonates

Weight	Tube Size (ID) (MM)	Oral Length (cm)	Nasal Length (cm)
<0.7	2.0	5	5
<1.0	2.5	5.5	7
1.0	3.0	6	7.5
2.0	3.0	7	9
3.0	3.0	8.5	10.5
3.5	3.5	9	11

agents, so deep volatile anaesthesia must be avoided.

Sevoflurane can cause apnoea at high concentrations and the induction concentration should not exceed 6%. The neonate might require assisted mask ventilation until an airway is secured as they may hypoventilate during induction; take care to turn the inspired concentration of volatile agent down if assisted ventilation is used, otherwise the child will become very deep, very quickly. Halothane is more likely to cause myocardial depression and the induction concentration should be kept less than 2%. Halothane can cause arrhythmias in high concentrations, especially if the CO₂ is also high. Atropine (20mcg.kg⁻¹ IM or IV) should be considered prior to induction to reduce bradycardia, particularly if halothane is used.

Alternatively, intravenous induction with ketamine (2mg.kg⁻¹) or thiopentone (2-4mg.kg⁻¹) can be performed; induction of anaesthesia will be rapid (the anaesthetist must be confident they can manage the airway), and recovery may be delayed. Ketamine is particularly useful for the critically unwell neonate as cardiovascular depression is minimised. Use glycopyrrolate (10mcg.kg⁻¹ IV) or atropine (20mcg.kg⁻¹ IV or IM) to minimise the secretions caused by ketamine.

Maintenance of anaesthesia

Anaesthesia is maintained with volatile, oxygen and air or nitrous oxide. A ketamine infusion run at 2-4mg.kg.hr⁻¹ is a useful alternative in unstable neonates.

Attention must be paid when positioning the patient and pressure points must be protected. Whenever the child is moved, check the position of the tracheal tube as it is very easy to displace the tracheal tube in neonates, which could have potentially catastrophic consequences.

Pain management

It is important to consider pain management in all neonates. Pain pathways are fully developed before birth, and neonates display the physiologic, hormonal, and metabolic markers of the stress response.¹ Preterm infants have been shown to have an increased sensitivity to pain and even non-painful stimuli may be perceived as painful.² Long-term effects on pain responses have been documented in neonatal boys who were circumcised without analgesia.³ However, immature metabolic pathways for drugs and immature respiratory control mean that neonates are more sensitive to the side effects of analgesics commonly used during surgery.

Multimodal analgesia should be used for all neonates. Options include paracetamol (7.5mg.kg⁻¹ IV, or 20mg.kg⁻¹ PR), opioids such as fentanyl or morphine titrated to effect (fentanyl 1mcg.kg⁻¹ IV, morphine 10-20mcg.kg⁻¹ IV). Regional anaesthesia or infiltration of local anaesthetics should be

used where possible. Non-steroidal anti-inflammatory drugs (NSAIDs) should be avoided because of the immature renal system. Non nutritive sucking, sucrose and breast milk have also been shown to be safe and effective for reducing pain associated with procedures such as cannulation.^{4,5}

Invasive monitoring

Invasive monitoring (intra-arterial and central venous pressure) may be indicated depending on the type of surgery and the physiological status of the patient. Invasive monitoring is mandatory in circumstances such as cardiac surgery where there is the potential for rapid changes in blood pressure, use of inotropes or potential for large volume blood loss. In other circumstances, for instance neonatal laparotomy, the risks/benefits should be considered. Invasive monitoring is time consuming to insert, associated with complications and may delay the start of surgery. If the surgery is sufficiently urgent it may be necessary to proceed without. 24G or 22G catheters may be inserted into radial or femoral arteries for arterial monitoring, but distal limb perfusion must be checked. 4Fr or 5Fr central lines may be inserted into the femoral or internal jugular vein, ideally with ultrasound guidance. Near infrared spectroscopy (NIRS) can be used, if available, as a non-invasive monitor of tissue perfusion.⁶

Oxygen

Unmonitored oxygen therapy leading to hyperoxia in neonates is associated with retinopathy of prematurity, bronchopulmonary dysplasia and damage to the developing brain. Neonatal exposure to 100% oxygen is rarely necessary, and should be avoided except prior to interventions such as intubation. Hypoxia is also harmful, so targeting oxygen saturation levels between 91% and 95% is probably the safest practice.⁷⁻⁹ In low income countries where it may not be possible to deliver a variety of inspired oxygen mixtures, an air/oxygen mix should be used if possible and oxygen saturations should be monitored before, during and after anaesthesia.

Postoperative apnoea

Apnoea can be defined as a pause in breathing of more than 20 seconds or cessation of respiration of any duration accompanied by bradycardia or oxygen desaturation. Preterm infants are particularly at risk apnoeas due to an immature respiratory control centre. This effect is potentiated by general anaesthetic agents, and all term neonates <44 weeks post-conceptual age (PCA) and pre-term neonates (<60 weeks PCA) are at risk of postoperative apnoea. Infants with multiple congenital abnormalities, a history of apnoea and bradycardia, chronic lung disease and anaemia (Hb <10g.dl⁻¹) are at particular risk for postoperative apnoeas.¹⁰

Prophylactic caffeine (10mg.kg⁻¹ orally) can be given to prevent

post-operative apnoea in premature neonates.¹¹ Intravenous aminophylline (5mg.kg⁻¹) is an alternative although it has more side effects including tachycardia, jitteriness, irritability, feed intolerance, vomiting and hyperglycaemia.

It is important to allow sufficient time for neonates to wake up at the end of the operation, and they should be closely monitored in recovery until the anaesthetist is happy that they have returned to their normal awake state. All neonates <44 weeks post-conceptual age (PCA), ex-preterm infants up to 60 weeks PCA and any patients with whom there is any concern regarding the possibility of post-operative apnoeas should have post-operative apnoea and oxygen saturation monitoring for 24 hours.

Hypoglycaemia and hyperglycaemia

Persistent, recurrent or severe hypoglycaemia (blood glucose <2.5mmol.l⁻¹) may lead to irreversible neurological injury in neonates. Preterm infants and those with intrauterine growth retardation (IUGR) are at particular risk of hypoglycaemia. Fasting times should be minimized, blood glucose should be monitored and glucose containing maintenance fluids should be continued if they have been required prior to surgery.

Treat hypoglycaemia with a bolus of 2ml.kg⁻¹ of 10% dextrose. Hyperglycaemia (blood glucose >10mmol.l⁻¹) is also detrimental and is associated with increased mortality and sepsis in extremely low birth weight infants, so do not use boluses of 50% glucose.¹²

Perioperative fluids

Assessment of the fluid status of the neonate will help to guide peri-operative fluid replacement. It is helpful to consider preoperative maintenance fluids, intraoperative fluids and postoperative maintenance.

Preoperative maintenance fluids

A neonate may require preoperative maintenance fluids if they are unable to take fluids by mouth before surgery. In the first few days of life, the sodium requirement is not high, and typically 10% dextrose is recommended. After the post-natal diuresis has occurred at around day 3 of life, an isotonic fluid containing 5% dextrose and sodium should be used, and electrolytes and plasma glucose monitored (Table 2).

Table 2. Routine maintenance fluids in neonates

Day 1 of life	2ml.kg.hr ⁻¹ (50ml.kg.day ⁻¹)
Day 2 of life	3ml.kg.hr ⁻¹ (75ml.kg.day ⁻¹)
Day 3 of life and thereafter	4ml.kg.hr ⁻¹ (100ml.kg.day ⁻¹)

Intraoperative fluids

During surgery, isotonic fluids such as Hartmann's or Ringer's lactate must be used for resuscitation, replacement and maintenance to maintain intravascular fluid volume, replace fluid deficits and avoid hyponatraemia. Blood glucose should be monitored.

The decision whether to order or administer blood or blood products will depend on the cardiovascular status of the neonate, presence of haemorrhage, type of surgery, the most recent blood results and the normal expected values (Table 3). Once the decision to transfuse has been taken it may be worth transfusing to higher haemoglobin levels to avoid exposure to further donors. Ideally, the haematocrit should be measured during surgery using near-patient testing device such as a HemoCue[®] or a blood gas machine. The British Committee for Standards in Haematology (BCSH) has a suggested transfusion 'trigger' for neonatal top-up transfusion (Transfusion Guidelines for Neonates and Older Children (<http://www.bcsghguidelines.com>) (see Table 4). Suggested transfusion doses for blood and blood products are described in Table 5.

Transitional circulation

In utero, the pulmonary vascular resistance is high and there is very little blood flow to the lungs as the placenta is the source of gas exchange. After birth as the neonate takes the first few breaths, a chain of events is set in place that results in the transition from the foetal circulation to the neonatal circulation with closure of the foetal shunts (foramen ovale, ductus venosus and ductus arteriosus). During the first few weeks of life the pulmonary vasculature is highly reactive; an increase in pulmonary vascular resistance can lead to reopening of the foetal shunts, in particular the arterial duct between the pulmonary artery and the aorta. As a result there is right-to-left shunting from the pulmonary artery (deoxygenated blood) to the aorta, causing profound hypoxia. The oxygen saturation measured in the right hand may be normal ('pre-ductal'); the oxygen saturation in the other limbs ('post-ductal') will be low.

During the perioperative period it is important to prevent factors that increase pulmonary vascular resistance such as sepsis, hypoxia, acidosis, hypercapnoea, pain and hypothermia. When post-ductal oxygen saturations drop in relation to preductal oxygen saturations it may indicate a return to a foetal circulation.

Table 3. Normal haematological ranges for term and preterm babies (adapted from United Kingdom Blood Services Handbook of Transfusion Medicine, p54 4th Edition 2007 TSO)

	Term	Preterm	Adult
Haemoglobin g.l ⁻¹	140 - 240	140 -240	115 - 180
Platelets x 10 ⁹ .l ⁻¹	150 - 450	150 -450	150 - 400
PT (sec)	10 -16	11 - 22	11 - 14
APTT (sec)	31 - 55	28 - 101	27 - 40

Table 4. The British Committee for Standards in Haematology (BCSH) transfusion 'trigger' for neonatal top-up transfusion - reproduced from British Committee for Standards in Haematology (BCSH) Transfusion Guidelines for Neonates and Older Children - <http://www.bcsghguidelines.com> with permission

Postnatal age	Suggested transfusion threshold (g.l ⁻¹)		
	Ventilated	On oxygen/CPAP	Off oxygen
First 24 hours	<120	<120	<100
≤ week 1 (days 1-7)	<120	<100	<100
week 2 (days 8-14)	<100	<95	<75-85
≥ week 3 (days 15 onwards)		<85	Depending on clinical situation

Table 5. Suggested transfusion doses for blood and blood products (Joint United Kingdom (UK) Blood Transfusion and Tissue Transplantation Services Professional Advisory Committee <http://www.transfusionguidelines.org/transfusion-handbook/10-effective-transfusion-in-paediatric-practice/10-2-neonatal-transfusion>)

Product	Suggested transfusion dose
Packed cells	10-20ml.kg ⁻¹
Platelets	10-20ml.kg ⁻¹
FFP	12-15ml.kg ⁻¹

Neurodevelopmental effects of anaesthetics in neonates

Inadequate anaesthesia and analgesia have been shown to be detrimental to neonates, and associated with increased mortality. However, many animal model studies have been published recently that have demonstrated accelerated neuronal cell death ('apoptosis') and long-term behavioural changes after animals are exposed to anaesthetic agents in the neonatal period. The situation in humans remains unclear.¹³ The risks and benefits of surgery in neonates should be considered carefully, and non-essential elective surgery should be avoided in the neonatal period where possible.

Transfer of neonates

Neonatal surgery should ideally be undertaken in an environment where the facilities and expertise are available for definitive treatment and on-going care. In certain situations, if the baby is unstable and not suitable for transfer to theatre, it may be necessary to undertake surgery on the NICU itself. In certain situations the baby may need to be transferred to a specialist centre. In low-income countries this may not be an

option and treatment may not always be possible.

Prior to transfer the appropriate personnel, equipment, drugs and fluids should be prepared and checked using a transfer checklist (Table 6). The neonate should be carefully assessed for stability for transfer or if necessary transfer may need to be delayed for further resuscitation and optimisation. Check that the monitoring is functional and the patient is adequately fluid resuscitated. Take time to ensure that the neonate is stable prior to transfer on the current drug infusions and mode of ventilation.

Careful monitoring during transfer is extremely important and will highlight clinical trends. A detailed handover is essential for good continuity of care.

SPECIFIC NEONATAL PATHOLOGIES

Inguinal hernia repair

Inguinal hernia is common in premature neonates. The timing of surgery depends on the risk of incarceration, bowel strangulation or testicular atrophy versus the risk of

postoperative apnoea and the potential harm to neurodevelopment. The major anaesthetic risk is post-operative apnoea, which has been shown to vary from 4.7% to 49% of patients.^{10,14}

Some units prefer spinal anaesthesia for inguinal hernia repair, others use a balanced anaesthetic technique using general anaesthesia with intubation, supplemented with a regional technique. There is currently not enough evidence to show whether the incidence of apnoea is lower using spinal anaesthesia, and the choice is usually determined by local preference of the surgeon and anaesthetist.¹⁵

Editors' note: As this edition of Update goes to press, the editors are aware that the GAS study is reporting its preliminary findings on apnoea comparing GA and spinal in >700 neonates, publication of full results is expected late 2018.

Caudal anaesthesia using 0.25% bupivacaine 0.75ml.kg⁻¹ provides excellent supplementary analgesia for inguinal hernia repair under general anaesthesia. Alternatively, an ilioinguinal block can be performed with 0.5-1.0ml.kg⁻¹ 0.25% bupivacaine. These patients may require post-operative apnoea monitoring dependent on their PCA, as discussed earlier, and some premature infants will require post-operative ventilation or CPAP for treatment of apnoea. Paracetamol (7.5mg.kg⁻¹ IV or 20mg.kg⁻¹ rectal suppository) provides adequate post-operative analgesia.

Anorectal malformations

Anorectal malformations (ARM) occur in approximately 1:5000 live births. They represent a wide spectrum of disease, from a simple membrane involving the distal rectum and anus to more complex anomalies involving the genital and urinary tract. Spinal anomalies are frequently found in these patients. These include spinal dysraphism, low lying cord (LLC) and tethered cord.¹⁶ Plain spinal Xrays and spinal ultrasound are used to screen for these abnormalities although they may be normal in occult dysraphism. Caudal anaesthesia may be beneficial and can be used in ARM if there is certainty that anomalies of the spine and spinal cord have been excluded.¹⁷ ARM may be associated with other anomalies including Vertebral, Anorectal, Cardiac, Tracheoesophageal, Renal and Limb abnormalities, collectively known as the VACTERL association.

Primary surgical repair can be undertaken in the neonatal period although more commonly a colostomy is performed and a definitive repair is carried out at a later date.

If caudal anaesthesia is contraindicated an opioid-based technique is used (fentanyl 1-2mcg.kg⁻¹ or morphine 20-50mcg.kg⁻¹ [0.02-0.05mg.kg⁻¹], with infiltration with local anaesthetic. Rectal suppositories cannot be used but

intravenous paracetamol is a useful adjunct if available. Standard monitoring is usually all that is required. Opioids should be carefully titrated as the usual aim is to extubate at the end of surgery.

The patient may be positioned supine or prone depending on the surgical technique. Prone positioning is associated with increased risk to pressure areas, abdominal compression resulting in difficulty with ventilation, endobronchial intubation or tracheal tube displacement. Long-term outcome is variable depending on the complexity of the anorectal malformation. These patients usually require serial anal dilatations following repair.

Intestinal malrotation

Malrotation occurs in approximately 1:500 live births. Normal intestinal rotation around the superior mesenteric artery (SMA) and fixation during foetal development is interrupted. It may also be associated with congenital diaphragmatic hernia, exomphalos and gastroschisis.

Nearly 50% of cases will present in the first week of life most commonly with bilious vomiting secondary to duodenal obstruction.¹⁸ This may be due to a midgut volvulus, or physical compression secondary to peritoneal tissue bands or abnormal locations of the duodenum and its surrounding structures. If the condition is diagnosed early the neonate may be relatively well with only subtle abdominal signs. The neonate may present late with frank sepsis and peritonitis secondary to perforated or necrotic bowel. The gold standard radiological investigation is an upper GI contrast series. Plain X-rays are useful if there is concern of another diagnosis or to exclude visceral perforation.

These patients require adequate volume resuscitation and electrolyte replacement for ongoing fluid losses and should be taken to theatre as soon as is feasible. A nasogastric tube is inserted to suction the stomach. Prophylactic antibiotics such as co-amoxiclav or benzylpenicillin, gentamicin and metronidazole are required. Ideally invasive monitoring is inserted although it should not delay surgery in the sick neonate. If the gut has been compromised, inotropes may be needed and any coagulopathy will require correction. A central venous line may be required for ongoing total parenteral nutrition in the septic neonate. An opioid based technique can be used although a caudal may be considered if the patient is haemodynamically stable, there are no other contra-indications and extubation is anticipated. Post-operative NICU care and ventilation is often necessary.

Long-term outcomes depend on the extent of the necrotic bowel. Some patients will develop short bowel syndrome and if there is extensive bowel necrosis the mortality is 100%.

Necrotising enterocolitis (NEC)

Necrotising enterocolitis occurs in approximately 0.5 – 5:1000 live births. More than 90% of infants diagnosed with NEC are preterm.¹⁹ Morbidity and mortality are inversely proportional to the infant's post-conceptual age (PCA) and birth weight.

The aetiology of NEC is multifactorial. Risk factors include vascular compromise of the gastrointestinal tract, commencement of enteral feeding, immature gastrointestinal immunity and sepsis. Hypoxia or ischaemia combined with reduced splanchnic blood flow can occur with patent ductus arteriosus (PDA), cyanotic heart disease, respiratory distress syndrome, shock, asphyxia and with the use of umbilical catheters.

NEC may present with subtle gastrointestinal signs including abdominal distension, intolerance of feeds, abdominal tenderness, blood in the stool and bilious vomiting or may present with perforation and peritonitis with systemic signs including shock, temperature instability, acidosis and disseminated intravascular coagulopathy. Supine and decubitus plain Xrays may show the presence of hepatic venous gas, free intraperitoneal air, dilated bowel loops, ascites and asymmetric bowel gas patterns along with pneumatosis intestinalis.

Initial management includes discontinuation of enteral feeds, insertion of a nasogastric tube and commencement of broad-spectrum antibiotics such as benzylpenicillin, gentamicin and metronidazole. Ongoing fluid and electrolyte management with parenteral nutrition will be required. Frequent clinical monitoring of systemic and abdominal signs together with radiographic examination, monitoring of laboratory values and acid-base status guides further management. The only absolute indication for surgery is bowel perforation although the decision to proceed to surgery may be made if there is a clinical deterioration.

The preoperative assessment should evaluate and optimise any cardiovascular instability, metabolic acidosis, coagulopathy and respiratory compromise. If the patient is too unstable it may be necessary to carry out surgery on the NICU.

These patients are often already intubated and ventilated. A high dose fentanyl technique (10-20mcg.kg⁻¹) may be used to promote cardiovascular stability and reduce the systemic stress response.²⁰ Nitrous oxide should be avoided because of the risk of bowel distension. Low cardiac output state, organ hypoperfusion and acidosis secondary to large fluid shifts is common, and large volumes of intravenous fluids are frequently required. Invasive monitoring is useful to guide fluid management and allow frequent arterial blood gas sampling although this must be balanced against the risk of limb ischaemia in the preterm neonate. Insertion of an arterial

or a central line should not delay the start of surgery in the sick infant. There is a significant risk of coagulopathy and significant blood loss, and inotropes are often required. Packed red cells should be available and fresh frozen plasma and platelets are often indicated based on laboratory results or clinical evidence of bleeding. Hypothermia and glucose instability are common and should be managed appropriately.

Mortality remains significant and long term complications include short bowel syndrome and neurodevelopmental delay.

Oesophageal atresia and tracheoesophageal fistula

Congenital tracheoesophageal fistula (TOF) occurs in approximately 1:3,000 live births. It arises during foetal development as a result of incomplete separation of the oesophagus from the laryngotracheal tube. It is classified based on the site and presence of the fistula and whether there is oesophageal atresia (Figure 2). There may be other associated VACTERL anomalies.

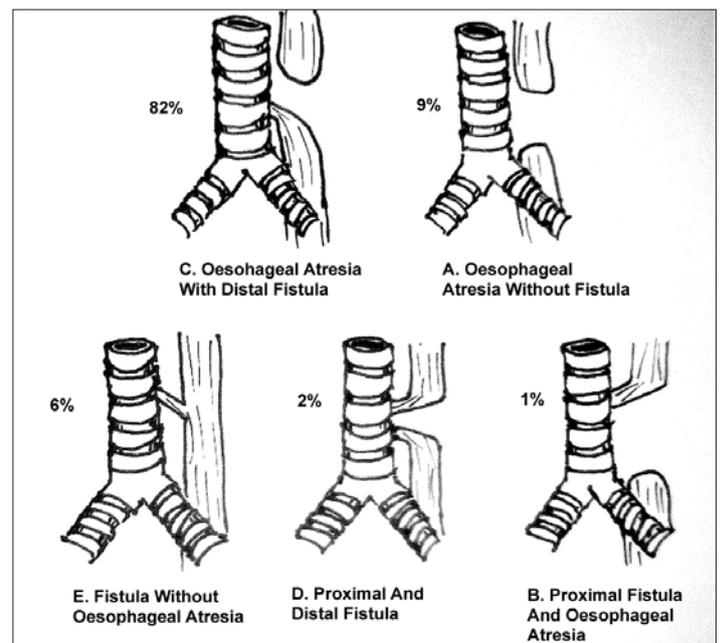


Figure 2. Incidence and types of oesophageal atresia/tracheo-oesophageal fistula

Neonates with TOF classically present within a few hours of birth with frothy sputum as they are unable to swallow oral secretions; delayed diagnosis is associated with episodes of coughing and choking associated with cyanosis, particularly if feeding is attempted. There may be copious oral secretions and abdominal distension due to gastric insufflation via the fistula. Left untreated the neonate will develop aspiration pneumonia. The diagnosis of oesophageal atresia is confirmed if it is not possible to pass a nasogastric tube and the chest Xray will show the nasogastric tube coiled in the proximal blind-ending

oesophagus (Figure 3). There may be an absent gastric bubble in isolated oesophageal atresia without a tracheoesophageal fistula.

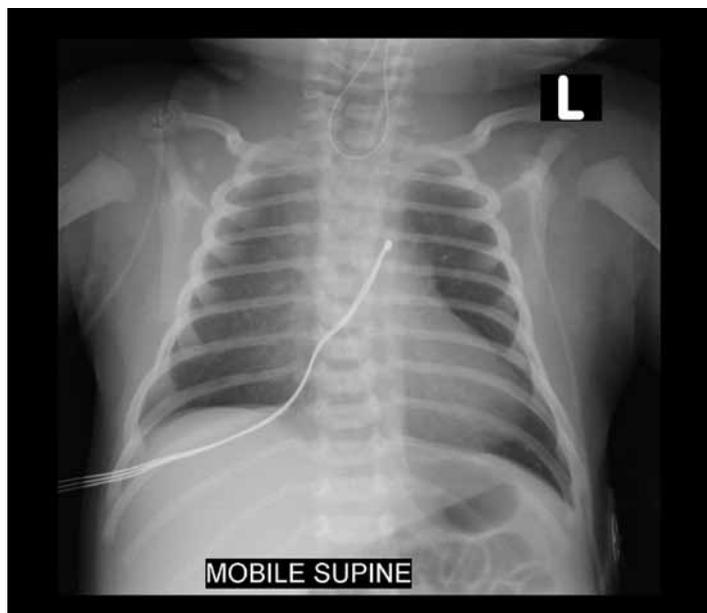


Figure 3. Chest Xray showing nasogastric tube curled in the upper oesophageal pouch in a child with oesophageal atresia

The goals of pre-operative management are to stabilise the child, minimise respiratory embarrassment and assess for timing of surgery. A nasogastric tube is inserted into the upper oesophageal pouch to drain secretions. The patient must be nursed head up or on the side to minimise the risk of aspiration. Intravenous fluids and prophylactic antibiotics should be commenced. This allows time for investigations such as an echocardiogram to exclude other associated congenital abnormalities.

Our preferred technique is to induce anaesthesia after pre-oxygenation and to maintain spontaneous ventilation initially with volatile or intravenous anaesthesia. Prior to repair the surgeons may perform flexible or rigid bronchoscopy to assess the level of the fistula and to see if there is a second or proximal fistula. Take note of the distance measured from the cords to the fistula to guide tracheal tube placement; the fistula is mid-tracheal in two thirds of cases, and located at level of the carina in one third of cases. Muscle relaxants and gentle mask ventilation may be given prior to intubation. If possible the tracheal tube is placed distal to the fistula, with the bevel of the tracheal tube facing anteriorly.

A right thoracotomy is performed with the patient on the side with a roll under the chest. The tube position must be checked and effective ventilation confirmed after the change of position. The lung is then retracted which often results in difficulty with ventilation, hypercapnoea and acidosis. Periods

of manual ventilation may be required. If gastric distension occurs prior to ligation of the fistula, the tracheal tube should be disconnected intermittently to decompress the stomach via the airway. An arterial line is useful to facilitate arterial blood gas measurement as end tidal CO_2 measurement is unreliable. Alternatively, transcutaneous CO_2 monitoring can be used. Hypercapnoea and acidosis is of particular importance in the presence of certain cardiac anomalies as the increased pulmonary vascular resistance can lead to right-to-left shunting and severe hypoxia. Other pitfalls include ligation of the wrong structure, intubation of the fistula and endobronchial intubation.

A balanced anaesthetic should be given, with bolus fentanyl analgesia as required ($1-2\text{mcg.kg}^{-1}$). Blood loss should be minimal and Ringer's lactate $10-20\text{ml.kg}^{-1}$ is usually all that is required. The wound should be infiltrated with local anaesthetic at the end of surgery. Some term infants born in good condition and with normal preoperative pulmonary function may be extubated at the end of surgery; most are likely to require post-operative ventilation and they should be transferred to a facility able to provide this level of care for their surgery. Many patients will require serial dilatation of the oesophagus during infancy.

Congenital diaphragmatic hernia

Congenital diaphragmatic hernia (CDH) occurs in approximately 1:3000 live births. In most cases the aetiology remains unknown. A defect in the diaphragm, usually on the left side, results in herniation of midgut structures into the thoracic cavity. Pulmonary vascular structure and reactivity are abnormal and there is a varying degree of lung hypoplasia. Diagnosis is made on antenatal ultrasound or on plain Xray postnatally when the abnormal bowel loops can be seen within the thoracic cavity.

Morbidity and mortality is related to the degree of pulmonary hypertension, right ventricular dysfunction and lung hypoplasia.²¹ A pre-operative echo is performed as a significant proportion of CDH have associated cardiac anomalies. Mortality still remains high in patients with significant co-existing congenital cardiac disease.²² It is generally accepted that delaying surgery, usually for 24-48 hours, allows a period of stabilisation. The reduction in pulmonary artery pressures and improvement in right ventricular dysfunction may improve outcome.

There has been a significant improvement in survival over the past 20 years due to the introduction of 'gentle ventilation' strategies.²³ These include permissive hypercapnoea ($\text{PaCO}_2 < 70\text{mmHg}$), limiting inflation pressures (avoid $\text{PIP} > 25\text{cm H}_2\text{O}$ and $\text{PEEP} > 5\text{cm H}_2\text{O}$) and accepting relative hypoxaemia

(aim for pre-ductal SpO₂ 90-95%). Surgery may need to be performed whilst the neonate is on high-frequency oscillation ventilation or extra-corporeal membrane oxygenation.²⁴

Preoperative assessment must pay particular attention to the presence of significant pulmonary hypertension, ventilation requirements, and associated cardiac anomalies. If the infant is not already intubated, anaesthesia is induced with care to avoid gastric insufflation with bag valve mask ventilation and further lung compression. A nasogastric tube is inserted to decompress the stomach. Invasive monitoring is required to allow serial blood gas measurement. There is a risk of blood loss and a unit of packed red cells should be available. In patients with significant pulmonary hypertension, having nitric oxide available in theatre may be critical for treatment of pulmonary hypertensive crises.

A subcostal or transverse abdominal incision is made and the herniated viscera are reduced into the abdomen. The diaphragmatic defect is then either closed primarily or with a prosthetic patch if the defect is large. Thoracoscopic repair is being undertaken in some centres. Following abdominal closure, raised intra-abdominal pressures may lead to difficulty with ventilation and a risk of developing abdominal compartment syndrome, and delayed closure may be necessary. Lung compliance decreases post-operatively and post-operative ventilation is usually necessary. These patients often suffer from chronic respiratory disease, gastro-oesophageal reflux disease and neurodevelopmental delay.

Gastroschisis and exomphalos (omphalocele)

Gastroschisis and exomphalos are both ventral wall defects resulting in herniation of abdominal viscera. Diagnosis is ideally made on antenatal ultrasound scan.

Gastroschisis occurs in approximately 1:3000 live births. The herniated viscera are not covered by a sac. It is thought to occur secondary to an ischaemic insult during abdominal wall development or due to early rupture of the hernia of the umbilical cord. A relatively small percentage (10-20%) are associated with other congenital abnormalities and these predominantly involve the gastrointestinal tract.²⁵

Exomphalos occurs in approximately 1:5000 live births. Failure of normal embryological development results in the bowel remaining within the umbilical cord and not returning to the abdomen. The herniated viscera is covered by a sac. There is a high incidence of associated congenital abnormalities including cardiac anomalies. Specific chromosomal associations include trisomies 13, 15, 18 and 21 and it can be associated with Beckwith-Wiedemann syndrome.

To avoid bowel injury the baby is delivered by caesarean section. The operating theatre should be warmed, the baby

dried, any exposed bowel covered with plastic and a nasogastric tube is inserted to decompress the stomach. Fluid resuscitation is commenced, a urinary catheter inserted and broad spectrum antibiotics started. Co-existing congenital abnormalities, especially cardiac, should be assessed. A renal or cranial ultrasound may also be indicated.

Surgery is more urgent in gastroschisis due to the ongoing fluid losses and electrolyte and metabolic derangement. If primary closure is not possible then a 'silo' is placed over the exposed bowel, which may require a general anaesthetic if the defect needs extending to fit the device. The silo is suspended above the patient postoperatively, and the bowel is gradually reduced into the abdominal cavity under gravity over the ensuing few days in the NICU. When the patient is stable and spontaneous reduction of the bowel has reached a plateau, then surgery for reduction and closure of hernia is performed. Surgery for exomphalos is less urgent, unless the sac has ruptured. If the patient is stable and the defect is small a primary repair can usually be done. In large defects, if the sac has not ruptured, it may be treated with topical silver sulfadiazine to allow epithelisation with definitive surgery at a later stage.

The neonate will require intubation and ventilation for surgery. Expect significant ongoing fluid and heat losses due to the exposed viscera. Peripheral intravenous access may be all that is required, but central venous pressure monitoring and an arterial line are useful to help guide fluid administration. Avoid the femoral vessels as there is a risk of decreased perfusion with the increased abdominal pressures. Placing the post-ductal oxygen saturation probe on either lower limb helps to give an indication if there is poor perfusion. Muscle relaxants will assist the surgeons in reducing the abdominal contents. Reduction of the bowel may cause abdominal compartment syndrome, diaphragmatic splinting and high ventilation pressures. If the intra gastric pressures are >20mmHg or the peak inspiratory pressures exceed 30cm H₂O then a staged repair is indicated.²⁶

Unless there is a very small defect the infant will require post-operative ventilation and a generous opioid-based anaesthetic technique can be used (fentanyl 10-20mcg.kg⁻¹). These patients often require parenteral nutrition and a significant proportion present for further abdominal surgery.

CONCLUSION

Improving outcomes in neonatal anaesthesia is dependent on a thorough understanding of the unique anaesthetic requirements of the neonate and a detailed knowledge of the different pathologies that present during this period. Unnecessary surgery should be avoided during the neonatal period as anaesthesia and surgical stress may have detrimental effects on the very immature child.

REFERENCES

1. Anand KJ, Aynsley-Green A. Measuring the severity of surgical stress in newborn infants. *J Pediatr Surg* 1988; **23**: 297-305.
2. Anand KJ. Clinical importance of pain and stress in preterm neonates. *Biol Neonate* 1998; **73**: 1-9.
3. Taddio A, Katz J, Illersich AL, Koren G. Effect of neonatal circumcision on pain response during subsequent routine vaccination. *Lancet* 1997; **349**: 599-603.
4. Stevens B, Yamada J, Lee GY, Ohlsson A. Sucrose for analgesia in newborn infants undergoing painful procedures. *Cochrane Database Syst Rev* 2013 1:CD001069.
5. Shah PS, Herbozo C, Aliwalas LL, Shah VS. Breastfeeding or breast milk for procedural pain in neonates. *Cochrane Database Syst Rev* 2012 12:CD004950.
6. Cerbo RM, Maragliano R, Pozzi M, Strocchio L. Global perfusion assessment and tissue oxygen saturation in preterm infants: where are we? *Early Hum Dev* 2013; **89**(S1): S44-6.
7. Bancalari E, Claure N. Oxygenation targets and outcomes in premature infants. *JAMA* 2013; **117**: 2161-2.
8. Stenson B, Brocklehurst P, Tarnow-Mordi W. U.K. BOOST II trial; Australian BOOST II trial; New Zealand BOOST II trial. Increased 36-week survival with high oxygen saturation target in extremely preterm infants. *N Engl J Med* 2011; **364**: 1680-82.
9. Schmidt B, Whyte RK, Asztalos EV et al. Canadian Oxygen Trial (COT) Group. Effects of targeting higher vs lower arterial oxygen saturations on death or disability in extremely preterm infants: a randomized clinical trial. *JAMA* 2013; **309**: 2111-20.
10. Cote CJ. Postoperative apnea in former preterm infants after inguinal herniorrhaphy: A combined analysis. *Anesthesiology* 1995; **82**: 809-22.
11. Henderson-Smart DJ, Steer P. Prophylactic caffeine to prevent postoperative apnea following general anesthesia in preterm infants. *Cochrane Database Syst Rev* 2001;4:CD000048.
12. Kao LS, Morris BH, Lally KP, Stewart CD, Huseby V, Kennedy KA. Hyperglycemia and morbidity and mortality in extremely low birth weight infants. *J Perinatol* 2006; **26**: 730-6.
13. Olsen EA, Brambrink AM. Anesthetic neurotoxicity in the newborn and infant. *Curr Opin Anaesthesiol* 2013 August 29 (e-pub ahead of print).
14. Malviya S, Swartz J, Lerman J. Are all preterm infants younger than 60 weeks postconceptional age a risk for post-anesthetic apnea? *Anesthesiology* 1993; **78**: 1076-81.
15. Craven PD, Badawi N, Henderson-Smart DJ, O'Brien M. Regional (spinal, epidural, caudal) versus general anaesthesia in preterm infants undergoing inguinal herniorrhaphy in early infancy. *Cochrane Database Syst Rev* 2003;3:CD003669.
16. Teo AT, Gan BK, Tung JS, Low Y, Seow WT. Low-lying spinal cord and tethered cord syndrome in children with anorectal malformations. *Singapore Med J* 2012; **53**: 570-6.
17. Bozza P, Morini F, Conforti A, Sgrò S. Stress and ano-colorectal surgery in newborn/infant: role of anesthesia. *Pediatr Surg Int* 2012; **28**: 821-4.
18. Millar AJ, Rode H, Cywes S. Malrotation and volvulus in infancy and childhood. *Semin Pediatr Surg* 2003; **12**: 229-36.
19. Llanos AR, Moss ME, Pinzón MC, Dye T. Epidemiology of neonatal necrotizing enterocolitis: a population-based study. *Paediatr Perinat Epidemiol* 2002; **16**: 342-9.
20. Anand KJ, Sippell WG, Aynsley-Green A. Randomised trial of fentanyl anaesthesia in preterm babies undergoing surgery: effects on the stress response. *Lancet* 1987; **1**: 62-6 [published correction appears in *Lancet* 1987; **1**: 234].
21. Wynn J, Krishnan U, Aspelund G, Zhang Y et al. Outcomes of congenital diaphragmatic hernia in the modern era of management. *J Pediatr* 2013; **163**: 114-9 e1.
22. Graziano JN. Cardiac anomalies in patients with congenital diaphragmatic hernia and their prognosis: a report from the Congenital Diaphragmatic Hernia Study Group. *J Pediatr Surg* 2005; **40**: 1045-9.
23. Guidry CA, Hranjec T, Rodgers BM, Kane B. Permissive hypercapnia in the management of congenital diaphragmatic hernia: our institutional experience. *J Am Coll Surg* 2012; **214**: 640-645.
24. Fallon SC, Cass DL, Olutoye OO, Zamora IJ et al. Repair of congenital diaphragmatic hernias on Extracorporeal Membrane Oxygenation (ECMO): Does early repair improve patient survival? *J Pediatr Surg* 2013; **48**: 1172-6.
25. Molik KA, Gingalewski CA, West KW et al. Gastroschisis: a plea for risk categorization. *J Pediatr Surg* 2001; **36**: 51-5.
26. Yaster M, Scherer TL, Stone MM et al. Prediction of successful primary closure of congenital abdominal wall defects using intraoperative measurements. *J Pediatr Surg* 1989; **24**: 1217-20.

Major elective surgery in children, and surgery in remote and rural locations

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INTRODUCTION

Safe, effective care of children presenting for surgery is challenging, even more so when the child presents with advanced pathology, the systems to support safe anaesthesia care are not well-developed, and the need for surgery is large. In many low-income countries, more than 50% of the population is less than 15 years old, and more than 85% of children are predicted to need some form of surgical intervention by the age of 15. The overall morbidity and mortality in this group of patients is alarmingly high when compared to similar patients in high-income countries, and the mortality would be expected to be even higher in the emergency cases.

Elective paediatric cases in children older than 6 months old are performed at many hospital levels in the rural and the urban setting. Common elective procedures in rural hospitals include:

- Inguinal and umbilical hernia
- Circumcision
- Incision and drainage of soft tissue and orthopaedic infections.

Larger more challenging cases often undertaken in urban centres include:

- Hypospadias repair
- Excision of abdominal mass (e.g. mesenteric cyst, Wilm's Tumor, ovarian mass)
- Head and neck surgery.

These patients are likely to be challenging even in a large national referral centre and good understanding and preparation is required.

This article will give a brief overview of the general principles of anaesthesia in children,

and describe some clinical cases with practical solutions to equipment requirements and clinical goals, which we hope will be useful to anaesthesia providers working in any setting.

GENERAL PRINCIPLES

Cardiac

Infants and young children have a relatively fixed cardiac output compared to adults due to immature myocardial function. The resting heart rate is high and there is limited ability to increase the stroke volume in response to a fluid challenge. Babies become bradycardic (heart rate slows) in response to hypoxia, but children older than 6 months of age have developed a balance between the sympathetic and parasympathetic nervous systems, and have the more usual tachycardia in response to hypoxia. The fall in the heart rate due to hypoxia in babies causes a fall in blood pressure; your immediate response should be to control the ventilation with 100% oxygen, or whatever level of oxygen is available in your setting, rather than attend to the blood pressure – i.e. you should not grab for the atropine but rather open the airway and/or assist ventilation, as hypoxia will be the source of the bradycardia in the vast majority of the situations. Your response to hypoxia must be rapid as babies have twice the oxygen consumption of adults, and quickly become desaturated. Always prepare your age appropriate airway equipment and full range of drugs prior to the start of a case, and make sure you have an assistant.

Children presenting for elective surgery, particularly those with congenital conditions associated with midline defects such as hypospadias, cleft lip and cleft palate, may also have associated congenital heart disease (CHD). If a murmur is present, or the child has a history

Summary

Children presenting for elective paediatric surgery in sub-Saharan Africa may have a high morbidity and mortality. Understanding the basic anatomy, physiology and pharmacology together with appropriate equipment for the paediatric patient will improve the anaesthesia mortality statistics. The development of paediatric surgical centres in both the rural and urban settings will allow for greater experience to be obtained in paediatric anaesthesia, which will improve care. The most valuable asset for these paediatric centres is to have well-trained physicians and nurses who can provide high quality care for children with the advanced surgical pathology encountered, taking account of the lack of infrastructure and the limited supplies that are a common problem. A successful perioperative course can be expected even for children requiring surgical intervention in austere environments.

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suggestive of cardiac disease (failure to thrive, frequent chest infections and/or cyanosis), then a further work-up may be indicated prior to surgical intervention. Most children will present to a general hospital without an adult cardiologist, let alone a paediatric cardiologist. Work-up for such a child should include a good history, including questions about feeding, weight gain, chest infections, 'funny turns' and exercise tolerance; examination to listen for murmurs or cyanosis and clubbing; chest Xray (to exclude cardiomegaly or pulmonary edema); and room air oxygen saturation measured with a pulse oximeter to exclude cyanotic heart disease or a chest infection in a child with cardiac failure. If the child is well and asymptomatic, not cyanosed, the murmur is not loud, not diastolic, there is no thrill and the chest Xray is normal, it should be safe to proceed as the murmur is likely to be benign. These cases will not require antibiotic prophylaxis unless the surgeons request an antibiotic for their surgical procedure.

Acquired rheumatic valvular heart disease is always a possible finding in a low resource setting, as the prevalence of this disease is higher and is increasing. If the patient is asymptomatic with good exercise tolerance and oxygen saturation, an elective surgical case can be performed safely.

Respiratory

Anaesthetic induction and intubation in babies requires special care because the oxygen saturation will decrease much faster than in an adult due to the higher oxygen consumption, high minute ventilation and reduced functional residual capacity (FRC) per kg body weight compared to adults.

The narrowest part of the airway in the child is the cricoid cartilage, not the vocal cords as in the adult. The glottis is more anterior and the epiglottis is less rigid and tends to fall back to occlude the glottic opening during intubation. The large occiput in infants tends to cause neck flexion, the tongue is relatively large, and it is easy to press on the floor of the mouth to push the tongue up, which means the airway is easily obstructed during facemask anaesthesia. The airway cartilages are soft and easily compressed; if cricoid pressure is used, the pressure should be gentle otherwise the trachea can collapse and you may not be able to ventilate due to obstruction of the trachea itself. Lateral displacement is also a frequent factor when an assistant is applying cricoid pressure with too much enthusiasm. If this is the case, ask your assistant to release the pressure to improve your view of the larynx. The combination of these anatomical differences and the limited oxygen reserve, relatively high oxygen requirement, and poor tolerance of hypoxia means that intubation can be more difficult in a neonate compared to an adult, but with skill and experience, safe intubation becomes routine.

The following airway equipment must always be prepared and checked prior to surgery. This preparation is a necessary, essential and professional aspect of the anaesthetic plan:

- Stethoscope
- Pulse oximeter
- Functioning laryngoscope with blades
- Endotracheal tube and stylet
- Oropharyngeal airways
- Mask.

It is essential to have the appropriate sized paediatric airway equipment available, including laryngoscope blades (Miller 0, 1, 2 blades), endotracheal tubes, facemasks and oral airways. A stylet can be helpful but, if not available, can be made with a flexible metal tube that has been blunted on both ends to prevent tracheal damage or endotracheal tube perforation. A precordial stethoscope and a pulse oximeter are essential monitoring equipment for all cases.

The technical skill of intubation should be practised in adults or older children before attempting to perform even an elective intubation in a small child; many paediatric airway disasters could have been avoided with better training, also with assistance on induction. The anaesthetist should always ask the surgeon or a nursing colleague to help with induction and intubation as respiratory disasters can happen very quickly in children.

Renal

Glomerular filtration rate (GFR) reaches adult levels by one year of age, and most children presenting for elective surgery have normal renal function. Routine questions such as "Any wet diapers/nappies, and when?" will be useful to assess hydration and renal function healthy children. Routine electrolytes and creatinine are not necessary except for in renal surgery where it is useful to record baseline values. A urinary catheter is rarely required, except for hypospadias repair or Wilm's tumours. The smaller sizes of urinary catheter are often not available, and it is much better to avoid damage to the urethra rather than to insert an inappropriately large catheter. The bladder can be emptied by the surgeon pressing gently on the lower abdomen during the case (if the area is surgically prepped), and urine collected in a diaper. Urine output can be estimated from the difference in weight of the diaper pre-surgery and post-surgery; one mg increase in weight in the diaper is equivalent to one ml of urine. A scale capable of measuring small weights must be used. All doses of drugs should be calculated and drawn up accurately, especially renal toxic drugs such as gentamicin.

Fluid balance should be assessed carefully before surgery, particularly if the child has been starved for a long period of time; many children are starved for far too long preoperatively. Patients in an arid climate often have a chronically low intravascular volume; the additive effect of the pre-existing deficit and the fasting deficit may be revealed on induction of anaesthesia with inhalational agents such as halothane, which results in a dramatic fall in blood pressure. A bolus dose of 10-20ml.kg⁻¹ 0.9% saline or Ringer's provides adequate hydration (and rehydration) for most minor elective cases. Weigh swabs and assess fluid balance carefully for children undergoing major surgery with on-going fluid and blood losses. Most children presenting for elective general surgery do not require dextrose containing fluids as the stress response to surgery causes the blood sugar to increase; neonates or malnourished children should have their blood sugar checked to make sure they are not hypoglycaemic before surgery starts.

Make sure the child is well hydrated at the end of surgery; and, if possible, avoid IV fluids on the ward unless the child has undergone major surgery and is not able to drink. This is important even in the elective patient as it is difficult to monitor IV fluids on the wards as the patient:nurse ratio is often very high. Patients frequently do not get an intravenous line in good time, the IV fluids may not be available or may not be infused accurately.

Temperature regulation

Temperature monitoring is not available in many countries, but low technology devices to detect hypothermia are available, and valuable, particularly in younger children. A simple thermometer can be used for axillary skin temperature measurement to help monitor trends. Children have a relatively large surface area and little fat for insulation, especially if malnourished. This can lead to dangerous hypothermia (low temperature) during surgery, which may result in clotting problems, hypoventilation and even cardiac arrhythmias. Basic heating pads and fluid warmers are helpful but need very close monitoring as they may also cause burns if not used properly. In particular, the heating pad should never be applied directly to the skin, and rarely placed on "high".

Intraoperative hypothermia can be avoided in the following ways:

- Avoid excessive betadine to 'prep' the surgical field. If you are not watching the surgical technician's use of betadine, the patient will be floating in cold fluid for the entire case. Pooling of betadine (or alcohol) against the child's skin may also cause a chemical burn.
- If possible, warm the theatre (22-25°C, depending on the age of the child). An ambient heating unit is useful to warm the room and reduce early heat loss.

- Do not let the room get too cold. An open window producing a breeze for the surgical team may cool the patient down too much. If air conditioning is available, make sure the temperature is not turned down too low.
- Keep the patient covered with warm sheets whenever possible, including at the start of the case when IV lines are being inserted, and/or prior to cleaning and draping by the surgical team. It is very helpful if the early heat loss caused by vasodilation at the start of anaesthesia can be reduced.

Haematology

Anaemia with haemoglobin level (Hb) less than 8g.dl⁻¹ is common in children presenting for elective surgery. Most will be nutritional, but you should consider other causes such as:

- Malaria
- Sickle cell disease
- Intestinal worms
- Drug-induced anaemia (rare).

It is feasible for children to have elective surgery when their haemoglobin is less than 8 g.dl⁻¹, but they have reduced tissue oxygen delivery, and may need supplemental oxygen to maintain oxygen saturations, especially if they have upper abdominal surgery and impaired respiratory function due to pain. Minor procedures such as hernia repair can be undertaken safely with Hb as low as 6-7g.dl⁻¹, but any major surgery needs a starting Hb above 8g.dl⁻¹. If the child is anaemic and is presenting for elective surgery, and they live relatively close to the hospital, they should be treated with a course of iron supplements for 3 months before re-booking.

Acute malaria can produce unexpected complications and increased morbidity. All children presenting for elective surgery who have malaria should be treated and surgery postponed.

Sickle disease is associated with increased perioperative complications, usually due to sickle chest or painful crisis. Children with sickle disease presenting for elective surgery should not be allowed to become dehydrated, and should be transfused using fresh whole blood if the Hb is below 8g.dl⁻¹. Ideally, freshly donated blood should be used for sickle cell disease patients, particularly if it is not possible to measure the sickle status of the donated blood. Typically a top up transfusion of 20ml.kg⁻¹ will be sufficient, along with the normal sickle cell precautions, page 35. If the temperature is high after blood transfusion in a malaria endemic area, the child should have blood taken to test for malaria parasites and treatment for malaria considered.

CLINICAL CASES

Case 1

A three-year-old male child was referred to a tertiary referral hospital in East Africa with an 8 month history of enlarging abdominal mass. The child was previously healthy, travelled from a neighbouring country, had been examined by multiple medical care providers, and was very malnourished. His mother came with a CT scan of the mass and an exploratory laparotomy was scheduled by the surgical team. See Figure 1.

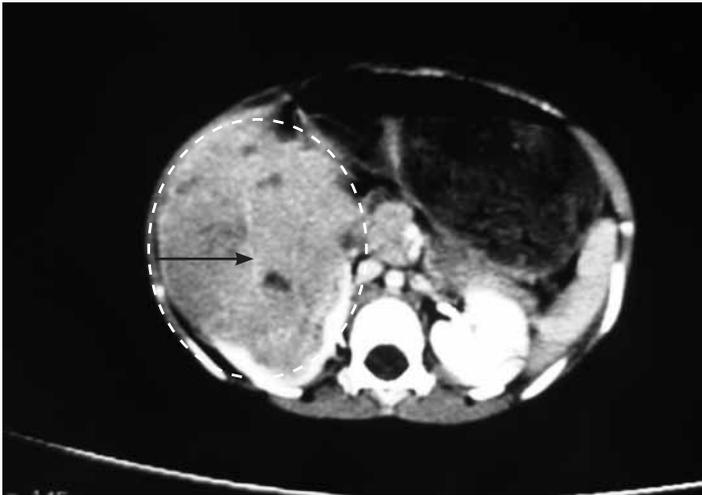


Figure 1. CT scan: large renal mass on the patient's right side (the left side of the scan) - indicated within broken line

Preoperative evaluation and plan

Nephroblastoma (Wilms' Tumor) is the most common abdominal tumour in children, with a reported incidence of 1:20,000 to 1:100,000. Typically tumours in sub-Saharan Africa are more advanced and in Nigeria, nephroblastoma is reported to be Stage III and IV in 72% of the children with an undocumented overall survival.¹ Advanced disease suggests delayed presentation, poor surgical infrastructure or, possibly, more aggressive tumours compared to children in western research studies. Anaesthesia and surgery for advanced tumour cases can be very challenging.

A CT scan and a chest Xray are essential to plan surgery, to determine the mass effect of the tumour, location within the vascular system (e.g. identify involvement of the inferior vena cava or renal vein), presence of bilateral renal involvement (8-12% of cases), and spread to the pulmonary parenchyma. Each of these would help determine if surgery should be altered or even cancelled due to extensive spread and unnecessary risks, although a surgical palliative mass debulking could still be considered. Many of these patients present in a state of malnutrition and their response to inhalation agents such as halothane may be more dramatic with more cardiovascular depression and drop in blood pressure than anticipated.

Each child needs an accurate weight documented in the chart for drug dosing, fluid requirements, and overall health assessment. Essential laboratory measurements include: haemoglobin, platelet count, creatinine (allows comparison of pre and postoperative renal function) and blood type and cross match, anticipating the potential for significant blood loss. A minimum of two adult units of type specific blood must be ready prior to skin incision. In addition, two family members with a matching blood type, or other appropriate donors, must be available in the theatre waiting area, with direct communication from the surgical team. You will need to have a minimum of two blood transfusion sets in theatre, in case one becomes obstructed with blood clots during the case. Additional labs are unnecessary and costly.

Postoperative care must be planned before surgery, including where the child will be cared for after surgery. The postop ward must allow for close observation with access to:

- Oxygen
- Suction
- Bed which has the capability to elevate the head (improving respiratory efforts)
- Focused nursing care and monitoring of vital signs.

This is often the key component to a successful hospital discharge from the surgical procedure and should never be overlooked in the preparatory phase of an anaesthesia plan. Many hospitals do not have an ICU, but good care is possible as a substitute for a formal ICU if the patient is positioned in the area closest to the nursing station with access to close monitoring. The vital signs and fluid input and output must be monitored (bladder catheter required), and a key person identified who will show an active response to the postoperative observations.

Induction and maintenance of anaesthesia

A large intra-abdominal tumour may predispose the patient to regurgitation of gastric contents on induction of anaesthesia. The diaphragm will be pushed up which will limit the lungs from fully expanding thus causing a faster oxygen desaturation. Both of these factors should prompt the anaesthesia care provider to ask for assistance during the induction phase of anaesthesia. (See Figure 2)

This patient should have an IV induction with careful application of cricoid pressure (NOT an inhalation induction). Remember, if you are having difficulty viewing the glottis, ask your assistant to reduce the cricoid pressure and/or change their compression direction to a more midline position. This patient could have any induction agent (thiopentone, propofol,



Figure 2. Induction of a child with a large Wilms' tumor: note multiple assistants, head of bed elevated, cricoid pressure, stethoscope ready, senior anaesthesia care provider ready to assist in intubation in teaching scenario

or ketamine); and muscle relaxation (succinylcholine or rocuronium) to provide good conditions for rapid intubation. I would not use any narcotics at this stage, but would wait until the airway has been secured. Children can have a more dramatic drop in oxygen saturation when they are apnoeic compared to adults, due to higher oxygen consumption, and in this case, the child will also have a reduced functional oxygen reserve, so will require efficient intubation. If the mass is very large, the head of the bed can be elevated slightly to reduce the effect of the mass compressing on the diaphragm, which may assist during the induction period. The lung volumes will be reduced due to elevation of the diaphragm, so check more than once that the endotracheal tube is not down too far and is in the proper position in the trachea. This patient will do best with a cuffed endotracheal tube, if available, due to increased intra-abdominal pressure during surgical tumour manipulation. If an uncuffed endotracheal tube is all that is available, place the appropriate size tube that only has a leak around the tube at higher pressures (between 20-30cmH₂O).

Higher inspiratory pressures than normal may be required due to the mass effect of the tumour on the lungs, as would apply to any intra-abdominal pathology such as bowel obstruction that pushes up on the diaphragm. If the chest is not moving well, recheck the position of the endotracheal tube and adjust the inspiratory pressure; this should be undertaken as a priority rather than waiting for desaturation or carbon dioxide retention to occur.

Two large bore intravenous catheters should be inserted into the upper limbs for surgery. The cannulas are placed in the hands or arms because the tumour could involve the inferior vena

cava (IVC), which will then need to be clamped for surgery. If the IV access is in the legs in this situation you would not be able to transfuse fluid to reach the central circulation and your lines would be useless.



Figure 3. Anaesthetic set up for Wilms' Tumour. Note peripheral IV lines x2 in the arms, arm at 90 degrees to keep an infusion port next to the anaesthesia provider (no extension tube available); bladder catheter to monitor urine output, eyes taped. Note, a sterile small (6-8 Fr) nasogastric tube can be used if there is no urinary catheter available

During the surgical exposure of the tumour, the surgical team could decrease venous return to the heart by compression of the IVC or by torsion on the liver, which would result in a sudden drop in the blood pressure without any signs of blood loss. You must watch the surgery closely so that you can anticipate blood loss and be aware of the manipulation of the tumour; you should alert the surgeons when the blood pressure drops. There will be times when you need to have the blood in the room and be ready to transfuse. If you are warming the blood in a bath of warm water, make sure that it is not too hot; if you cannot keep your hand in the water for more than 5 seconds then it is too hot and must not be used as you can cause haemolysis and massive infusion of potassium. Remember that 98% of the potassium in blood is intracellular; if the blood becomes haemolysed, the potassium will flood out of the cells and cause arrhythmias and even cardiac arrest when you transfuse the blood. With this child, the blood will need to be given in a 30-60 ml syringe, so that you can keep an accurate measurement of blood transfusion volume. Ideally, place a three-way stop cock in the infusion line, which will allow you to keep the syringe attached and to aspirate from the bag and infuse into the patient without a break in the blood line.

Children having major tumour excision need to have a urinary catheter inserted. An arterial line is not possible in many

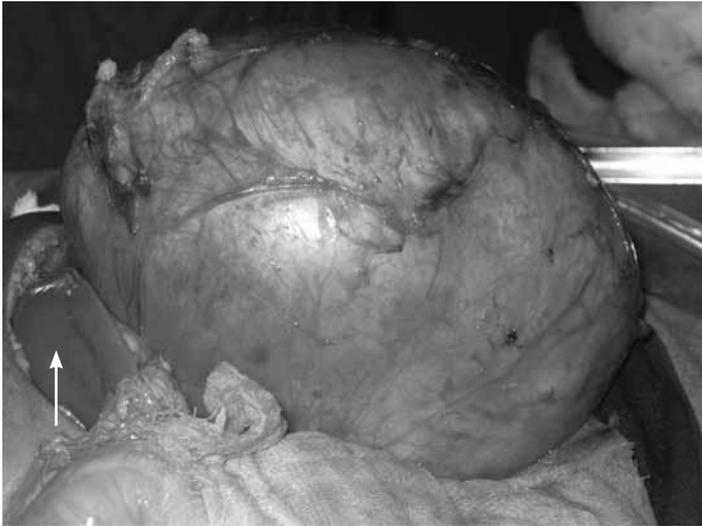


Figure 4. Wilm's Tumor intraoperative manipulation: liver compression (left side of photo - arrowed) can decrease venous return to the heart and the blood pressure will decrease, which should prompt communication with the surgical team



Figure 5. Wilm's tumour, pathological specimen: note the tumour located in the upper regions of the kidney and the abnormal tissue in the upper aspect

settings, so accurate non-invasive blood pressure monitoring needs to be done every two minutes, ideally using an automated cuff. As one can see in the pathological specimen, these tumours will involve a large section of the kidney and one can see haematuria at times. In cases of bilateral tumour involvement, the surgeons may need to do renal sparing procedures (hemi-nephrectomy), which can be associated with very large blood loss and high risk for renal dysfunction postoperatively.

If the surgery is successful and the blood loss is minimal, the child can be extubated at the end of the procedure but needs to have good pain management. It is helpful if, in addition

to opioids given during the procedure, the surgeon infiltrates the wound edges with a safe dose of local anaesthetic at the end of surgery (lignocaine 2% 3mg.kg⁻¹ or bupivacaine 0.25% 2mg.kg⁻¹). The surgery will be associated with significant postoperative pain, which should be managed by small doses of morphine or pethidine titrated to effect in the recovery room. If morphine is the drug available, a dose of 0.05mg.kg⁻¹ should be given then an observation period before adding to reach a maximum doses of 0.1mg.kg⁻¹ every 4-6 hours depending upon the patients condition. Postoperatively, these patients need to be observed in a setting with a higher nurse to patient ratio, with a bed that can have the head elevated, oxygen in the room, and careful monitoring of fluid intake and output by the nursing team. If close observation is not possible, intramuscular opioids, at the appropriate dose, may be safer than intravenous narcotics in these settings. The appropriate dosing based upon accurate weight is critical when dealing with the paediatric surgical patient. The surgeons will usually request a nasogastric tube to be inserted as the child is likely to have a postoperative ileus after this large intra-abdominal tumour is removed.



Figure 6. Wilm's tumour incision. Postoperative pain management after upper abdominal surgery will require careful treatment, with small doses of opioids titrated to effect, and close monitoring of respiratory rate by the ward nurses

Case 2

A 6-year-old female living in a very rural and resource poor area of Africa has had a one year history of abdominal swelling which has not responded to medical treatment. She is afebrile, with normal vital signs, no past medical or surgical history, and neither the family nor the medical facility has access to CT or MRI. She has travelled for two days to for a surgical consult by your outreach team as the area she lives in has minimal

access to paediatric surgery. A portable ultrasound machine revealed a large intra-abdominal cystic mass and the surgeon would like to proceed to surgery. The hospital is without piped gases or oxygen tanks, no anaesthesia machines, and has one electrically powered oxygen concentrator that produces flow up to 6 litres.minute⁻¹. The lighting is poor, but there is a diesel generator available. The child weighs 17kg.

Preoperative evaluation and plan

The surgical skill level available in the rural hospital is critical when considering this type of surgery. Is this an experienced surgeon who can adjust to the environment and will be able to retreat and stop surgery if direct visualization of the mass demonstrates a very difficult excision? Can the surgeon operate safely and efficiently (i.e. fast)? You need to consider these types of questions when working in extremely remote regions with challenging surgical and anaesthesia cases.

The only laboratory test needed in this scenario would be a haemoglobin level and a type and crossmatch for one unit of blood. If payment for treatment is required, do not spend the family's limited funds on laboratory tests that will not change your anaesthetic management. In even the very remotest of locations, there should be a blood bank for family donors and a pharmacy near the hospital that sells the blood and IV giving sets for the family to purchase and bring to the operating theatre. A laboratory that is able to check for HIV, hepatitis, malaria and blood typing would be beneficial. Always remember in an emergency situation a full cross match does not need to be done; give type-specific blood if the patient's vital signs cannot wait for the full cross match. This specific case would prompt the purchase of two blood giving sets so that if bleeding occurs and one filter blocks, you would have a secondary giving set ready.

Induction and maintenance of anaesthesia

A suitable anaesthesia plan in this situation would be total intravenous anaesthesia (TIVA) (combination of ketamine and propofol), and endotracheal intubation. A peripheral IV catheter is placed, 18 or 20 gauge, and intravenous induction performed with succinylcholine and thiopental, propofol or ketamine. Succinylcholine has a short duration of action, which will allow for spontaneous respiration to return quickly and avoid the need for positive pressure ventilation. This allows for a greater margin of safety in case the generator powered oxygen concentrator malfunctions and you are forced to use a self-inflating "Ambu" bag with room air only. Any size "Ambu" bag will be suitable, but take care to avoid excessive tidal volumes if you use an adult "Ambu" bag for a small child. Monitor the chest expansion carefully and do not squeeze the bag totally: note that some "Ambu" bags can give up to 2 litres of volume.



Figure 7. Abdominal mass and remote anaesthesia and surgery. There is no anaesthesia machine, and anaesthesia for exploratory laparotomy is delivered using an oxygen concentrator and propofol/ketamine TIVA mixture.

The propofol and ketamine mixture for this rural anaesthesia TIVA technique ("Ketofol") can be made by adding 20ml propofol 10mg.ml⁻¹ (200mg) and 2ml ketamine 100mg.ml⁻¹ (200mg) to the burette of a paediatric buretrol (microdrop) intravenous giving set. The concentration of propofol (10 mg.ml⁻¹) and the ketamine (100 mg.ml⁻¹) allows one to combine 20mls of propofol and 2mls of ketamine in an approximate 1:1 mg:mg combination for infusion, which simplifies the dosing. Each ml will have 10mg of propofol and 10 mg of ketamine. Most paediatric buretrols have 60 drops of fluid being equivalent to 1ml of fluid which translates to the infusion rates in the table. Confirm the dropper calibration with your specific buretrol being used.

At times, you may need a small dose of muscle relaxant (succinylcholine) but most surgeons can operate with a spontaneously ventilating patient. The goal in the remote environment is to maintain spontaneous breathing, which will add a level of safety. The TIVA technique could be combined with a spinal anaesthetic if the anticipated blood loss is minimal. This would allow for a lower dose of the ketofol, which would be less expensive.

Decrease the infusion rate 15-20 minutes before the projected completion of surgery and stop completely 5 minutes before the end. The patient should be extubated awake, and should be monitored closely postoperatively in the bed closest to the nursing station for 48 hour postoperatively, ideally in a bed that can elevate the head to 30 degrees. Intestinal perforation is a potential risk for this procedure and should be considered if there is any concern about sepsis postoperatively.

Table 1: "Ketofol" doses²

Phase of anaesthesia	Dose in ml	Dose in buretrol drops assuming 60 drops = 1ml
Mean induction dose	0.2 ml.kg ⁻¹	12 buretrol drops.kg ⁻¹
Maintenance infusion: deep sedation (e.g. orthopaedic procedures)	0.2 ml.kg ⁻¹ .hour ⁻¹	12 buretrol drops.kg.hour ⁻¹
Maintenance infusion: anaesthesia for laparotomy	0.3-0.4 ml.kg ⁻¹ .hour ⁻¹	18-24 buretrol drops.kg.hour ⁻¹
Incremental increases in infusion rate if the patient demonstrates signs of pain [respiratory rate, heart rate, movement]	0.1 ml.kg.hour ⁻¹ extra	6 buretrol drops.kg.hour ⁻¹ extra



Figure 8. Mesenteric cysts. Intestinal damage, intraoperative blood loss and postoperative ileus need to be considered in the perioperative plan

SUMMARY

Children presenting for elective paediatric surgery in sub-Saharan Africa may have a high morbidity and mortality.³ Understanding the basic anatomy, physiology and pharmacology together with appropriate equipment for the

paediatric patient will improve the anaesthesia mortality statistics. The development of paediatric surgical centres in both the rural and urban settings will allow for greater experience to be obtained in paediatric anaesthesia, which will improve care. The most valuable asset for these paediatric centres is to have well-trained physicians and nurses who can provide high quality care for children with the advanced surgical pathology encountered, taking account of the lack of infrastructure and the limited supplies that are a common problem. A successful perioperative course can be expected even for children requiring surgical intervention in austere environments if the basic foundations of anaesthesia are adhered to and if there is a high level of surgical skill available.

REFERENCES:

1. Hadley LG, Rouma BS, Saad-Eldin, Y: Challenge of pediatric oncology in Africa. *Seminars in Pediatric Surgery* 2012; **21**: 136-141.
2. Weatherall A, Venclovas R: Experience with a propofol-ketamine mixture for sedation during pediatric orthopedic surgery. *Pediatric Anesthesia* 2010; **20**: 1009-16.
3. Walker IA, Wilson IH: Anaesthesia in developing countries - a risk for patients. *Lancet* 2008; **371**: 968-9.

Anaesthesia for paediatric ear, nose, and throat surgery

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ADENOTONSILLECTOMY

Tonsillectomy is one of the most frequently performed surgical operations in children. According to the Department of Health Hospital Episode Statistics (<http://www.hesonline.nhs.uk>), 25 000 tonsillectomies and 6500 adenoidectomies were performed in children under 15 years of age in England in 2003.⁴ The tonsils and adenoids are lymphoid tissues forming part of the Waldeyer's ring encircling the pharynx. They appear in the second year of life, are largest between 4 and 7 years of age and then regress. Children with adenotonsillar hypertrophy can present with nasal obstruction, recurrent infections, secretory otitis media and deafness (secondary to Eustachian tube dysfunction), and obstructive sleep apnoea (OSA). Tonsillectomy is indicated in children with recurrent tonsillitis if they have had five or more episodes of sore throat per year because of tonsillitis, or if symptoms have persisted for at least 1 year and are disabling, that is, interfering with normal functioning (SIGN publication no. 34, available from <http://www.sign.ac.uk>). Other indications for tonsillectomy include chronic tonsillitis, peritonsillar abscess, and OSA. Adenoidectomy is indicated when there is evidence of enlarged adenoids causing nasal obstruction, OSA, or hearing loss. In the presence of OSA, adenotonsillectomy eliminates obstruction in 85 – 95% of children, yielding improvement of symptoms and quality of life.

Preoperative assessment

In your preoperative assessment you need to elicit features of OSA, especially in the younger child, in whom obstructive symptoms rather than recurrent infections are commonly the indication for surgery (prevalence of OSA 1 – 3%).

Symptoms of OSA include heavy snoring,

apnoeas, restless sleep, extended neck position during sleep, and daytime hypersomnolence. Over time, this can lead to neurocognitive impairment, behaviour problems, failure to thrive, and rarely cor pulmonale.

Children with severe OSA have a higher incidence of perioperative complications and may need postoperative HDU/ICU care. Specifically, they are at an increased risk of desaturation, laryngospasm, and developing airway obstruction during induction of anaesthesia.² They have increased sensitivity to the respiratory depressant effects of sedatives and opioids and a diminished ventilatory response to CO₂ compared with normal.^{2,3} The overall incidence of postoperative respiratory complications in children with severe OSA is 16 – 27% compared with an incidence of 1% in children without OSA. Other risk factors for respiratory complications include age >3 years, craniofacial abnormalities, neuromuscular disorders, failure to thrive, and obesity.³

Preoperative investigations are not routinely indicated for patients undergoing adenotonsillectomy (NICE Guideline on Preoperative Tests, available from <http://www.nice.org.uk>). It is difficult to confirm the diagnosis and quantify the severity of OSA. The gold standard for diagnosis is nocturnal polysomnography, but there is a great deal of variability in scoring methods between different sleep laboratories, and the test is expensive to perform.

Recent studies suggest that overnight oximetry to score the frequency and depth of desaturation events may be useful in identifying patients with severe OSA.⁴ In children with long-standing OSA, a full blood count will reveal polycythaemia and an ECG may show a right ventricular strain pattern.

Summary

Children account for approximately one-third of all patients undergoing ear, nose, and throat (ENT) surgery. Procedures range from simple day-case operations, such as myringotomy, to complex airway reconstruction surgery undertaken in specialist centres. This article describes the anaesthetic management of some of the commonly performed paediatric ENT procedures, including adenotonsillectomy, oesophagoscopy, and middle ear surgery. Paediatric bronchoscopy is discussed in detail in a separate article, page 173.

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Anaesthetic considerations

The main areas of anaesthetic concern are airway management, provision of analgesia, and prevention of postoperative nausea and vomiting (PONV).

Airway management

Sharing the airway with the surgeon, remote access, and the need to prevent soiling of the respiratory tract are factors that need to be taken into consideration in airway management. Two techniques are commonly used: the tracheal tube and the reinforced laryngeal mask airway (LMA).^{5,6} The advantages and disadvantages of these techniques are compared in Table 1.

The tracheal tube provides a definitive airway, and a 'south-facing' RAE tube positioned in the midline provides good surgical access. The disadvantages of intubation are that muscle paralysis or a deep plane of anaesthesia are required, bronchial intubation or accidental extubation can occur with surgical movement of the neck, and there is variable protection against airway soiling. The dilemma of whether to extubate the patient when fully awake and able to protect their airway or still deeply anaesthetized to avoid a stormy emergence and bleeding always exists. The reinforced LMA offers a good airway with no soiling of the respiratory tract, avoidance of the use of neuromuscular blocking agents, smooth emergence, and airway protection until awake. To avoid soiling the laryngeal inlet, the LMA should be removed with the cuff still inflated. To ensure best surgical access, the smallest LMA for size should be used, and when positioned correctly, the cuff should not be visible once the Boyle-Davis gag has been opened to its fullest extent. An incorrectly sized LMA, or too large a blade on the mouth gag, can cause obstruction.

The main disadvantages of the LMA are that it does not offer the definitive airway provided by a tracheal tube and it may restrict surgical access in younger patients. However, with both the tracheal tube and the LMA, dislodgement or compression can occur during positioning of the mouth gag, and airway patency must be re-confirmed before surgery proceeds.

A postal survey of anaesthetic techniques used in paediatric tonsillectomy in the UK in 1996 – 7 suggested that only 16% of anaesthetists used the reinforced LMA routinely.⁷ I.V. induction with propofol, tracheal intubation with succinylcholine, and spontaneous ventilation with isoflurane were the commonest anaesthetic techniques.⁷ Concern about the danger of succinylcholine-induced hyperkalaemic cardiac arrest in children with undiagnosed muscle disease has led to a decline in the use of this drug for elective intubation. Alternative techniques for intubation include deep inhalation anaesthesia, combinations of propofol with a short-acting opioid, or the use of a short-acting non-depolarizing neuromuscular blocking agent during light anaesthesia.

Analgesia

Adequate postoperative analgesia is best provided with a combination of simple analgesics and small doses of opioids. Paracetamol⁸ and NSAIDs have a morphine-sparing effect. The concerns around the potential for increased perioperative bleeding with NSAIDs have largely been discounted. The exception is ketorolac, which you should avoid. Administering the simple oral analgesics before operation is safe and ensures effectiveness by the end of surgery. Alternatively, the rectal route can be used after induction of anaesthesia. However, this route is less acceptable to many patients and will not achieve

Table 1. Comparison of the LMA and the tracheal tube for tonsillectomy

	LMA	Tracheal tube
<i>Advantages</i>	Straightforward airway No soiling of airway with blood Smooth emergence Paralysis not required Airway protection until awake Minimizes trauma to the airway	More secure airway Good surgical access
<i>Disadvantages</i>	Less secure airway May impair surgical access	Risk of airway trauma Oesophageal/bronchial intubation Requires paralysis Soiling of airway with blood Problems associated with extubation

therapeutic levels by the end of surgery in most cases.

A single dose of dexamethasone $0.1 - 0.5\text{mg.kg}^{-1}$ has also been shown to reduce postoperative analgesic requirements, whereas local anaesthetic infiltration of the tonsillar bed has not been found to be superior to placebo. Regular doses of paracetamol and an NSAID after operation provide good analgesia. Recently, concerns have been raised about respiratory depression and even death following use of codeine for postoperative analgesia. The current UK recommendation is to avoid codeine in any child with history of OSA undergoing (adeno)tonsillectomy, and in any child under 12 years old.⁹

Prevention of postoperative nausea and vomiting (PONV)

The incidence of PONV can be as high as 70% after adenotonsillectomy and a multimodal approach is indicated to combat this.

Minimizing starvation, avoiding the use of nitrous oxide (N_2O), and balanced analgesia with prophylactic administration of antiemetics reduce the incidence of PONV. A combination of ondansetron $0.1 - 0.2\text{mg.kg}^{-1}$ and dexamethasone $0.1 - 0.5\text{mg.kg}^{-1}$ (maximum 8mg) intraoperatively has been shown to greatly reduce the incidence of PONV.⁹ Intraoperative fluid administration has also been shown to decrease the incidence of postoperative nausea. Rescue antiemesis can be provided by further doses of ondansetron with or without cyclizine $0.5 - 1\text{mg.kg}^{-1}$ (up to 50mg).

Special considerations

Severe OSA

In general, sedative premedication and long-acting opioids are best avoided in patients with severe OSA. Inhalation induction is preferred, as airway obstruction commonly occurs during induction, and children with associated craniofacial anomalies may prove to be difficult to intubate.² Consideration should be given to the use of a small dose of fentanyl to supplement simple analgesia, as this is associated with less postoperative respiratory depression.

The incidence of complications varies with the time of day that the procedure is performed. Children undergoing surgery in the morning have fewer desaturations than those undergoing the same procedure in the afternoon. Close postoperative monitoring and the availability of an ICU bed is required.

Day-case tonsillectomy

Successful and safe implementation of day-case tonsillectomy requires careful patient selection. Exclusion criteria include age >3 years, significant co-morbidity, OSA, and living further than a one hour drive from the hospital or having no private transport. Thought also needs to be given to the risk of early

haemorrhage and the management of postoperative pain and PONV.

The incidence of early postoperative bleeding is $<1\%$ and the majority of these occur within the first 4h after surgery. An extended observation period of 4 – 6h before discharge is therefore recommended; this limits surgery to morning lists. A multimodal analgesic and antiemetic regimen as previously discussed is very important, as the main reasons for overnight admission are PONV, pain, and poor oral intake.

Bleeding tonsil

Haemorrhage is the most serious complication after tonsillectomy and can occur within the first 24h (primary haemorrhage) or up to 28 days after surgery (secondary haemorrhage). In the National Prospective Tonsillectomy Audit (July 2003 – September 2004), the incidence of post-tonsillectomy haemorrhage patients was 3.5% and the overall rate of return to theatre was 0.9%. The incidence of primary haemorrhage was 0.6% and the majority of these occurred within the first 6h after operation. Factors influencing haemorrhage rates were age (lower rates in children than adults), indication for surgery (highest rates with quinsy and recurrent tonsillitis, lowest with obstructive symptoms), and surgical technique (higher rates with use of diathermy and disposable equipment, lowest with blunt dissection).

The anaesthetic considerations in bleeding tonsil include hypovolaemia, the risk of pulmonary aspiration (swallowed blood with or without oral intake), potential for a difficult intubation because of excessive bleeding obscuring the view with or without oedema after earlier airway instrumentation, a second general anaesthetic, and the stress to both child and parents. Blood loss is because of venous or capillary ooze from the tonsillar bed and is difficult to measure, as it occurs over several hours and is partly swallowed.

Excessive blood loss may lead to the child spitting blood. In these cases, the child is likely to be seriously hypovolaemic, anaemic, and potentially difficult to intubate because of poor visualization of the larynx. Tachycardia, tachypnoea, delayed capillary refill, and decreased urine output are early indicators of hypovolaemia, whereas hypotension and altered sensorium are indicators of advanced volume depletion. Preoperative resuscitation (guided by trends in monitoring) is essential, even if this requires the insertion of an interosseous needle. Induction of anaesthesia in a hypovolaemic child can precipitate cardiovascular collapse. Haemoglobin and coagulation variables should be checked. Blood and blood products should be immediately available and transfused as necessary. Before induction, in addition to the standard equipment, a selection of laryngoscope blades, smaller than expected tracheal tubes, and two suction catheters should be immediately available. Anaesthesia is induced once the

child is haemodynamically stable. Preoxygenation and rapid sequence induction with slight head-down positioning of the patient ensures rapid control of the airway and protection from pulmonary aspiration. Consider adopting the left lateral position if bleeding is excessive. Controlled ventilation provides good conditions for haemostasis.

Fluid resuscitation and transfusion of blood and blood products should continue intraoperatively as necessary. Once haemostasis is achieved, a large-bore stomach tube is passed under direct vision and the stomach emptied. Neuromuscular block is antagonized and the trachea is extubated, with the child fully awake in the recovery position. After operation, the child should be monitored closely for any recurrence of bleeding.

OESOPHAGOSCOPY

Rigid oesophagoscopy is performed for the removal of an ingested foreign body. History of ingestion, dysphagia, and odynophagia are the usual presenting symptoms, whereas a previous stricture is a predisposing factor for obstruction. The commonest site of impaction of the foreign body is at the level of the cricopharyngeus muscle. Oesophagoscopy should be performed in all cases of suspected impacted foreign body to prevent complications of perforation, mediastinitis, and fistula formation.

Anaesthetic considerations include management of the shared airway and the risk of pulmonary aspiration or oesophageal perforation during the procedure. A rapid sequence induction protects against pulmonary aspiration and ensures rapid control of the airway. The tracheal tube should be secured on the left side to allow easier access for the endoscopy. Adequate depth of anaesthesia and muscle relaxation during the procedure are essential to reduce the risk of oesophageal perforation. Analgesia is provided by a combination of intravenously or rectally administered simple analgesics and a small dose of opioid. The patient is extubated when fully awake. If oesophageal perforation is suspected, withhold oral intake, commence IV antibiotics, and observe the patient closely for features of mediastinitis, such as severe chest pain, pyrexia, and subcutaneous emphysema.

EAR SURGERY

The most common surgical procedures on the ear are those performed to treat otitis media and its complications. Otitis media is the second most prevalent illness of childhood. This is because of a combination of factors including Eustachian tube dysfunction and an increased susceptibility to upper respiratory tract infection (URTI) in early childhood. The short Eustachian tube in young children predisposes to reflux of nasopharyngeal secretions into the middle ear space and thus to recurrent infections. Oedema of the Eustachian tube mucosa

secondary to recurrent URTI, and mechanical obstruction of the Eustachian tube orifice by enlarged adenoids, lead to a negative pressure in the middle ear and a transudative effusion (secretory otitis media). Children with otitis media present with deafness and complications such as perforation, ossicular chain damage, and cholesteatoma. Surgery is performed to improve hearing and to eradicate middle-ear disease.

MYRINGOTOMY

Myringotomy and insertion of pressure-equalizing tubes are used to improve middle-ear aeration and hearing in chronic otitis media. It is a short procedure performed as a day-case. The preoperative assessment should elicit features of URTI, as otitis media is associated with recurrent URTI and these children can consequently have increased airway irritability. A small percentage of this population may also display symptoms of OSA secondary to adenoidal hypertrophy. The anaesthetic technique usually involves the patient breathing spontaneously via a facemask or LMA, with the head positioned to one side. Mild postoperative pain can occur in up to 75% of patients, but this can be avoided with the preoperative administration of paracetamol, NSAIDs, or both.¹¹

MYRINGOPLASTY, TYMPANOPLASTY, AND MASTOIDECTOMY

Children with complications of chronic otitis media need more complex ear surgery. Myringoplasty involves repair of a tympanic membrane perforation in a dry ear. Tympanoplasty is performed when there is extensive middle-ear damage and involves reconstruction of the tympanic membrane and the ossicular chain. The approach to the ear can be permeal or postaural, the latter providing better surgical access. Two surgical techniques of tympanic membrane grafting are used, the underlay and the overlay. The underlay technique involves elevation of a tympanomeatal flap and placing the graft material underneath (or medial to) the eardrum.

The overlay technique involves stripping the lateral epithelium off the eardrum and placing the graft material on the outer side of (or distal to) the eardrum. Various graft materials may be used, the most common being temporalis fascia, tragal perichondrium, and fat.

Mastoidectomy is performed to eradicate chronic suppurative middle-ear disease. The anaesthetic considerations associated with these three procedures are similar; therefore, we shall describe their anaesthetic management collectively.

Anaesthetic considerations

Typically, these procedures are performed in the older child or teenager and can be of prolonged duration. The main factors that have a bearing on anaesthetic management are the effect of N₂O on the middle ear, the need for a bloodless operative

field, the use of facial nerve monitoring by the surgeon, and the high associated incidence of PONV.

As the relative solubility of N₂O in blood is 34 times that of nitrogen, it diffuses across into the non-compliant middle-ear cavity much more rapidly than nitrogen can leave. This can lead to pressures as high as 350 mm H₂O within 30 min of commencing N₂O, especially in the presence of Eustachian tube dysfunction.¹²

Displacement of tympanoplasty grafts, worsening of deafness, rupture of the tympanic membrane, and increased PONV have all been associated with elevated middle-ear pressures. In addition, after discontinuation of N₂O, rapid re-absorption of the gas leads to negative pressures in the middle ear and this can lead to 'lifting off' of the underlay tympanic membrane graft. As the middle ear remains open until the surgeon places the graft over the tympanic membrane, N₂O can be used up to 10 – 15min before graft placement and then discontinued. However, it may be best to avoid its use in middle-ear surgery completely.

Any bleeding during middle-ear surgery distorts the view through the operating microscope and can make the procedure difficult. Venous ooze can be minimized by a head-up tilt of 10° – 15° and ensuring unimpeded venous drainage. Epinephrine infiltration by the surgeon, relative hypotension (mean arterial pressure 10 – 20%, normal), and avoidance of tachycardia minimize arterial bleeding.

In its course through the temporal bone, the facial nerve runs through the middle ear in close relation to the ossicles and through the mastoid before emerging from the stylomastoid foramen. Therefore, it is vulnerable to damage during middle-ear surgery, especially as the disease process can distort the anatomical relationship of the nerve to the ear structures and make identification difficult. Intraoperative facial nerve monitoring is useful for identification and preservation of the nerve during ear surgery. A single dose of a short-intermediate acting relaxant can be used to aid tracheal intubation, its effects should have worn off sufficiently before the stage in the operation when facial nerve monitoring is required. However, it may be prudent to avoid the use of relaxants altogether by using other agents to facilitate intubation or by avoiding intubation. Whether using a tracheal tube or an LMA, the patient requires controlled ventilation for this procedure. Much of the surgery is performed using an operating microscope; therefore, if paralysis is to be avoided, a deep plane of anaesthesia is required to guarantee immobility. Controlled ventilation also allows control of the end-tidal CO₂, which helps to minimize bleeding.

The options for airway management are a tracheal tube or a

reinforced LMA. The advantages of a tracheal tube over an LMA are a secure airway and ease of controlled ventilation, though a stormy emergence contributing to graft displacement is a potential problem. Smoother emergence can be ensured by tracheal extubation in a deep plane of anaesthesia. A reinforced LMA has the potential advantages of less airway stimulation and smooth emergence, but care must be taken to limit airway inflation pressures in order to prevent gastric distension during controlled ventilation.

For either technique and where available, maintenance of anaesthesia with propofol and remifentanyl, or sevoflurane and remifentanyl, offers many advantages. They allow controlled ventilation without neuromuscular blocking agents, thus permitting unimpeded facial nerve monitoring. Remifentanyl provides a titratable degree of hypotension while maintaining a stable heart rate and provides excellent operating conditions. The use of TIVA is also associated with a lower incidence of PONV.¹³

Analgesia and antiemesis

A multimodal approach provides good analgesia and minimizes opioid-induced PONV. Oral paracetamol and NSAIDs given before operation are better accepted by the older child; alternatively, these can be given rectally or intravenously during surgery. As remifentanyl has no residual analgesic effect after termination of the infusion, a small dose of morphine should be given 30 – 40 min before the end of the procedure to ensure adequate analgesia on awakening. A greater auricular nerve block has been shown to reduce postoperative opioid requirement. Postoperative analgesia is provided by regular, simple analgesics and small doses of opioids if necessary.

Routine prophylactic ondansetron and dexamethasone are indicated because of the emetogenic potential of middle-ear surgery. Avoiding prolonged starvation, adequate hydration, avoiding N₂O, use of TIVA, and balanced analgesia also help decrease PONV.

BONE-ANCHORED HEARING AID

The bone-anchored hearing aid (BAHA) is a surgically implantable system for the treatment of conductive deafness in children with chronic ear infections or congenital external auditory canal atresia who cannot benefit from conventional hearing aids. It allows sound to be conducted through the bone rather than via the middle ear, a process known as direct bone conduction. The procedure involves two short operations. Firstly, a titanium fixture is implanted into the mastoid bone and this over time integrates with the bone of the skull. Around 6 months later, at a second operation, an external abutment is placed over the fixture and this allows a sound processor to be connected.

The majority of children presenting for BAHA implant have associated congenital anomalies, the commonest being Goldenhar's syndrome (26%) and Treacher Collins syndrome (21%).¹³ There is also a high incidence of congenital heart disease (19%) and craniofacial anomalies. The main anaesthetic concern is an increased incidence of difficult intubation. In most instances, after inhalation induction, the airway can be safely and easily maintained using a reinforced LMA. However, equipment for fiberoptic intubation and appropriately trained staff should be available in the event of a need for intubation. Analgesia is provided with a combination of paracetamol, NSAID, and a small dose of opioid. Routine antiemetics are indicated, as PONV is common.

REFERENCES

Ravi R, Howell T. Anaesthesia for paediatric ear, nose and throat surgery. *Update in Anaesthesia* (2008), **24**(1):18-23. Available from: http://www.wfsahq.org/components/com_virtual_library/media/cf2070c8974e043763936f7aeffad69-6922932df850570c39edd579d02ab2bf-Anaesthesia-for-Paediatric-ENT-Surgery--Update-24-1-2008-.pdf

- Dix P. Bronchoscopy for a foreign body in a child. See article in this edition, page 173.
- Warwick JP, Mason DG. Obstructive sleep apnoea syndrome in children. *Anaesthesia* 1998; **53**: 571 – 9.
- Rosen GM, Muckle RP, Mahowald MW, Goding GS, Ullewig C. Postoperative respiratory compromise in children with obstructive sleep apnoea syndrome: can it be anticipated? *Pediatrics* 1994; **93**: 784 – 8.
- Nixon GM, Kermack AS, Davis GM, Manoukian JJ, Brown KA, Brouillette RT. Planning adenotonsillectomy in children with obstructive sleep apnoea: the role of overnight oximetry. *Pediatrics* 2004; **113**: 19 – 25.
- Williams PJ, Bailey PM. Comparison of the reinforced laryngeal mask airway and tracheal intubation for adenotonsillectomy. *Br J Anaesth* 1993; **70**: 30 – 3.
- Webster AC, Morley-Forster PK, Dain S, Ganapathy S, Ruby R, Au A, Cook MJ. Anaesthesia for adenotonsillectomy: a comparison between tracheal intubation and the armoured laryngeal mask airway. *Can J Anaesth* 1993; **40**: 1171 – 7.
- Hatcher IS, Stack CG. Postal survey of the anaesthetic techniques used for paediatric tonsillectomy surgery. *Paediatr Anaesth* 1999; **9**: 311 – 5.
- Korpela R, Korvenoja P, Meretoja OA. Morphine-sparing effect of acetaminophen in pediatric day case surgery. *Anesthesiology* 1999; **91**: 442 – 7.
- Guidance for the administration of codeine and alternative opioid analgesics in children: Joint guidance from the RCoA, APA and RCPCH. Published November 26th 2013 Available from: http://www.apagbi.org.uk/sites/default/files/images/Codeine_Nov2013.pdf
- Steward DL, Welge JA, Myer CM. Steroids for improving recovery following tonsillectomy in children. *Cochrane Database Syst Rev* 2003; 1: CD003977.
- Watcha MF, Ramirez-Ruiz M, White PF, Jones MB, Lagueruela RG, Terkonda RP. Perioperative effects of oral ketorolac and acetaminophen in children undergoing bilateral myringotomy. *Can J Anaesth* 1992; **39**: 649 – 54.
- Chinn K, Brown OE, Manning SC, Crandell CC. Middle ear pressure variation: effect of nitrous oxide. *Laryngoscope* 1997; **107**: 357 – 63.
- Mukherjee K, Seavell C, Rawlings E, Weiss A. A comparison of total intravenous with balanced anaesthesia for middle ear surgery: effects on postoperative nausea and vomiting, pain and conditions for surgery. *Anaesthesia* 2003; **58**: 176 – 80.
- Jones SEF, Dickson U, Moriarty A. Anaesthesia for insertion of bone- anchored hearing aids in children: a 7 year audit. *Anaesthesia* 2001; **56**: 777 – 98.

Anaesthesia for paediatric eye surgery

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INTRODUCTION

Unlike adults, children requiring eye surgery do not tolerate sedation or local anaesthetic techniques and therefore almost always require general anaesthesia. This update will present a general review of the principles of anaesthesia for children undergoing eye surgery and a description of anaesthesia for some specific procedures.

GENERAL PRINCIPLES OF ANAESTHESIA FOR PAEDIATRIC EYE SURGERY

Preoperative considerations

Most children presenting for eye surgery are healthy, ASA I or II and may be managed as day cases. A small number have underlying conditions, often of a chromosomal or metabolic nature, which pose more specific anaesthetic challenges.^{1,2} Examples of these are described in appendix 1.

Ophthalmic medications

Many children requiring eye surgery receive eye drops. Knowledge of commonly used drugs and potential side effects is useful (See table 1). Medications may be absorbed through the pharyngeal mucosa via the nasolacrimal ducts to cause systemic effects, although this is rarely a significant problem.^{2,3,4}

Anaesthetic considerations

Premedication and induction of anaesthesia

The decision to premedicate the child and the choice of induction technique, intravenous (IV) or inhalational, should be tailored to the needs of the child and to the preferences of the anaesthetist. Children with visual impairment should be handled in a careful and sensitive manner.

Airway management

Airway management should be tailored to the procedure. For measurement of intraocular pressure (IOP), spontaneous respiration via a facemask should be used, as intubation will raise the intraocular pressure. For simple procedures such as examination under anaesthesia (EUA) it may be more convenient to maintain spontaneous respiration through a reinforced laryngeal mask airway (LMA), particularly where a sterile field is required.

The reinforced LMA may be used in older children for most eye procedures. It is possible to use controlled ventilation with muscle relaxants, and coughing is reduced at the end of the surgery.

Intraocular surgery requires a still eye with low intraocular pressure. The airway is best managed by intubation with paralysis and controlled ventilation.

Summary

- Children require general anaesthesia for ophthalmic procedures/surgery, but can generally be managed as day cases.
- The oculocardiac reflex may be induced during eye surgery and risks provoking dangerous bradycardias. Prevent these by premedicating with anticholinergic agents.
- Postoperative nausea and vomiting is common after eye surgery in children.

Table 1. Common ophthalmologic preparations and their side effects.

Eye preparations	Indication	Systemic side effect
Beta-blockers: Timolol maleate or Betaxolol hydrochloride	Glaucoma	Bradycardia refractory to atropine; bronchospasm in asthmatics
Carbonic anhydrase inhibitors: Acetazolamide (diamox)	Glaucoma	Metabolic acidosis, electrolyte abnormalities, allergies, including Stevens-Johnson syndrome
Antimuscarinic agents: Cyclopentolate or Atropine	Pupil dilatation	Dry mucous membranes, nausea and vomiting, tachycardia
Alpha-adrenergic sympathomimetic agents: Phenylephrine 2.5%	Pupil dilatation	Hypertension, tachycardia
NSAIDs: Diclofenac sodium, Ketorolac trometamol 0.5%	Pain relief	Potential to worsen or precipitate acute asthma
Local anaesthetic agents: Amethocaine (Tetracaine), Oxybuprocaine, Proxymetacaine	Pain relief or prevention	Local anaesthetic toxicity, particularly preterm neonates

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Similarly, eye surgery in very young children is best managed with intubation and controlled ventilation to ensure a secure airway. Access to the airway will be restricted during the surgery so it is important to secure the tracheal tube firmly. A preformed south facing RAE tube is ideal, but this may be too long in neonates; a reinforced flexible tracheal tube (ETT) may be preferable in this situation.

Maintenance of anaesthesia

As with induction, the choice of maintenance technique rests largely on the preferences of the anaesthetist and the availability of different agents.

The incidence of dysrhythmias is increased with halothane, particularly if there is hypercapnia and eye preparations containing atropine or adrenaline are used. Isoflurane or sevoflurane may be preferable.

Propofol has anti-emetic effects. Total intravenous anaesthesia (TIVA) with propofol reduces the risk of postoperative nausea and vomiting (PONV). Remifentanyl can reduce volatile requirements.^{5,6}

Nitrous oxide is of limited value in eye surgery as it increases PONV and diffuses into gas filled spaces. It should be avoided in vitreoretinal detachment surgery where intraocular gas bubbles of sulphur hexachloride or perfluoropropane are introduced into the eye to tamponade detached surfaces as it will cause significant rise in intraocular pressure. It should also be avoided for any patient who has undergone recent vitreoretinal detachment surgery as the bubble may last several weeks. Alternatively, if nitrous oxide was used from the start of the anaesthetic, prior to placement of the gas bubble, it will diffuse out of the bubble on completion of the anaesthetic and increase risk of re-detachment.

Anaesthetic agents and intraocular pressure

Normal intraocular pressure (IOP) ranges from 10 to 20mmHg. Most anaesthetic agents will decrease this. Table 3 describes the effects of commonly used anaesthetic agents on IOP.^{7,8,9,10,11,12} If serial measurements of IOP are being made, it is important to be consistent with the type of anaesthetic used on different occasions.

Anaesthetic techniques and intraocular pressure

Physical and physiological variables have an important effect on IOP.

Laryngoscopy, coughing, straining, crying, bucking and the process of tracheal extubation may all cause a rise in IOP. This effect may be attenuated by lidocaine 1mg.kg⁻¹ IV 3 minutes prior to intubation or extubation. Use of the LMA allows smoother induction and emergence from anaesthesia and has much less effect on IOP.^{13,14,15}

Hypoxia and hypercapnia both increase IOP.

Hypocapnia and hypothermia decrease IOP.

The oculocardiac reflex

The oculocardiac reflex is common during eye surgery in children, and is seen in up to 60% of children undergoing strabismus surgery. It is essential to use continuous heart rate monitoring with an ECG during eye surgery in children. The reflex takes its afferent innervations from the ophthalmic division of the trigeminal nerve, relays via the sensory nucleus in the 4th ventricle, with the efferent impulse passing through the vagus nerve.^{1,2}

Surgical traction on the extra-ocular eye muscles or pressure on the globe causes a sinus bradycardia, and occasionally junctional rhythms, atrioventricular block, atrial ectopics or ventricular ectopics. The reflex is most commonly induced by traction on the medial rectus muscle, rather than the smaller lateral rectus muscle. The bradycardia resolves almost immediately after the stimulus has been removed and weakens with repetition of the stimulus.

Atropine 20mcg.kg⁻¹ IV or glycopyrrolate 10mcg.kg⁻¹ IV at induction will block the oculocardiac reflex. If not given at induction, it is important to have the drugs drawn up and ready to administer if bradycardia should occur.

The reflex may be attenuated by application of topical local anaesthetic agents to the eye (such as tetracaine eye drops), or by blocking the afferent limb of the reflex with a peribulbar block, although this block is not usually used in children due to the risk of globe perforation.^{16,17}

The oculocardiac reflex is less common with sevoflurane compared to halothane, and less common with deep anaesthesia compared to light anaesthesia.¹⁸ The likelihood of significant bradycardia is doubled if hypercarbia is present, so controlled ventilation should be considered. The oculocardiac reflex is more likely to occur with rocuronium compared to atracurium.¹

Table 2. Anaesthetic agents and their effects on intraocular pressure.

Anaesthetic agent	Effect on intraocular pressure
Propofol, thiopentone	IOP reduced by 20-30% (3-7mmHg)
Halothane, sevoflurane, isoflurane, desflurane	IOP reduced by 20-30% (3-7mmHg)
Opioids	Minimal to no effect on IOP
Ketamine	Dose dependent increase in IOP (minor); marked when dose exceeds 5mg.kg ⁻¹
Atropine	No effect on IOP
Non-depolarising muscle relaxants	Minimal to no effect on IOP
Suxamethonium	Significant increase in IOP within 30 sec of administration (approx 8mmHg), effect lasts for 5-7 minutes, less if given with agents that reduce IOP
Acetazolamide, mannitol, dextrans	Used for acute reduction of IOP perioperatively

Children who exhibit the oculocardiac reflex are more likely to develop PONV¹⁹ and should receive an antiemetic during anaesthesia.

Extubation and emergence from anaesthesia

It is important to avoid coughing and bucking on the tracheal tube at the end of surgery, particularly for children who have undergone intraocular surgery. For this reason, many anaesthetists use a laryngeal mask airway for eye surgery. If a tracheal tube has been used, the child should be extubated deep, if possible. If deep extubation is contraindicated, for instance if the child has a full stomach, lidocaine 1mg.kg⁻¹ IV can be given to reduce rises in IOP.

Principles of pain relief and postoperative care

Pain after eye surgery is usually mild to moderate and can be managed with simple analgesics such as paracetamol, NSAIDs and topical local anaesthetic agents. These may be given pre-emptively as oral preparations preoperatively or rectally/IV at induction.

Squint surgery, evisceration and vitreoretinal surgery is associated with more severe pain. Analgesia should include an opioid such as fentanyl IV, paracetamol, NSAIDs, and topical local anaesthetic if possible. Multimodal analgesia should be continued into the postoperative period, with the addition of codeine phosphate or tramadol, escalating to morphine if required. The use of opioids increases the risk of PONV and antiemetics are essential.

PONV is extremely common after paediatric eye surgery, and for strabismus surgery can be as high as 60% if no prophylaxis is given. The combination of ondansetron 0.15mg.kg⁻¹ IV, and dexamethasone 0.1–0.2mg.kg⁻¹ IV reduces PONV to 10% in strabismus surgery.^{20,21,22} It is wise to leave the IV cannula in place postoperatively where PONV may be a problem so that further antiemetics and IV fluids can be given.

Ketamine is associated with emergence phenomena and the child should be recovered in a quiet area with minimal stimulation.¹²

Most paediatric eye procedures are treated as day cases and children may resume oral intake as soon as they are able. Occasionally PONV results in an unplanned overnight admission.

ANAESTHESIA FOR SPECIFIC OPHTHALMIC CONDITIONS AND PROCEDURES

EUA and measurement of IOP

For an examination of the eyes under anaesthesia, either an inhalational or intravenous induction technique and airway maintenance with a facemask will suffice. It may be technically easier to place an LMA for a longer EUA.

Most anaesthetic agents decrease IOP, which may potentially mask a high IOP.

Some anaesthetists advocate the use of ketamine 1–2mg.kg⁻¹ IV or 5–10mg.kg⁻¹ IM for IOP measurements, as it does not drop IOP. Although it may slightly raise IOP, this may be safer than having a falsely low reading. Ketamine increases secretions so should be given with either atropine 20mcg.kg⁻¹ IV or glycopyrolate 10mcg.kg⁻¹ IV. Airway reflexes are maintained and instrumentation of the airway is rarely required.^{1,11,12}

Alternatively, inhalational induction may be used. The ophthalmologist should be present in the room so that the IOP can be measured as soon as the child is still. The child should not be too deeply anaesthetised, the eyes should be central and the facemask must not press on the eyes.

Regardless of the technique used, the IOP should always be measured before laryngoscopy or LMA insertion, although there is little evidence to prove that the latter significantly raises IOP. A consistent technique should be ensured if serial measurements of IOP are to be made.

Syringing and probing of nasolacrimal ducts

Children with blocked nasolacrimal ducts will usually present early in life with increased tearing. Most respond to probing of their nasolacrimal ducts, which is a short procedure for which an LMA will suffice.

Should simple probing fail, the surgeon might place a silicone catheter through the duct where it is secured for a few weeks. Alternatively the inferior turbinate bone may be fractured to relieve the obstruction.

Dacrocystorhinostomy is a more extensive procedure that involves exposure of the duct and creation of a new opening into the nasal cavity.¹

Anaesthetic considerations

The main problem is bleeding from the nasal mucosa:

- Topical vasoconstrictors reduce bleeding from the nasal mucosa.
- Hypotensive anaesthesia may be required to reduce bleeding, for instance, relatively deep anaesthesia with moderate head up tilt.
- The airway should be protected from blood, ideally with a throat pack, and the nasopharynx should be suctioned before extubation.
- Opioids may be required for analgesia for this procedure.

Strabismus surgery

Squint is a common problem that affects 3 – 5% of the population, making strabismus surgery the most commonly performed eye operation in children. It affects males and females equally.

Squints are usually idiopathic, but may also be secondary to intracerebral space occupying lesions, trauma, infection or inflammation causing muscle palsies. Most patients are healthy, but occasionally squints may be associated with a family history, prematurity, and disorders of the central nervous system such as cerebral palsy, hydrocephalus and myelomeningocele. Patients may have occult myopathies and there is a threefold increase in the incidence of masseter spasm. Anecdotal evidence of an increased association with malignant hyperpyrexia remains unproven.²

Squint correction is achieved by lengthening (recession), shortening or tightening (resection) or transposition of any of the four rectus and two oblique extra-ocular muscles, or combinations of any of the above.

Surgeons may use forced duction testing to distinguish a paretic muscle from one that has restricted motion. Botulinum toxin may be injected into the extra-ocular muscle for minor abnormalities.

This requires electromyogram (EMG) control and muscle relaxants should be avoided.¹

In older children an adjustable suture may be used that allows fine adjustments to be made 24 to 48 hours postoperatively under topical local anaesthetic once the patient is awake.

Anaesthetic considerations

- Induction technique, the method of airway control and choice of ventilation may be guided by the preference of the anaesthetist.
- TIVA with propofol reduces PONV. Alternatively, anaesthesia may be maintained with a volatile agent and air/oxygen.
- Consider atropine 20mcg.kg⁻¹ IV or glycopyrolate 10mcg.kg⁻¹ IV to block the oculocardiac reflex.
- PONV is common postoperatively, up to 50–75%. Give two anti-emetic agents such as ondansetron 0.1mg.kg⁻¹ IV and dexamethasone 0.1-0.2 mg.kg⁻¹ IV.^{20,21,22}
- Extubate the child deep if possible.
- Analgesia should include topical tetracaine or oxybuprocaine, NSAIDS such as ibuprofen or diclofenac and paracetamol, unless contraindicated.
- Intraoperative opioids should be avoided if possible.
- A peribulbar block is effective for analgesic requirements and reduces PONV, possibly by blocking the ophthalmic division of the trigeminal nerve that passes to the vomiting centre in the medulla. The risk of globe perforation in children makes most practitioners cautious of this.¹⁷
- A sub-Tenon block performed intraoperatively by the surgeon provides effective analgesia.

Glaucoma

The pressure within the eye is maintained through a balance between the production of aqueous humor, primarily by the ciliary body in the posterior chamber, and drainage via the trabecular network to the canal of Schlem in the anterior chamber.

In glaucoma the normal IOP of 10–20mmHg is elevated so that capillary blood flow to the optic nerve is reduced, which compromises the function of the optic nerve.

The causes of glaucoma are varied:

- Primary congenital glaucoma is caused by a failure of the development of the trabecular network, reducing the drainage of aqueous humor. It is bilateral in 75% of cases and has a prevalence of 1:10000 births. There is a male to female ratio of 35%: 65% and it is more common in children below the age of 3 years.
- Secondary glaucoma is usually caused by blockage of existing drainage channels by infection, inflammation or trauma. It is also seen in some rare syndromes such as Sturge-Weber Syndrome, Axenfeld Syndrome and in association with aniridia in 20% of patients with Wilm's tumour.²

Treatment may be medical or surgical. Medical treatment consists of drugs used to reduce IOP. Acetazolamide 15–30mg.kg⁻¹.day⁻¹ PO in

3 – 4 divided doses suppresses aqueous production, but its usefulness is limited since the causes of glaucoma are usually structural and related to drainage.

Surgical treatments may vary:

- Goniotomy involves visualising the anterior chamber with a gonioscope and making an incision into the trabecular meshwork to allow drainage.
- Trabeculotomy involves the insertion of a fine probe into Schlem's canal to create a new drainage channel.
- Trabeculectomy involves the creation of a new drainage channel from the anterior chamber into sub-Tenon's space where the aqueous is absorbed.
- Cyclocryotherapy is the ablation of part of the ciliary body by a cryoprobe at - 60 to - 80 degrees Celsius to reduce the production of aqueous humor.²

Anaesthetic considerations

- Avoid raising the IOP by ensuring a smooth induction and deep emergence without coughing.
- Maintain a motionless eye; consider muscle relaxants and controlled ventilation to avoid hypercapnia.
- Analgesia with paracetamol and NSAIDS is usually adequate, however when cyclocryotherapy is used opioids may be necessary.
- High incidence of PONV, give routine anti-emetics.

Cataract extraction

Cataracts are a major cause of childhood morbidity worldwide, predominantly in the developing countries of Africa and Asia (85% of cases).

Aetiology may be varied:²

- Hereditary cataracts are autosomal dominant and present in otherwise healthy children.
- Syndromes may be associated with cataracts. Some include Lowe's oculo-cerebro-renal syndrome (X-linked recessive), Down syndrome (trisomy 21), Edward syndrome and Cri-du-chat syndrome.
- Metabolic causes of cataract may include glucose-6-phosphate dehydrogenase deficiency, hypoglycaemia, hypocalcaemia and galactosaemia.
- Blunt or penetrating trauma over time may cause unilateral cataracts.
- Inflammation such as the uveitis associated with juvenile chronic arthritis may cause cataracts.
- Tumours, such as retinoblastoma can cause cataracts.
- Intrauterine infections, including rubella, cytomegalovirus (CMV), toxoplasma and toxocariasis.
- Radiation for leukaemia might cause cataracts.
- Chronic steroid use can result in cataracts.

Treatment involves surgical implantation of an intraocular lens. This needs to be done very early (as early as 4 weeks old) in order to allow

stimulation of the retina and visual development. The procedure takes about 30 - 60 minutes, but complications can be more frequent than in adults and include uveitis, glaucoma, endophthalmitis, iris damage or prolapse, retinal detachment and thickening of the posterior lens capsule.²

Anaesthetic considerations

- Aim for a motionless eye either with deep anaesthesia or muscle relaxants.
- Avoid high IOP with a smooth induction and emergence.
- Consider controlled ventilation to avoid hypercapnia.
- Give anti-emetics.

Enucleation and evisceration

Enucleation is the removal of the whole eye. This may be done for surgical treatment of a retinoblastoma, significant eye trauma or for cosmetic reasons where an eye is blind. It involves the dissection of the extra-ocular muscles off the globe. There is a similar risk here for the oculocardiac reflex as in squint surgery, although less risk of PONV. IV atropine/glycopyrrolate should be available.

Evisceration involves the removal of the contents of the globe, but retention of the sclera. This procedure is often painful and opioid analgesia will be required.¹

Penetrating eye injury

A penetrating eye injury is a relatively common injury in children, primarily boys between 3 and 9 years. Surgery is required to close the defect or remove a foreign body. Up to 30% of these injuries may be associated with trauma to the head, orbit or adnexa, and the risks of anaesthesia under these circumstances need to be weighed against the benefits of early closure.²³ Surgery is urgent, as anything that raises IOP (coughing, straining) may cause the globe to extrude its contents.^{2,3}

This presents two conflicting anaesthetic problems. First, the child may have a full stomach so a rapid sequence intubation with suxamethonium is indicated in order to prevent aspiration. Second, there is a need to protect the globe from a rise in IOP that could result in extrusion of the structures of the anterior chamber or the vitreous humor. The transient rise in intraocular pressure produced by the use of suxamethonium could theoretically cause this.²⁴

- One approach recommends the use of a large dose of non-depolarising muscle relaxant (NDMR) and ventilation with cricoid pressure until intubating conditions are achieved, providing the child has a normal airway.¹⁰
- Another view is that there have been no documented reports of vitreous extrusion after the use of suxamethonium, and protection of the airway is paramount;²⁵ hence the use of suxamethonium and a traditional rapid sequence induction is indicated.

Other considerations include:

- Crying, coughing and straining should be avoided; consider light oral sedation and analgesia preoperatively.
- Direct laryngoscopy of a poorly paralysed airway can cause coughing and bucking, whichever technique is used.

- Consider blunting the intubation response prior to laryngoscopy. Administer lidocaine 1-2 mg.kg⁻¹ IV given 3 minutes prior to rapid sequence intubation during preoxygenation.

Vitreoretinal surgery

Vitreoretinal surgery is performed for the repair of a detached retina, and although uncommon, may be necessary in children. Detachment may be primary where it is related to a defect in the retina, or secondary to an underlying illness. The surgery involves creating a choroiretinal scar with cryotherapy and placing a scleral buckle towards the back of the eye, which serves to oppose the neuroretina and retinal pigment epithelium. The surgeon may place an intraocular bubble of either sulphur hexafluoride or perfluropropane to tamponade the detached surfaces together.^{1,2}

Anaesthetic considerations

- Avoid nitrous oxide if an intraocular gas bubble is used, both during surgery and for several weeks after. Parents should be given clear instructions in this regard for future anaesthetics.
- Controlled ventilation and muscle relaxants should be used
- This procedure is painful and analgesia including opioids should be considered.
- Anti-emetics should be used routinely
- Avoid raised IOP during extubation - usually achieved by deep extubation.

CONCLUSION

This update has reviewed the general principles of anaesthesia for paediatric eye surgery, as well as considerations for some common procedures. Key learning points include:

- Children require general anaesthesia for ophthalmic procedures and surgery, but most are healthy and can be managed as day cases.
- The oculocardiac reflex may be induced during eye surgery and risks provoking dangerous bradycardias, which can be prevented by premedicating with anticholinergic agents.
- Postoperative nausea and vomiting is common after eye surgery in children and might delay discharge if suitable prophylaxis is not given.

REFERENCES

- Anaesthesia for paediatric eye surgery. *Anaesthesia Tutorial of the Week* **144** (2009). Available from: <http://update.anaesthesiologists.org/wfsa-education-resources/tutorial-of-the-week/atotw-archive/Archive%20TotW/AToTW%20Articles%202009/Paediatrics/144%20Anaesthesia%20for%20paediatric%20eye%20surgery/detail>
1. James I. Anaesthesia for paediatric eye surgery. *Continuing Education in Anaesthesia, Critical Care and Pain* 2008; **8**: 5 – 10.
 2. Morrison A. Ophthalmology, plastics, oncology, radiology, thoracic, and dental surgery. In: Doyle E, eds. *Paediatric Anaesthesia*. Oxford, Oxford University Press. 2007: 298 – 307.
 3. Steward DJ, Lerman J. *Manual of Pediatric Anesthesia*. Churchill Livingstone, 2001: 225 – 33.
 4. BNF for children. *Notes on drugs and preparations: Eye*. BMJ group, RPS Publishing. 2008: 606 – 27.

5. Reimer EJ, Montgomery CJ, JC Bevan, et al. Propofol anaesthesia reduces early postoperative emesis after paediatric strabismus surgery. *Canadian Journal of Anaesthesia* 1993; **40**: 927 – 33.
6. Marsh DF, Hodgkinson B. Remifentanyl in paediatric anaesthetic practice. *Anaesthesia* 2009; **64**: 301 – 8.
7. Sator S, Wilding E, Schabering C, et al. Desflurane maintains intraocular pressure at an equivalent level to isoflurane and propofol during unstressed non-ophthalmic surgery. *British Journal of Anaesthesia* 1998; **80**: 243 – 45.
8. Sator – Katzenschlager S, Deusch E, Dolezal S, et al. Sevoflurane and propofol decrease intraocular pressure equally during non-ophthalmic surgery and recovery. *British Journal of Anaesthesia* 2002; **89**: 764 – 67.
9. Sator-Katzenschlager SM, Oehmke MJ, Deusch E, et al. Effects of remifentanyl and fentanyl on intraocular pressure during the maintenance and recovery of anaesthesia in patients undergoing non-ophthalmic surgery. *European Journal of Anaesthesiology* 2004; **21**: 95 – 101.
10. Chiu CL, Jaais F, Wang CY, Effect of Rocuronium compared with succinylcholine on intraocular pressure during rapid sequence induction of anaesthesia. *British Journal of anaesthesia* 1999; **82**: 757 – 61.
11. Craven R. Ketamine. *Anaesthesia*. 2007; **62**: 48 – 53.
12. Pun MS, Thakur J, Poudyal G, et al. Ketamine anaesthesia for paediatric ophthalmology surgery. *British Journal of Ophthalmology* 2002; **87**: 535 – 38.
13. Lerman J, Kiskis AA. Lidocaine attenuates the intraocular pressure response to rapid intubation in children. *Canadian Anaesthetists' Society Journal* 1985; **32**: 339 – 45.
14. Drenger B, Pe'er J. Attenuation of the ocular and systemic responses to tracheal intubation by intravenous lignocaine. *British Journal of Ophthalmology* 1987; **71**: 546 – 48.
15. Gulati M, Mohta M, Ahuja S, et al. Comparison of Laryngeal Mask Airway with tracheal Tube for Ophthalmic Surgery in Paediatric patients. *Anaesthesia and Intensive Care* 2004; **32**: 383 – 90.
16. Ruta U, Mollhoff T, Markodimitrakis H, Brodner G. Attenuation of the oculocardiac reflex after topically applied lignocaine during surgery for strabismus in children. *European Journal of Anaesthesiology* 1996; **13**: 11 – 15.
17. Gupta N, Kumar R, Kumar S, et al. A prospective randomised double blind study to evaluate the effect of peribulbar block or topical application of local anaesthesia combined with general anaesthesia on intra-operative and postoperative complications during paediatric strabismus surgery. *Anaesthesia* 2007 **62**: 1110 – 13.
18. Yi C, Jee D. Influence of the anaesthetic depth on the inhibition of the oculocardiac reflex during sevoflurane anaesthesia for paediatric strabismus surgery. *British Journal of Anaesthesia* 2008; **101**: 234 – 238.
19. Allen LE, Sudesh S, Sandramouli S, Cooper G, et al. The association between the oculocardiac reflex and postoperative vomiting in children undergoing strabismus surgery. *Eye*. 1998; **12**: 193 – 96.
20. Rose JB, Martin TM, Corrdry DH, et al. Ondansetron reduces the incidence and severity of post strabismus repair vomiting in children. *Anaesthesia and Analgesia* 1994; **79**: 486 – 489.
21. Sennaraj B, Shende D, Sadhasivam S, et al. Management of post-strabismus nausea and vomiting in children using ondansetron: a value-based comparison of outcomes. *British Journal of Anaesthesia* 2002; **89**: 473 – 78.
22. Tramer M, Moore A, McQuay H. Prevention of vomiting after paediatric strabismus surgery: a systematic review using numbers-needed-to-treat method. *British Journal of Anaesthesia* 1995; **75**: 556 – 61.
23. Sinha A, Baumann B. Anesthesia for ocular trauma. *Current Anaesthesia and Critical Care* 2010; **21**: 184 - 88
24. Murphy DF. Anaesthesia and Intraocular pressure. *Anaesthesia and Analgesia* 1985; **64**: 520 – 30.
25. Vachon CA, Warner DO, Bacon DR. Succinylcholine and the open globe: tracing the teaching. *Anesthesiology* 2003; **99**: 220 – 23.

Appendix 1. Conditions associated with eye abnormalities requiring special precautions.

Underlying condition	Eye condition	Special precautions
Neonates and premature babies	Congenital cataracts	As for neonatal surgery – warming, glucose management, monitoring for postoperative apnoea
Craniosynostosis syndromes (Crouzon, Apert and Pfeiffer syndromes)	Glaucoma, cataracts, squint, exophthalmos	Difficult to maintain airway with facemask (mid-face hypoplasia); improved by oral airway; intubation usually easy
Craniofacial syndromes (Goldenhar, Treacher Collins, Smith-Lemli-Opitz)	Glaucoma, cataracts, squint	Micrognathia/facial asymmetry – difficult intubation
Mucopolysaccharidoses (Hunter and Hurler syndromes)	Corneal opacities, retinitis pigmentosa	Difficult airway and intubation, cardiomyopathy, cervical spine instability
Down syndrome, Edward syndrome, Cri-du-Chat syndrome	Cataracts, strabismus	Difficult intubation, cervical spine instability in Down syndrome
Hallerman-Striess syndrome	Congenital cataract	Difficult intubation
Stickler syndrome	Glaucoma, chorioretinoid degeneration, lens dislocation	Cleft palate and associated airway problems
Homocystinuria	Lens dislocation	Hypoglycaemia
Marfan syndrome	Lens dislocation	Aortic root dilatation, aortic/mitral valve regurgitation
Neuro-oculo-cutaneous disorders (Neurofibromatosis Sturge-Weber syndrome, tuberous sclerosis, Von-Hippel-Lindau syndrome)	Retinal vascular disorders	Seizures, intracranial lesions, cardiac lesions and phaeochromocytoma

Anaesthesia for cleft and lip palate surgery

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Summary

Cleft lip and palate (CLP) is one of the commonest congenital deformities. Surgery aims to restore form and function, and modern techniques can leave many defects undetectable. Airway management problems, dealing with associated abnormalities and young patients all present anaesthetic challenges.

INTRODUCTION

Cleft lip and palate (CLP) is one of the commonest congenital deformities. The associated facial disfigurement causes feeding, speech and dental development problems and has significant psychosocial consequences. Surgery aims to restore form and function, and modern techniques can leave many defects undetectable. Airway management problems, dealing with associated abnormalities and young patients all present anaesthetic challenges.

CLASSIFICATION AND ANATOMY

Figure 1. *Incomplete unilateral cleft lip*

Cleft lip (CL) is a unilateral or bilateral fissure in the upper lip. Complete CL extends across the whole lip and into the nostrils. Incomplete CL ranges from small indentations to large defects with little connecting tissue between the two clefts.

Cleft palate (CP) is a unilateral or bilateral fissure in the soft palate that may extend into the hard palate. CP may occur with CL when the lip fissure extends beyond the incisive foramen and includes the sutura palatina. CP without CL is an aetiological and embryologically distinct entity.

The hard palate is formed by the palatine processes of the maxillae and the horizontal plates of the palatine bones. It is continuous with the soft palate, a movable fibromuscular fold from which the uvula hangs. The incisive foramen lies immediately behind the central maxillary incisors - the primary palate is anterior to the incisive foramen and secondary palate posterior.

Complete CP involves both primary and secondary palates whereas incomplete CP affects the secondary palate alone. A mucosal covering may obscure palatal defects often delaying diagnosis until the child develops subsequent speech problems.

INCIDENCE

The overall worldwide incidence is 1 in 7-800 live births and in the UK affects approximately 1000 babies

each year. Two-thirds involve the lip with or without the palate and the remainder the palate alone. CL is unilateral in 80% of cases and occurs on the left in over 70% of cases. Approximately 85% of infants with a bilateral CL and 70% with a unilateral CL will have an associated cleft palate.

The incidence of CL with or without CP is strongly influenced by race. At 3.6 per 1000 live births it is most common in Native Americans compared with 1.0 per 1000 Caucasians and 0.3 per 1000 Afro-Caribbean births. CP alone occurs more evenly across races at around 0.4 per 1000 live births. CLP is more common in males and the more severe the cleft the wider the sex discrepancy becomes. In contrast isolated palatal clefts are more common in females.

EMBRYOLOGY

Lip and palate development occurs in the first trimester, the critical period being between 6 and 9 weeks gestation. The upper lip and primary palate are formed from the fusion of the frontonasal and bilateral maxillary prominences - CL occurs when this fusion fails on either or both sides.

The secondary palate is formed from lateral palatal processes arising from the deep portions of the maxillary prominences. Initially these lie vertically alongside the tongue, but as mandibular development proceeds the tongue moves inferiorly allowing the palatal shelves to assume a horizontal alignment. Fusion of the two shelves occurs in an anterior to posterior direction - incomplete fusion produces CP.

CL may be reliably diagnosed at the 18-20 week scan. CP is harder to see and can only be excluded on examination after delivery.

AETIOLOGY

The cause of CLP is unknown but appears to be multifactorial with genetic and environmental influences. It is familial; affected parents have a 3-5% chance of an affected child, and with one affected child the sibling risk is 20-40%. Monozygotic twins show the same defect in 40-50% of cases, but only 5% in dizygotic twins.

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Some cases may result from mechanical obstruction. Impaired mandible development can prevent the tongue descending, which then obstructs fusion of the palatal shelves. Teratogen exposure associated with CLP includes maternal alcohol use and smoking, anticonvulsants (phenytoin, benzodiazepines), salicylates and cortisone. The risk increases with rising maternal and paternal age. Folic acid 400mcg.day⁻¹ has a role in preventing CLP.

Associated Conditions

CLP is associated with over 200 syndromes or sequences and several have significant anaesthetic implications (Table 1). Children with CLP may have multiple abnormalities without a recognised syndrome. Additional abnormalities are most likely to be found with isolated CP (particularly submucous CP) and least likely with isolated CL. Craniofacial abnormalities are most common, followed by CNS abnormalities e.g. mental retardation and seizures, congenital cardiac disease, renal and abdominal defects.

Estimates of CLP patients with associated abnormalities range from 10-60%. Studies of birth records produce lower values compared with those subjecting patients to detailed clinical and genetic examination.

SURGICAL REPAIR

CL is classically repaired between 6 and 12 weeks but there is an increasing trend to operate in neonatal period. More popular with parents, this has a positive influence on the associated sleep disordered breathing problems, and also produces better aesthetic results and may promote bonding. CP is usually repaired later, between 3 and 9 months, in a one or two stage operation to promote normal speech development and reduce nasal regurgitation. Surgery may be delayed due to associated abnormalities, or more commonly lack of access to appropriate services CLP patients are likely to require further surgery either related to the primary problem e.g. plastic improvements to a CL repair, or for associated abnormalities. Around 20% will require pharyngoplasty for velopharyngeal dysfunction at around 4-6 years.

Primary palatoplasty disrupts normal palate growth and despite orthodontic treatment, some will require significant maxillofacial



Figure 1. Large adult cleft palate

Table 1: Syndromes and sequences associated with cleft lip and palate

Syndrome or sequence	Clinical details
Velocardiofacial syndrome, also known as Sprinzen's syndrome; includes DiGeorge syndrome (22q11 deletion)	Most common syndrome associated with CLP Cleft palate in ~30%, velopharyngeal incompetence Congenital cardiac disease, immune deficiency
Pierre Robin syndrome	Cleft palate in 80% of cases Micrognathia Glossoptosis Intubation becomes easier with age due to mandible growth
Stickler syndrome	Progressive connective tissue disorder (autosomal dominant) Midface hypoplasia, micrognathia, Pierre Robin sequence Retinal detachment and early cataracts, deafness Hypermobility of joints
Treacher Collins	Cleft palate in ~30% of cases Hypoplasia of zygomatic bones and mandible Eye and ear abnormalities, deafness Choanal stenosis or atresia Significant risk of airway obstruction in neonatal period Intubation may become more difficult with increasing age
Down syndrome	Macroglossia, microstomia Atlantoaxial subluxation and instability Small stature, mental retardation Congenital cardiac disease
Goldenhar syndrome (Hemifacial microsomia)	Incomplete development of palate, lip, nose, ear and mandible on one side of the face Scoliosis, renal and lung abnormalities Intubation may become more difficult with increasing age
Foetal alcohol syndrome	Small palpebral fissures, smooth philtrum, thin upper lip Growth deficiency, CNS abnormalities, microcephaly

surgery in their teens to correct midface hypoplasia and maxillary retrusion

ANAESTHESIA FOR CLP

Anaesthesia for primary CLP surgery may be successfully provided in a wide range of environments from the well-equipped dedicated paediatric hospital, to isolated resource poor clinics. In the UK CLP surgery is limited to 10 specialist centres; anaesthetising CLP patients without oxygen would be an anathema here, yet it is a reality borne of necessity for many. There are a variety of acceptable anaesthetic techniques and practice should be modified by experience and available facilities.

Pre-operative assessment

History and examination should assess general fitness for anaesthesia and surgery. Particular attention should be paid to associated abnormalities. Some issues specific to CLP patients are explored below.

Airway management

Over 70 years ago Magill recognised problems with airway management in children with CLP, but predicting which children may have difficult airways is problematic. CLP does not make upper airway obstruction inevitable and when obstruction occurs it is more often due to associated structural or neuromuscular problems

Some syndromes are well associated with difficult intubation (see Table 1). In non-syndromic patients difficult laryngoscopy and intubation were strongly associated with retrognathia and bilateral cleft lip (due to the protruding maxilla). Fewer problems occur with increasing age and are very uncommon over the age of 5.

Patients with previous CLP repairs more commonly have difficult airways. Nasal intubation is acceptable except with history of pharyngoplasty when it should be avoided. The nostril of choice is the side of the original cleft as this will be wider. Laryngeal mask airways (LMAs) have been widely used in children with previous repairs without reported adverse effects although rotation on insertion is not advised.

Upper respiratory tract infection (URTI)

Chronic rhinorrhoea is common in children with CLP due to food reflux into the nasal passages and they may present with overt, often recurrent, URTI. Even if clinically well, pre-operative antibiotics for children with low grade infection (positive nasal swabs) reduces the incidence of post-operative respiratory complications (PRC). Surgical repair reduces rhinorrhoea and URTI so risks of anaesthesia and PRC should be individually balanced against the benefits of surgery.

The risk of PRC increases with the severity of the defect. Infants with bilateral CLP have a significantly higher risk of PRC (9%) than infants with isolated CL or unilateral CLP (2 and 3% respectively), even when clinical scoring pre-operatively gives no indication of infection.

Chronic airway obstruction

Snoring, apnoea during feeds or protracted feeding time may indicate chronic airway obstruction. Older children and adults may have chronic hypoxia, right ventricular hypertrophy and cor pulmonale. These patients are more sensitive to sedative drugs and have an

increased risk of airway obstruction at induction and post-operatively. If cardiac involvement is suspected an ECG and ECHO should be considered and a higher-level post-operative monitoring is advised.

Nutrition and hydration

CLP defects make it difficult for an infant to create a seal sufficient to suckle. Feeding difficulties are common and surgery should be deferred in malnourished or dehydrated children. Nutritional or physiological anaemia may occur (a nadir at around 9 weeks); measuring haemoglobin may be appropriate but CLP repairs are successful in resource-poor environments without laboratory facilities.

Premedication

Sedatives may precipitate airway obstruction and should be avoided. Atropine (20mcg.kg⁻¹ intramuscularly 30 minutes pre-operatively or 10-20mcg.kg⁻¹ intravenously at induction) is an effective drying agent and is advisable when difficult intubation is anticipated or anaesthesia planned with ether or ketamine.

Intraoperative management

Cleft lip repair in adults and older children can be performed with local anaesthetic infiltration and conscious sedation e.g. diazepam 0.05-0.1mg.kg⁻¹ All other patients will require general anaesthesia.

Induction

In general, choose a technique that maintains spontaneous ventilation. Gas induction with a volatile agent (e.g. sevoflurane, halothane) in oxygen is common; ketamine given intramuscularly (10-12.5mg.kg⁻¹) or intravenously (1-2mg.kg⁻¹) is an alternative. Intravenous access, if not already established, should be obtained as soon as the child is asleep and facemask ventilation confirmed before the use of any neuromuscular blocking drugs. A standard intravenous induction may be appropriate for older children or adults without anticipated airway difficulty e.g. propofol 4-6mg.kg⁻¹, thiopentone 3-5mg.kg⁻¹.

Difficult mask ventilation is unusual but should it occur options include nasal or oropharyngeal airways, a laryngeal mask airway or turning the patient lateral. These manoeuvres may achieve sufficient anaesthetic depth to allow intubation. However, CLP repair is not life-saving surgery – if the airway cannot be managed safely surgery should be deferred until they are older when maturation often makes airway management easier.

Endotracheal intubation may be performed under deep inhalational anaesthesia or using muscle relaxants e.g. suxamethonium 2mg.kg⁻¹ or a non-depolarising agent. Difficult laryngoscopy (Cormack and Lehane views grade III or IV) occurs in up to 10% of ASA I patients for CLP repair and the incidence rises in those with an associated syndrome. Large alveolar defects may hamper laryngoscopy, as there is a tendency for the laryngoscope to fall into the cleft; packing with gauze may help prevent this, as may the use of a straight blade.

A variety of techniques are available for difficult intubations; anterior laryngeal pressure, alternative laryngoscopes and the gum elastic bougie are simple, readily available and effective. The LMA has been successfully used to allow CLP repair in a child in whom intubation had proved impossible. It is more bulky and less secure than an endotracheal tube and its routine use is not advised.

Fibreoptic techniques often use an LMA as a conduit. A guide-wire may be threaded down the suction port of an adult endoscope, the LMA and endoscope are then removed and the wire used to railroad a tube.

Alternatively a paediatric endoscope may be used to introduce a pre-loaded tube directly through the LMA.

Rarely an emergency surgical airway may be required. A preformed oral south facing RAE tube is ideal for this surgery as it can be taped on the chin and improves surgical access although standard and reinforced tubes are both acceptable.

Intubation is not always necessary. In children older than one year, routine CL repair has been described using only ketamine, atropine and local anaesthetic infiltration. This requires considerable experience and co-operation between anaesthetist and surgeon but may be useful when resources are limited.

Maintenance

A head ring and roll under the patient's shoulders extend the neck and tip the head down, and throat packs are used to absorb blood and secretions. During palate surgery a gag inserted over the endotracheal tube keeps the mouth open and tongue clear. Tube problems are common during surgery with a shared airway and may occur at any time. Vigilance is needed to prevent inadvertent extubation, intubation of the right main bronchus and tube kinking or occlusion.

Maintenance is most often with an inhalational agent choice. Halothane should only be used if oxygen is available due to the risk of arrhythmias. Ether precludes the use of diathermy due to the explosion risk. There is growing interest in desflurane as it produces rapid recovery with early return of airway reflexes. However it is expensive, requires special vaporisers and is not suitable for gas inductions.

Intravenous bolus doses of ketamine may be given for maintenance ($0.25\text{mg}\cdot\text{kg}^{-1}$). It produces dissociative anaesthesia and has the advantage of maintaining respiration and cough reflex. However, experience is required to titrate the dose of ketamine correctly, particularly in infants or small children, and there are disadvantages of hypersalivation and emergence phenomena.

Spontaneous ventilation techniques are safer if there is a disconnection or inadvertent extubation but are not suitable for infants and small children.

Controlled ventilation with muscle relaxation reduces anaesthetic requirements promoting a more rapid wake up and recovery of reflexes, as well as allowing lower PaCO_2 , which may reduce blood loss.

If available, modern anaesthetic machines with integral ventilators allow the anaesthetist to select his or her preferred technique. Drawover systems are more common worldwide and are sufficient for CLP surgery. A standard system may include a vaporiser such as the Epstein Macintosh Oxford (EMO) for ether or Oxford Miniature Vaporiser (OMV) for halothane connected in series to an Oxford Inflating Bellows (OIB; Penlon, Abingdon, UK, See Drawover Anaesthesia article, page XXXXX). Drawover systems are not suitable for children who weigh less than 20kg due to high respiratory resistance. Attaching a Jackson Rees circuit to an OIB is an example of a suitable modification;

the positive-pressure ventilation required for neonates and infants is possible with the bag of the T-piece whilst the OIB can be used to generate a fresh gas flow.

Surgery usually lasts 1-2 hours. Although blood transfusion is uncommon, CP repairs have the potential for significant blood loss so facilities for cross matching should be available. Existing fluid deficits and intraoperative losses are replaced with crystalloid and a single dose of intravenous antibiotic e.g. augmentin given.

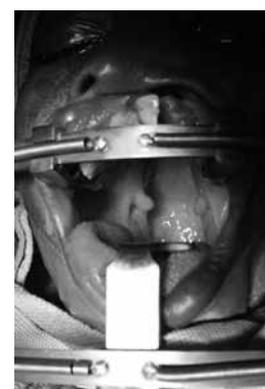


Figure 3. Cleft palate repair

Infiltration of local anaesthetic by the surgeon is recommended e.g. 1% lidocaine with 1:200,000 adrenaline. This provides intraoperative analgesia, reduces blood loss and improves the surgical field. The dose of adrenaline should be limited to $5\text{ micrograms}\cdot\text{kg}^{-1}$ if halothane is being used.

Paracetamol (acetaminophen) can be given orally as a pre-medication ($20\text{mg}\cdot\text{kg}^{-1}$) or rectally after induction ($30\text{--}40\text{mg}\cdot\text{kg}^{-1}$). Non-steroidal anti-inflammatory drugs (NSAIDs) are effective analgesics and most paediatric anaesthetists prescribe them to infants over 6 months old. They may increase the risk of post-operative bleeding thus some advocate delaying administration until 12 hours postoperatively.

Despite local anaesthetic infiltration, endotracheal tube movement can produce marked intraoperative stimulation, which can be obtunded with intraoperative opioids. For CL repairs short acting agents e.g. fentanyl $1\text{--}2\text{ micrograms}\cdot\text{kg}^{-1}$ are sufficient whereas for more painful CP repairs a longer acting agent is more appropriate e.g. morphine $0.05\text{--}0.1\text{mg}\cdot\text{kg}^{-1}$. Opioids have the advantage of promoting a smoother emergence with less crying, which may reduce swelling and bleeding from the surgical site.

The use of opioids in neonates and infants raises justifiable concerns regarding post-operative sedation, respiratory depression and consequent airway compromise. Where postoperative supervision is inadequate it is vital that the child leaves theatre fully awake and in control of their airway. If trained staff, pulse oximetry and apnoea monitors are not available then opioids should be avoided and alternative analgesia provided.

Infraorbital nerve blocks can provide effective post-operative analgesia for cleft lip repair. The infraorbital foramen may be palpated in children and adults; in neonates it may be found at the mid-point of a line drawn from the midpoint of the palpebral fissure to the angle of the mouth (approximately 7.5mm from the alar base). The nerve may be approached percutaneously or via the mucobuccal fold; only small volumes of local anaesthetic are required e.g. $0.5\text{--}2\text{ml}$ 0.5% bupivacaine depending upon weight (maximum bupivacaine dose $2\text{mg}\cdot\text{kg}^{-1}$ which is $0.4\text{ml}\cdot\text{kg}^{-1}$ 0.5% solution).

Extubation and postoperative care

The very real risk of postoperative airway obstruction is most likely to occur in children with pre-operative airway problems. Throat packs should be removed at the end of the operation and the oropharynx

inspected for blood clots and to check haemostasis. Thereafter keep suction to a minimum. If nondepolarising relaxants have been used they should be antagonised. Extubate the child once fully awake and protective reflexes have returned.

Airway obstruction may be due to swelling of the tongue from gag pressure, inadequate mouth breathing, laryngospasm, retained throat pack, blood clot or a combination of factors. A short period of CPAP may suffice as may turning the child lateral or prone. Nasopharyngeal airways (NPA) are effective and well tolerated; in patients at high risk of postoperative airway complications - in particular those who have a pharyngoplasty (Furlow procedure) as part of their surgery - they may be inserted before emergence. Insertion at this time will be less traumatic and greatly reduces the risk of disrupting the surgical suture line. The NPA can usually be removed the following day once swelling has subsided and the child has mastered mouth breathing. Avoid oropharyngeal airways due to the risk of disrupting the surgical repair. A small number of infants will require re-intubation and possibly tracheostomy.

Careful monitoring for 12-24 hours allows early detection of any airway obstruction or postoperative bleeding. Ideally this will be in a high dependency unit, although centres may be able to provide the required care outside such an environment. Patients at particular risk e.g. those with Pierre-Robin sequence should be cared for in an ICU. In addition to the risks of mechanical upper airway obstruction, the control of breathing alters around the time of surgery due to the different shape of the upper airway and change in breathing pattern

associated with this. If pulse oximetry is available it is highly advised while the child is asleep, to continue until you observe them to sleep without significant oxygen desaturation occurring.

Children can be reluctant to feed and intravenous fluids should be continued until adequate oral intake is established.

Post-operative analgesia includes opioids administered as intravenous boluses, continuous infusions or nurse controlled analgesia according to locally developed protocols, as well as regular paracetamol and NSAIDS (for those over 6 months).

REFERENCES

Rawlinson E. Anaesthesia for cleft lip and palate surgery. *ATOTW* (2007) **72**. Available from: http://www.wfsahq.org/components/com_virtual_library/media/bd407ebfb3607f1cb53510c04bf0bc1bd8b0d6dbfd48ef476d91c9415cc17a7d-72-Anaesthesia-for-cleft-lip-and-palate-surgery.pdf

1. Tolarova M, Oh H. Cleft Lip and Palate. May 2006 eMedicine <http://www.emedicine.com/ped/topic2679.htm>
2. Law RC, deKlerk C. Anaesthesia for Cleft Lip and Palate Surgery. Update in Anaesthesia Issue 14 (2002) Article 9. http://www.nda.ox.ac.uk/wfsa/html/u14/u1409_01.htm
3. Simpson S, Wilson I. Drawover Anaesthesia Review. Update in Anaesthesia Issue 15 (2002). Article 6. http://www.nda.ox.ac.uk/wfsa/html/u15/u1506_01.htm
4. Somerville N, Fenlon S. Anaesthesia for Cleft Lip and Palate Surgery. *CEACCP*, Jun 2005; 5: 76-79.

Anaesthesia for paediatric orthopaedic surgery

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INTRODUCTION

Paediatric orthopaedics in low or middle income countries (LMIC) ranges from simple fractures (but often complicated by delayed presentation, anaemia or nutritional deficiency), to chronic osteomyelitis and fracture non-union, to complex elective procedures in children with cerebral palsy. This article will consider the spectrum of disorders encountered in paediatric orthopaedic surgery in LMICs, the orthopaedic manifestations of specific conditions in childhood, specific orthopaedic procedures and anaesthetic management of these conditions. Practical aspects of regional anaesthesia are covered elsewhere in this issue of Update (page 99).

THE SPECTRUM OF DISORDERS SEEN IN PAEDIATRIC ORTHOPAEDIC SURGERY

Conditions can be considered under the following broad headings:

- Trauma: Simple and complex fractures (acute or delayed), burns, polytrauma, traumatic paraplegia, conflict related
- Common congenital conditions: Talipes equinovarus (club foot), scoliosis and other congenital limb deformities, achondroplasia, bone cysts
- Infections: Osteomyelitis (acute, untreated, chronic), TB, poliomyelitis
- Developmental abnormalities: Developmental dysplasias of the hip (congenital dislocation of the hip (CDH)), Perthes disease, slipped upper femoral epiphysis (SUFE), idiopathic scoliosis
- Neuromuscular conditions: Muscular dystrophies, progressive muscular atrophy, poliomyelitis, scoliosis

- Neurological conditions: Cerebral palsy, spina bifida
- Auto-immune conditions: Juvenile idiopathic arthritis (JIA)
- Tumours: Sarcomas, osteochondromas
- Rare congenital conditions: Osteogenesis imperfecta, neurofibromatosis, mucopolysaccharidosis (Hunter's, Hurler's), arthrogyrosis multiplex.

COMMON ORTHOPAEDIC CONDITIONS IN LMIC

Trauma

Acute fractures and burns are common in children in LMIC and can be associated with high morbidity. A high proportion of fractures are treated non-surgically with traction or simple casting, with fracture manipulation under anaesthesia one of the most common paediatric orthopaedic procedures undertaken. Paediatric musculoskeletal impairment (MSI) has a prevalence of 2.6-4.8% in children under 12 years; angular limb deformity and fracture non/mal-union are seen in a significant proportion of children presenting for elective surgery.^{1,2} Polytrauma and burns (acute and reconstructive procedures) are challenging problems associated with high mortality, which are considered elsewhere in this edition of *Update* [page 199 and 204].

Congenital talipes equinovarus (clubfoot)

Congenital talipes equinovarus (clubfoot) seems to have a higher prevalence in developing countries compared to elsewhere - for example, the incidence of clubfoot in Malawi is 2 per 1000 children, twice that of North America and Europe.^{3,4} Although clubfoot programmes

SUMMARY

Paediatric orthopaedic procedures range from fixing simple fractures in otherwise healthy children to more complex procedures in patients with major co-morbidities. Anaesthetic management is generally dependent on the complexity of the procedure as well as the child's general health. Pain relief is a particular challenge. Regional anaesthesia plays a very important role in managing intra- and postoperative pain. Availability of resources, training and safety are a major determinant of anaesthesia technique in LMICs.

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with Ponseti serial casting are widespread in LMIC, disability associated with untreated or partially treated clubfoot remains high.^{5,6}

CASE SELECTION

The success of orthopaedic programmes in LMICs depends on case selection and choice of surgical procedure. In general, interventions that have a good outcome are those that require little or no follow up, are cheap to perform with minimal instrumentation and implanted material, and do not require specialised surgical skills. The majority of children with cerebral palsy are best treated with prolonged physiotherapy; operative intervention and prolonged multidisciplinary follow-up are time consuming and expensive.

Five common orthopaedic conditions that benefit from operative intervention and where operative treatment is financially and practically feasible are:



Figure 1. *Recurrent club foot*

- Neglected or recurrent talipes (club foot): postero-medial release, posterior release, wedge tarsectomy or triple arthrodesis (see Figure 1)
- Chronic osteomyelitis: sequestrectomy and debridement. Some cases occur secondary to overlying soft tissue injury but many cases are caused by blood borne infection. If the osteomyelitis is adjacent to a joint there should be a high index of suspicion for intra-articular spread with septic arthritis. Systemic infections prevalent in the developing world include tuberculosis and Human Immunodeficiency Virus (HIV). Thanks to a successful global polio vaccination programme, the prevalence of this disease is now very low but it has yet to be eradicated.
- Angular limb deformities: Malnutrition with vitamin D deficiency and rickets, and developmental disorders such

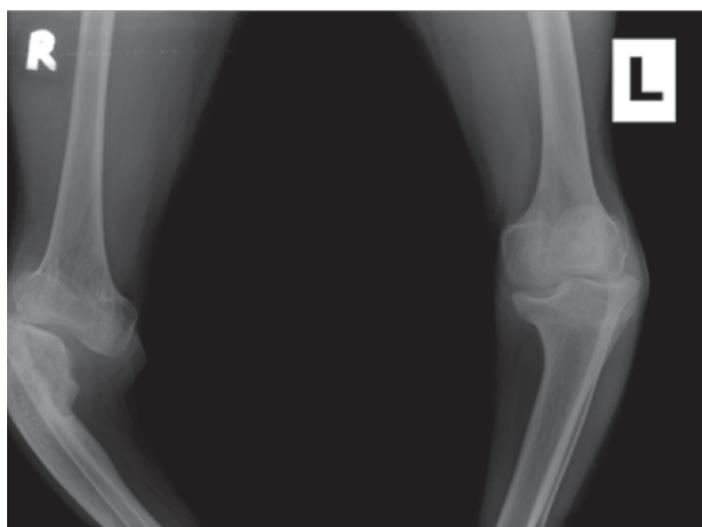


Figure 2. *Blounts Disease (Bow legs) showing angular deformities around the knee, more severe on the right leg*

as Blounts disease cause angular limb deformities, especially around the knee (see Figure 2). An open osteotomy is required to re-align the limb and the reduction is usually maintained by casting rather than metalwork.

- Burns contractures. Releases and skin grafting are common procedures undertaken by orthopaedic surgeons, particularly when involving the hands
- Open fracture requiring wash out and debridement

ANAESTHETIC MANAGEMENT OF CHILDREN PRESENTING FOR ORTHOPAEDIC SURGERY

Pre-operative assessment and preparation

All children should be seen pre-operatively, preferably with a parent present, and a full history and appropriate examination taken as routine (see p XX this *Update*). Pay particular attention to co-existing conditions, particularly for children with cerebral palsy or cardiorespiratory disease. Previous experience of anaesthesia can be a useful pointer as many of these children may have had other procedures, with positive or negative experiences. Review of previous anaesthetic charts is useful when available. A particular challenge is the small malnourished child presenting for the first time with a long history of an untreated fracture or osteomyelitis to a facility with minimal facilities for pre-operative investigation, but much can be gained from the patient history and examination.

Pain management is often a special concern for children and parents, and pain management plans should be discussed before surgery. For fractures, this is usually best provided by a cast, sling or traction, supplemented with oral analgesia. IV morphine may be required if pain is severe, or a regional block may be of benefit (e.g. femoral nerve block). Consent should be obtained if rectal analgesics are to be used.

Starvation: This should be as standard, but children with acute fractures or complex co-morbidities may well have delayed gastric emptying. Extra caution is required.

Pre-medication: This is not usually required, but those with learning difficulties or previous difficult experiences may benefit from sedative pre-medication. For a child undergoing multiple procedures, a calm, smooth induction of anaesthesia can help enormously with subsequent anaesthesia.

Simple oral analgesics such as paracetamol 20mg.kg⁻¹ PO or ibuprofen 5-10mg.kg⁻¹ PO can be given as a premed. This is easy to do, well-tolerated, inexpensive, may reduce opioid requirements, and improves postoperative pain management.

Examination: This should be a focused physical examination relevant to the history and practical aspects of anaesthesia, for instance, intravenous access, airway management and positioning.

Investigations: Children who are fit, healthy and having minor procedures do not require laboratory investigations. Those undergoing major surgery will need a haematocrit or full blood count with group and screen as a minimum. Investigation of cardio-respiratory disease is best done in consultation with a paediatrician. Screening with pulse oximetry, particularly an overnight assessment of SpO₂ may be useful if there is a concern about cardiorespiratory disease.

The anaesthetic plan should be explained to the parents and the child if possible, and consent obtained.

Anaesthetic technique

The surgery and available resources will determine the choice of technique. Some procedures such as tendon release in the Ponseti procedure are routinely performed under local anaesthesia. Many cases are safely anaesthetised with either IM or IV ketamine; regional anaesthesia may be used for older children. For peripheral surgery, a standard GA technique is appropriate for most cases, with spontaneous breathing with inhalational anaesthesia, ideally supplemented by regional block or local infiltration. Consider tracheal intubation if there is delayed gastric emptying, expected poor respiratory effort under anaesthesia (e.g. neurologically impaired), or a very long procedure.

Intravenous access is best practice for all cases. Short simple cases such as application of hip spicas and other plaster applications may be safely anaesthetised without IV access; infants, however, should have secure IV access even for these cases. Though simple and often quick, fracture manipulation can be very stimulating and an IV is useful for quickly deepening anaesthesia. Ketamine anaesthesia or regional blocks awake are safest with IV access in place.

Analgesia is best provided with a multi-modal technique (ketamine, oral analgesics, a regional block or local anaesthetic infiltration, +/- opioids if required). Postoperatively, use regular simple analgesics supplemented by stronger opioid drugs if required (given by agreed protocol), guided by age-appropriate pain assessment tools (e.g. FACES, FLACC) (see pain management page 72 this *Update*)

It is good practice to give intravenous fluids to rehydrate following a period of fasting and to replace intra-operative losses. Postoperative nausea and vomiting can be reduced with good hydration, and this is less expensive and more readily available than anti-emetic drugs.

Open procedures will require prophylactic antibiotics, which should be given before 'knife to skin'.

Regional anaesthesia in paediatric orthopaedic surgery

Regional anaesthesia can provide excellent perioperative analgesia and anaesthesia, but requires special training and equipment; regional anaesthesia is discussed elsewhere in this *Update*. (See article page 99).

Specific considerations for orthopaedic surgery are as below:

- *Choice of block:* Discuss with surgeons and ward nurses

when deciding which block will be the most useful (location and longevity required). The child and parents' previous experience of regional anaesthesia may guide decisions. Catheters can also be used in regional techniques to prolong analgesia.

- **Lower extremity surgery:** The simplest block to perform is caudal block, as this covers many lower limb procedures, and is safe and efficacious in all paediatric age groups.^{7,8,9} Unilateral limb blocks provide good quality analgesia; the most common and easy to perform being femoral/3:1 or popliteal sciatic blocks. Any procedure below the knee can be covered with popliteal blocks, supplemented locally or by infiltration of the saphenous nerve for lateral surgery at the ankle or foot. Ankle blocks are straightforward and can provide good pain relief for foot procedures.
- **Surgery at the hip:** This requires a higher block usually an epidural, as a one shot or catheter technique. This provides excellent prolonged analgesia for most lower limb surgery. Lumbar epidurals can only be safely performed in children under general anaesthesia.
- **Spinal anaesthesia:** This can be used in older children as an alternative to a GA. It is safe and cost effective in experienced hands and if the appropriate equipment is available, particularly when working with a fast surgeon. Consent is clearly an important issue.
- **Upper extremity surgery:** Brachial plexus block at the axilla provides excellent analgesia for wrist and hand surgery in children, and may be supplemented by local infiltration or ketamine IV or IM if required to provide anaesthesia for surgery. In expert hands, supraclavicular block or interscalene block may be used to provide anaesthesia or analgesia.¹⁰

BLOOD LOSS AND BLOOD CONSERVATION

It is important to develop good blood conservation practices during orthopaedic surgery in LMIC as chronic anaemia is common in children, access to safe blood transfusion is limited and costly, and this type of surgery has the potential for large blood loss.

Blood conservation is greatly aided by the use of tourniquets where possible, but much can be achieved by simple changes in anaesthetic technique, without the use of induced hypotension or expensive drugs.

The following techniques help to reduce the requirement for blood transfusion:

- In elective surgery, optimise preoperative haemoglobin: consider de-worming and iron supplements in at-risk populations
- A stable anaesthetic with good analgesia, with control of heart rate and blood pressure in the low normal range. Use sedative premedication if the child is anxious. If available, TIVA with propofol and remifentanyl is useful as anaesthetic depth can be varied easily according to the level of surgical stimulation
- Maintain normothermia to prevent cooling and hence coagulation problems; limit patient exposure from induction of anaesthesia onwards
- Position the child carefully to reduce venous congestion, avoid hypercarbia and avoid the child coughing. This will help to reduce bleeding from the surgical site
- Tranexamic acid 10-20mg.kg⁻¹ bolus IV at induction, repeat dose 10mg.kg⁻¹ IV at 4 hours. This is a safe drug and has been shown to reduce blood loss in trauma patients and should be increasingly available

Table 1. Different types of cerebral palsy

Description:	Spastic CP	Ataxic CP	Athetoid/dyskinetic CP	Mixed type
Incidence (% cases)	70%	10%	10%	10%
Clinical features:	Hemiplegic CP - one side of the body is affected. Diplegic CP - both legs are usually more affected than both arms Tetraplegic CP - all four limbs are involved	Intention tremor Head tremor Poor sense of balance with falls and stumbles	Dystonic - maintained twisting position of torso and extremities Athetosis - slow, purposeless, distal movements Chorea - quick, jerky, proximal	Spastic athetoid

- Use clearly defined transfusion protocols, in particular the trigger for transfusion. Healthy children can tolerate an acute drop in haemoglobin to 7g.dl⁻¹ with no adverse effects. A low haemoglobin threshold for transfusion is a cost free way of avoiding transfusion
- If available, intra- and post-operative cell salvage is very useful
- Temporomandibular joint dysfunction, increased incidence of malocclusion; possible difficult intubation
- Autonomic neuropathy; poor control of temperature and blood pressure. Intraoperative hypothermia is common, compounded by lack of fat and muscle in the malnourished child
- Difficult venous access due to spasticity or dystonia

If major blood loss occurs (i.e. more than half circulating blood volume), use whole blood to correct coagulopathy, or “reconstituted whole blood”, using FFP and platelets with each red cell transfusion.

SPECIFIC CONDITIONS REQUIRING ORTHOPAEDIC SURGERY

Cerebral palsy

Most children with cerebral palsy (CP) presenting for surgery in LMIC will have mild deficits affecting one or two limbs, but without the major complications associated with CP. These children will often be otherwise well, but with decreased mobility or limb deformity requiring correction. Rarely, children with severe comorbidity may also present (see Table 1).

Intellectual disability and epilepsy are common in children with spastic quadriplegic CP. Bulbar muscles are involved to a varying degree, resulting in poor control of the mouth, tongue and pharynx. Aspiration pneumonia is most likely in this group. Dyskinetic CP may be associated with deafness, dysarthria and drooling.¹¹

Anaesthetic implications of CP:^{11, 12}

- Cognitive, communication and behavioural disorders
- Epilepsy (30% of children)
- Gastro-oesophageal reflux is common; may cause recurrent respiratory problems
- Drooling of saliva may be related to pseudobulbar palsy with impaired swallowing or tongue thrusting
- Poor nutritional status, with potential for electrolyte imbalance or anaemia
- Respiratory problems; poor respiratory reserve, sub-clinical pulmonary aspiration from reflux, recurrent respiratory tract infections, and chronic lung disease
- Poor dentition; dental caries and loose teeth are common. The potential for bacteraemia during airway instrumentation has important implications for children receiving metal implants

Ideally, children with CP undergoing major surgery should be assessed by a paediatrician prior to surgery, to make sure that they are as fit as possible for the proposed surgery. Anticonvulsant therapy should be continued perioperatively and restarted as soon as possible postoperatively. These patients may also require physiotherapy, bronchodilators and antibiotics preoperatively. Several readings of the blood pressure should be taken to check the baseline. Keep the child warm and avoid exposure whenever possible.

Orthopaedic procedures in children with CP

Soft tissue releases to relieve contractures are commonly performed in these children. Repeat procedures are common. Recent trends are for single event multilevel surgery involving tenotomies and/or osteotomies at different levels on one or both limbs.

Conduct of anaesthesia

It is important to have a good rapport with these children and engage them in the discussions. Communication may be difficult and the child may be anxious, so it is useful to have their carer available at induction and in the recovery room. Sedative premedication can be considered but these patients can have an unpredictable response to them. Antacids, prokinetics and drugs to reduce secretions may be useful. Thiopentone is a useful IV induction agent in children with epilepsy. Intubation should be considered in children with reflux. The child should be positioned carefully to prevent pressure sores, nerve or muscle damage. Children with severe CP may have reduced volatile anaesthetic requirements and may take a long time to wake after surgery.

Muscle spasms are a particular problem in children with spastic CP, and for this reason, regional techniques are strongly recommended for intra-operative and postoperative analgesia, also to reduce opioid requirements. In children having extensive lower limb surgery, epidural analgesia is beneficial.¹¹ Pain assessment can be challenging, but should not prevent the anaesthetist seeking to provide good analgesia.

Postoperatively, drooling can present problems, and frequent suctioning may be necessary. Aspiration of gastric contents

Table 2. Classification of neuromuscular disorders

	Inherited	Acquired
Prejunctional	Peripheral neuropathies <ul style="list-style-type: none"> • Charcot-Marie Tooth • Friedreich's ataxia 	Motor neurone disease Multiple sclerosis Guillain-Barré syndrome Peripheral neuropathies
Junctional		Myasthenia Gravis Eaton-Lambert Syndrome
Post-junctional	Dystrophies <ul style="list-style-type: none"> • Duchenne (DMD) • Becker's Myotonias <ul style="list-style-type: none"> • Myotonic dystrophy • Myotonia congenital • Other myopathies • Hyper/hypokalaemic periodic paralysis Metabolic/mitochondrial disorders	Inflammatory myopathies Critical illness polyneuropathy and myopathy

may occur in children with pseudobulbar palsy. Children with CP are commonly irritable on emergence, but it may be difficult to elicit the cause.

Neuromuscular disorders

Pre-operative assessment

Children with significant neuromuscular conditions presenting for orthopaedic surgery usually have a clear diagnosis (see table 2). If there is limited information, it is important to ask how long the child has been weak, whether the weakness is stable or progressive, if the muscle weakness is associated with fatigability and what limits activity. All major cases should be discussed with a neurologist or paediatrician before surgery. All medication should be continued preoperatively, and restarted as soon as possible after surgery.

Anaesthetic assessment should include current status, airway, cardio-respiratory and any other system disorders. The principal anaesthetic risks relate to the airway, respiratory impairment, poor myocardial function, gastro-oesophageal reflux, abnormal drug reactions (principally MH), and excess bleeding in certain myopathies (e.g. Duchenne muscular dystrophy, DMD). Investigations are best undertaken in consultation with respiratory paediatricians and/or paediatric cardiologists if available. The family should be counselled to make sure that they are aware of the prognosis of the specific

condition, also that they are realistic as to their expectations about surgery.

Assessing functional capacity during exercise is very useful, as it will help identify to significant cardio- respiratory or airway compromise. However, if the child is inactive this may mask the severity of both respiratory and cardiac disease. Formal sleep studies are often used to indicate the need for non-invasive respiratory support postoperatively; overnight oxygen saturation monitoring can also reveal useful clinical information. Children already established on non-invasive ventilatory support can be safely anaesthetised; familiarity with the particular device and current settings is essential preoperatively (the parents are often expert). The device must be available for immediate use as the child wakes after surgery.

Progressive degeneration of cardiac muscle fibres, resulting in conduction defects and cardiomyopathy occurs in DMD; this occurs in later adolescence and is managed in the early phase with ACE inhibitors. Friedrich's ataxia is also associated with cardiomyopathy.

Dysphagia and decreased gastric motility are common.

Review of previous anaesthetics is useful but does not mean subsequent anaesthetics will be problem-free, for instance, relating to airway or respiratory events. Previous uneventful

use of suxamethonium or volatile agent does not guarantee that they can subsequently be used safely.¹³

Perioperative management

There should be full discussion with the family before surgery to discuss potential risks and benefits of surgery. Sedative pre-medication should be used with caution if at all, as it may cause respiratory depression or reduced respiratory muscle tone. However, anxiety can be high in these patients.

Spontaneous ventilation techniques are often not well tolerated in children with neuromuscular disease. TIVA with propofol and remifentanyl is effective and safe for these patients, provided cardiac function is not severely impaired.

Choice of anaesthetic technique can be very difficult in some children, but must be made on an individual basis. For instance, a child may have a history of difficult intubation, or severe respiratory impairment, but refuses surgery under regional anaesthesia. The child may be willing to undergo a regional anaesthetic block, but these techniques are often technically more challenging in children with neuromuscular disease. It is sensible to avoid large doses of opioids perioperatively, particularly where ventilatory support postoperatively is limited or unavailable.

All cases should be closely monitored, including core body temperature, as these children are prone to both hypo- and hyperthermia. Hyperthermia may occur due to increased muscle activity seen in myotonias, iatrogenic causes or malignant hyperthermia. Monitoring must be continued postoperatively, ideally in a high dependency area. Intensive care, ideally with ventilatory support, should be available for high-risk cases and/or those undergoing major surgery.

Children with neuromuscular diseases have increased sensitivity to non-depolarizing neuromuscular blocking drugs. The use of a nerve stimulator and short acting neuromuscular blocking agents is recommended. Suxamethonium must NOT be used. In the channelopathies, there may be a dramatic rise in serum potassium in response to suxamethonium.¹⁴ Malignant hyperpyrexia (MH) and anaesthesia-induced rhabdomyolysis (AIR) may also be precipitated. The only conditions shown to have a definite link to MH are King-Denborough syndrome, central core disease and Evans myopathy. Patients with other neuromuscular conditions have shown MH-like symptoms under general anaesthesia but the link with true MH remains unclear.¹⁵

Children presenting for muscle biopsy have a 10-20% chance of a positive finding and around half of these have a diagnosis of muscular dystrophy. In these circumstances, avoid volatile agents and use total intravenous anaesthesia (TIVA) or spinal anaesthesia with sedation in preference.

SPECIFIC ORTHOPAEDIC PROCEDURES

Surgery for osteomyelitis and septic arthritis

Antibiotics have a limited role in the treatment of septic arthritis and established osteomyelitis. Infection within a joint is an orthopaedic emergency and requires urgent open or arthroscopic wash out.

Infected bone needs thorough debridement of the connecting sinus tracts and ample curettage of the bone segment to remove dead bone (sequestrum), one of the major causes of recurrence of osteomyelitis. Extensive infection may require one side of the bone to be removed to allow adequate drainage, a process known as decortication. Soft tissues are allowed to heal by secondary intention and regular dressing changes are essential for healing. These children may be chronically unwell, anaemic, with poor nutrition, and they may also have impaired clotting.

Ponseti procedure

This technique involves manipulation and casting to correct congenital clubfoot without surgery (see Figure 3). The ligaments, joint capsules and tendons are stretched under gentle manipulations and a plaster cast is applied after each manipulation. The displaced bones are gradually brought into correct alignment and remodel. The initial phases of this procedure do not require anaesthesia. One stage requires a percutaneous tenotomy, which can be done under local infiltration with lignocaine and/or topical anaesthesia. In 10-30% of cases, a tibialis anterior tendon transfer to the lateral cuneiform may be required when the child is about 3 years old and this will require general anaesthesia with or without regional analgesia.

Application of hip spica casts

Spica casts create stability and immobilise femoral fractures and hip abnormalities. Anaesthesia is usually required to apply



Figure 3. Ponseti casts in a hospital in Nepal

a hip spica. In small children, hip spicas are applied with the child on an elevated narrow table that provides very little head support.

During cast placement, the goals of anaesthesia include haemodynamic stability, appropriate anaesthesia and safe patient positioning. The patient's health status and any pre-existing disease dictate the choice of anaesthetic technique. It is important to remain vigilant when the child is placed on the spica table to ensure that the airway is secure and to prevent nerve damage. Consider intubation in children who weigh <10kg, particularly as the abdomen may be compressed during the application.

Pelvic osteotomies for congenital dislocation of the hip (CDH)

These are used to correct developmental dysplasia of the hip joint to stabilise the hip, allow corrective remodelling and prevent early osteoarthritis. The choice of osteotomy will depend on the severity of the acetabular dysplasia, the presence or absence of congruent hip reduction and the age of the child.

Both general and regional anaesthesia can be used if no contraindications exist. These procedures can be long and can be associated with considerable blood loss. General anaesthesia combined with spinal or epidural analgesia can provide good surgical conditions as well as effective postoperative pain control. Postoperatively, oral pain medication can be given, including paracetamol PO, opioids PO and non-steroidal anti-inflammatory drugs (give NSAIDs short-term to reduce any interference with bone healing).

Slipped upper femoral epiphysis

Slipped upper femoral epiphysis (SUFE) is a condition seen in children who are growing rapidly, and usually presents between the ages of 8 and 15. It is due to weakness in the growth plate that results in slippage of the femoral head from the rest of the femur. The child will often complain of knee or hip pain and may present with a limp.

SUFE is an orthopaedic emergency if the child presents acutely, as further slippage may result in occlusion of blood supply to the femoral head resulting in avascular necrosis. Surgery involves the placement of one or two percutaneous cannulated screws into the femoral head to prevent further slippage. In 20-40% of cases the opposite hip may become affected and therefore the other side is also fixed. (See Figures 4 and 5)

There is a high association of obesity with SUFE. Other associations include endocrine abnormalities especially hypothyroidism and treatment with growth hormone. A high incidence of renal osteodystrophy is also found in this group of patients. Preoperative investigations should therefore include full blood count, urea and electrolytes, thyroid screen and a group and screen for blood.

Figure 4. *Slipped Upper Femoral Epiphysis (SUFE)*

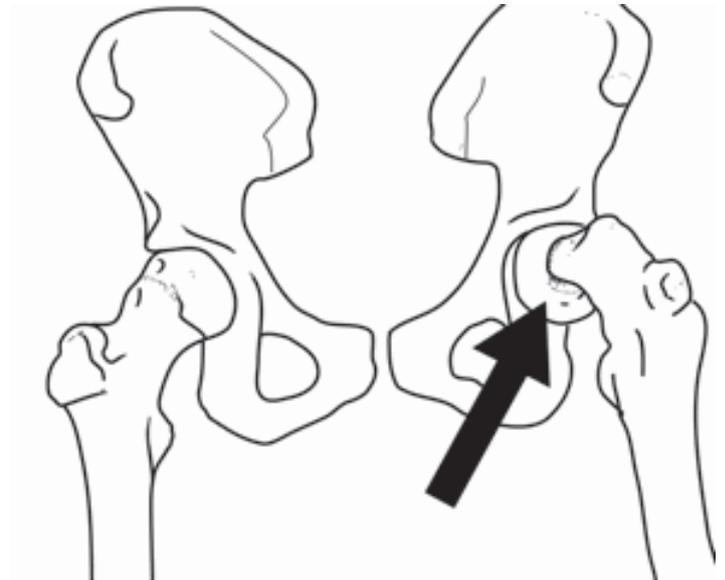


Figure 5. *Insertion of femoral screw for slipped upper femoral epiphysis*



Intraoperatively, the patient may be positioned supine or lateral. Take care to avoid nerve compression or injuries. General anaesthesia, in conjunction with regional anaesthesia, can be used if there are no contraindications to regional anaesthesia. A single shot caudal or epidural can be helpful.

Treatment of limb deformity and leg length inequality

There are many different conditions in childhood that can lead to deformity of a limb or differences in leg length. Treatment depends on the age of patient (how much growth is to be expected) and the pathology causing the limb length inequality.

Epiphysiodesis

This is surgery that slows down the growth of the longer leg over a period of time or corrects angular knee deformities and is only performed in growing children. Small incisions are made around the knee near the femoral and/or tibial growth plates. The growth plates are prevented from growing by the use of small screws and plates called 8 plates. If the desired effect has been achieved and there is still some limb growth left the 8 plates are removed and bone growth resumes.

Application of external fixator

The Taylor spatial frame is an example of an external fixator used to treat complex fractures and limb deformities. Anaesthesia techniques for this type of surgery will depend on the patient's general health and co-morbidities. General anaesthesia and regional analgesic techniques are usually employed. However in patients having an external fixator for acute fractures, use of a regional block must be discussed with the surgeon as it may mask signs of compartment syndrome postoperatively.

Scoliosis/spinal deformity surgery

Specialist orthopaedic teams perform scoliosis surgery. Surgery is generally for cosmetic reasons, particularly in teenagers with idiopathic scoliosis, but in children with cerebral palsy or muscular problems, surgery may improve sitting balance, pain or even prolong life if the deformity is severe. Most cases are idiopathic in origin, and the children are otherwise completely healthy; they may only be underweight or occasionally anaemic. Children with severe scoliosis due to neuromuscular disorders are much higher risk due to impaired cardiorespiratory function.

A posterior approach is the most common technique, with spinal fixation in older children or to insert a growing rod system in pre-pubertal children. A thoracotomy may occasionally also be required which will require lung collapse to optimise access to the spine. The repair is usually done as a single-staged rather than as a two-staged procedure.

Scoliosis surgery is historically an operation associated with major blood loss and significant risk of cord injury. The use of comprehensive blood conservation techniques may allow allogenic blood transfusion to be avoided. Modern techniques, including spinal cord monitoring during surgery by trained electrophysiologists, can minimise risks. Propofol and remifentanyl TIVA provide optimal anaesthesia for spinal cord monitoring and a smooth recovery (volatile anaesthetic anaesthetics can interfere with the spinal cord monitoring). Good postoperative analgesia can be provided with an epidural (if there is no contraindication or concern about cord function post-operatively), or with balanced intravenous analgesia with morphine supplemented with ketamine and oral medications.

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REFERENCES

1. Helander E. Prejudice and Dignity: An introduction to community based Rehabilitation. 2nd edition New York: UNDP; 1999.
2. Atijosan O et al. The orthopaedic needs of children in Rwanda: Results from a national survey and orthopaedic service implications. *J Paediatr Orthop* 2009; **8**: 948-951.
3. Canale ST, Beaty JH. Campbell's Operative Orthopaedics, 11th ed. Mosby, Philadelphia. 2007.
4. Mkandawire NC and Kaunda E. Incidence and patterns of congenital talipes equinovarus (clubfoot) deformity at Queen Elizabeth Central Hospital, Blantyre, Malawi. *East and Central African Journal of Surgery* 2004; **9**: 28-31.
5. Mkandawire NC, Chipofya E, Licoleche et al. Ponseti technique of correcting idiopathic clubfoot deformity. *Malawi Med J* 2003; **15**: 99-101.
6. Lavy CB, Mannion SJ, Mkandawire NC et al. Clubfoot treatment in Malawi - a public health approach. *Disabil. Rehabil.* 2007; **29**: 857-862.
7. Dalens B, Hasnaoui A. Caudal anaesthesia in paediatric surgery. *Anaes Anal* 1989; **68**: 83-89.
8. Patel D. Epidural analgesia for children. Continuing Education in Anaesthesia, *Critical Care Pain* 2006; **6**: 63-66.
9. Lerman J, Nolan J, Eyres R et al. Efficacy, safety and pharmacokinetics of levobupivacaine with and without fentanyl after continuous epidural infusion in children: a multicentre trial. *Anesthesiology* 2003; **99**: 1166-74.
10. Pande R, Pande M, Bhadani U et al. Supraclavicular brachial plexus block as a sole anaesthetic technique in children: an analysis of 200 cases. *Anaesthesia*. 2000; **55**: 798-802.
11. Nolan J. Anaesthesia and pain management in cerebral palsy. *Anaesthesia* 2000; **55**: 32-41.
12. Franklin DL, Luther F, Curzon ME. The prevalence of malocclusion in children with cerebral palsy. *European Journal of Orthodontics* 1996; **18**: 367-643.
13. Ragoonanan V, Russell N. Anaesthesia for children with neuromuscular disease. Continuing Education in Anaesthesia, *Critical Care and Pain* 2010; **5**: 143-147.
14. Driesson J. Neuromuscular and mitochondrial disorders: what is relevant to the anaesthesiologist? *Current Opinion Anesthesiology* 2008; **21**: 350-5.
15. Parness J, Bandschapp O, Girard T. The myotonias and susceptibility to malignant hyperthermia. *Anesthesia Analgesia* 2009; **109**: 1054-64.
16. Gregory's Pediatric Anaesthesia p757-776 5th Edition 2012: Wiley Blackwell.

Update in Anaesthesia

Large airway obstruction in children

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Summary

There are anatomical, physiological and developmental reasons for children to be particularly susceptible to airway obstruction.

Rapid clinical assessment, minimal disturbance, and rapid intervention are important. Summon senior experienced help if available.

PART 1: CAUSES AND ASSESSMENT

Opening and maintaining the airway is fundamental to the treatment of all emergency situations in paediatrics, as in adults. All resuscitation algorithms start with ABC (Airway, Breathing, Circulation) and must be qualified in trauma to include cervical spine control. The commonest cause of paediatric airway obstruction is still the child with depressed conscious level who is not positioned properly or whose airway is not opened adequately by Basic Life Support manoeuvres. Airway foreign bodies are also common and may need rapid intervention. The pattern of infective causes of airway obstruction has changed since the introduction of vaccination programmes against *Haemophilus influenzae* type B. There has been a marked reduction in the incidence of epiglottitis, with a relative predominance now of viral croup and bacterial tracheitis, usually caused by *Staphylococcus aureus*.

Why are children at increased risk from airway obstruction?

There are anatomical, physiological and developmental reasons for children to be particularly susceptible to airway obstruction.

The nares, upper and lower airways are smaller in absolute terms in children. Resistance to air-flow (and thus the work of breathing) increases during quiet, laminar flow breathing in inverse proportion to the fourth power of the radius. A small decrease in radius of the airway increases markedly the resistance to breathing. This is even more noticeable during crying when air-flow is turbulent as resistance is then related to the fifth power of the radius. An example of this amplification effect in the upset child is to compare the increase in airway resistance when the airway narrows from 4mm to 2mm: in the quiet child the airway resistance increases 16-fold but when the child cries the increase is 32-fold.

The infant has a relatively large tongue and the larynx is situated relatively high in the neck, with the epiglottis at the level of C1 at birth, C3 in the infant and C6 from puberty. The laryngeal inlet appears to lie more anteriorly because of its high position. In the infant, the epiglottis is long and omega shaped and angled away from the long axis of the trachea.

The larynx is funnel shaped and is narrowest at the level of the cricoid ring compared with the cylindrical adult conformation, which is narrowest at the level of the vocal cords. The airway is more compressible as cartilage support components are less well developed. Thus, extrinsic pressure from haematomas, neoplasms, vessels or enlarged heart chambers may more readily compress the airway. The collapse of the laryngeal inlet during inspiration is a feature of laryngomalacia and the collapse of the trachea and/or bronchi during expiration occurs in tracheo-bronchomalacia. If the intrathoracic airways are narrowed from whatever cause, the extra work of inspiration and of expiration leads to large swings in intrathoracic pressure and the potential for gas trapping and hyperinflation behind the obstructed airway causing further compression of small airways. During forced expiration efforts, the intrathoracic airways may collapse down exacerbating the gas trapping effect.

Hyperinflation and gas trapping also impair the function of the diaphragm which is unable to contract so efficiently from its optimal length. In infants the diaphragm has a smaller proportion of contractile elements and fewer fatigue resistant muscle fibres. The rib cage is cartilaginous and more compliant, so the diaphragm anchor points are more mobile, leading to wasted inspiratory work and the clinical sign of recession of the chest wall. The chest wall shape in cross-section is circular in the infant compared with the elliptical shape in the older child and the ribs are attached perpendicular to the vertebral column compared with the acute angle of attachment in the older child. This means that the contribution of the "bucket-handle" movement of the rib cage to inspiration is minimal in small infants and also the elastic recoil effect is much less during expiration. The intercostal muscles and accessory muscles of inspiration are also less well developed. Thus, the small infant is very reliant on the diaphragm's contribution to inspiration and thus has few reserves when work of breathing has to increase. This is on top of the already high basal demands placed on the infant respiratory system by the higher rate of metabolism in early life.

The small absolute size of airways in children means that secretions, small airway constriction, oedema or

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compression more readily lead to airway closure and either atelectasis or gas trapping. The interalveolar pores and bronchoalveolar channels do not develop until the ages of 1 year and 8 years respectively so collateral ventilation is not an option around an area of obstruction by these mechanisms.

Thus anatomical, physiological and developmental factors conspire to make the child susceptible to airway obstruction and is exacerbated in disease states (Table 1).

What are the symptoms and signs of airway obstruction?

Signs of foreign body aspiration

Sudden onset of respiratory compromise associated with coughing, gagging, choking, aphonia or stridor suggests foreign body aspiration and this may necessitate emergency basic life support measures for the choking child. Signs of gas trapping behind a foreign body (“ball-valve effect”) may be seen with hyper-resonance of the hemithorax, loss of percussion dullness over the liver, surgical emphysema, tracheal deviation and unequal breath sounds. It is particularly important to think of the possibility of pneumothorax and actively exclude it and treat it promptly. Pneumomediastinum, pneumopericardium and pneumoperitoneum may be seen. Collapse or consolidation of lobes or lungs with bronchial breathing, widespread crackles and expiratory wheeze may all be elicited depending on the cause, site and duration of the airway obstruction.

Signs of increased work of breathing

The increased effort of breathing caused by airway obstruction may produce an increase in respiratory rate for age. A rate >50bpm in an infant and >30bpm in a child may be considered abnormal. However, of even more concern would be respiratory distress associated with a normal respiratory rate, bradypnoea or apnoeic spells which indicate decompensation and exhaustion.

A “see-saw” pattern of chest and abdominal breathing movements is seen in airway obstruction. This sign occurs earlier in younger infants. Recession of the intercostal spaces, subcostal region and sternum are also seen early in young infants and reflect the forces generated by vigorous contractions of the diaphragm and the compliant chest wall. If recessions are seen in older children they indicate severe airway obstruction. Use of the accessory muscles of inspiration (sternomastoids, scalene muscles and intercostals) is associated with tracheal tug, suprasternal and supraclavicular recessions and nasal flaring. Often the child sits upright and may adopt the “tripod” position to improve the mechanical advantage of these muscles in moving the chest wall and that of the diaphragm. In the small infant, an opisthotonic posture may be seen in airway obstruction and head bobbing is a sign of accessory muscle contraction in the infant. Lack of effort associated with deteriorating conscious level may indicate exhaustion and decompensation.

Expiratory grunting is often noted in infants with respiratory distress who are trying to generate auto-CPAP or expiratory braking at laryngeal level to maintain a residual lung volume at end expiration.

Stridor during inspiration is usually a sign of airway obstruction at supraglottic or laryngeal level but can occur in tracheal obstruction also. Stridor during expiration is usually a sign of intrathoracic airway obstruction. Prolonged expiration with wheeze is usually a sign of small airways obstruction as in bronchiolitis or asthma but can occur in large airway obstruction especially due to foreign body or if there is an underlying anatomical abnormality. The volume of stridor or wheeze does not correlate with the degree of airway obstruction. Indeed, the most ominous sign is the “silent chest” where obstruction is so severe that no gas flow is occurring.

Signs of ineffective breathing

Cyanosis, depression of conscious level, slow respiratory rate, the

Table 1. Some causes of large airway obstruction in children

Depressed conscious level

Foreign body

Infection

- Viral: croup, papillomatosis
- Bacterial: epiglottitis, tracheitis, tonsillitis, abscess
- Adjacent to airway

Trauma

Thermal injury

Congenital abnormalities: choanal atresia, choanal stenosis, micrognathia, macroglossia, laryngomalacia, laryngeal web

Neoplasm: haemangioma, lymphoma, mediastinal mass

Peripheral neurological disease

Neuromuscular disease

Iatrogenic: subglottic stenosis, post-intubation stridor, neck haematoma

Anaphylactoid reactions

silent chest despite vigorous respiratory efforts or lack of adequate respiratory effort, apnoeic spells and bradycardia are most worrying signs of ineffective breathing.

Secondary effects of airway obstruction

Airway obstruction may produce hypoxaemia and hypercarbia. Tachycardia, sweating, confusion, restlessness, agitation, anxiety, dyspnoea, inability to speak, peripheral vasoconstriction with pallor or mottling, cyanosis, decreased conscious level, apnoeic spells and bradycardia may occur. Generalised convulsions may occur secondary to hypoxaemia. Hypertension and bounding pulses may be felt and pulsus paradoxus of greater than 20 mmHg may be elicited in older children. Chronic airway obstruction may cause chest wall abnormalities, pulmonary hypertension, right heart failure and obstructive sleep apnoea syndrome.

What investigations are helpful?

The assessment of the child in order to identify and manage airway obstruction is a clinical one. Do not try to examine the child's throat. The pulse oximeter is a very helpful, non-invasive and atraumatic monitor of arterial oxyhaemoglobin saturation and heart rate. However, the readings need interpretation in context with the clinical picture as they are affected by poor perfusion, movement, ambient light and carboxyhaemoglobinaemia (as may occur in smoke inhalation injury) and are less accurate at values below 70%.

Radiology should not be used in the child in extremis before intervening but in the less acute situation may help elucidate chest signs, such as pneumothorax, consolidation, collapse, foreign body, steeple sign in croup, mediastinal mass, etc. It should be carried out at the bedside.

Lateral soft tissue neck films are seldom indicated but may show a foreign body, thumb sign of epiglottitis, prevertebral or a retropharyngeal abscess. CT and MRI scanning have no place in emergency management but are very helpful in cases such as haemangioma, mediastinal mass, or abscess adjacent to the airway.

The process of obtaining arterial, capillary or venous blood gases is likely to cause undue distress which will worsen airway obstruction. In the obtunded child intervention should be immediate and should not await blood gas results. For less severe cases, trends in carbon dioxide levels, pH and oxygen values may be helpful in guiding treatment and in reinforcing the need to intervene. Chronic airway obstruction leads to a respiratory acidosis which induces renal compensatory mechanisms with retention of bicarbonate and a metabolic alkalosis reflected in a high serum bicarbonate level and often a near normal arterial pH.

Making the diagnosis

Some features of the history and examination may be particularly helpful in pointing to a specific diagnosis and they are summarised in Table 2. These clinical signs are suggestive only as each disease process has a spectrum of severity. In individual cases it can be difficult to differentiate between the infective causes and foreign body aspiration. Severe tonsillitis or abscesses near the airway can produce similar symptoms and signs. Oedema of the face, periorbital tissues, tongue and peripheries is suggestive of angioneurotic oedema or anaphylactoid reactions.

Severity scoring system

A scoring system for croup (Table 3) is helpful in assessing severity, response to therapy and of the need for intervention

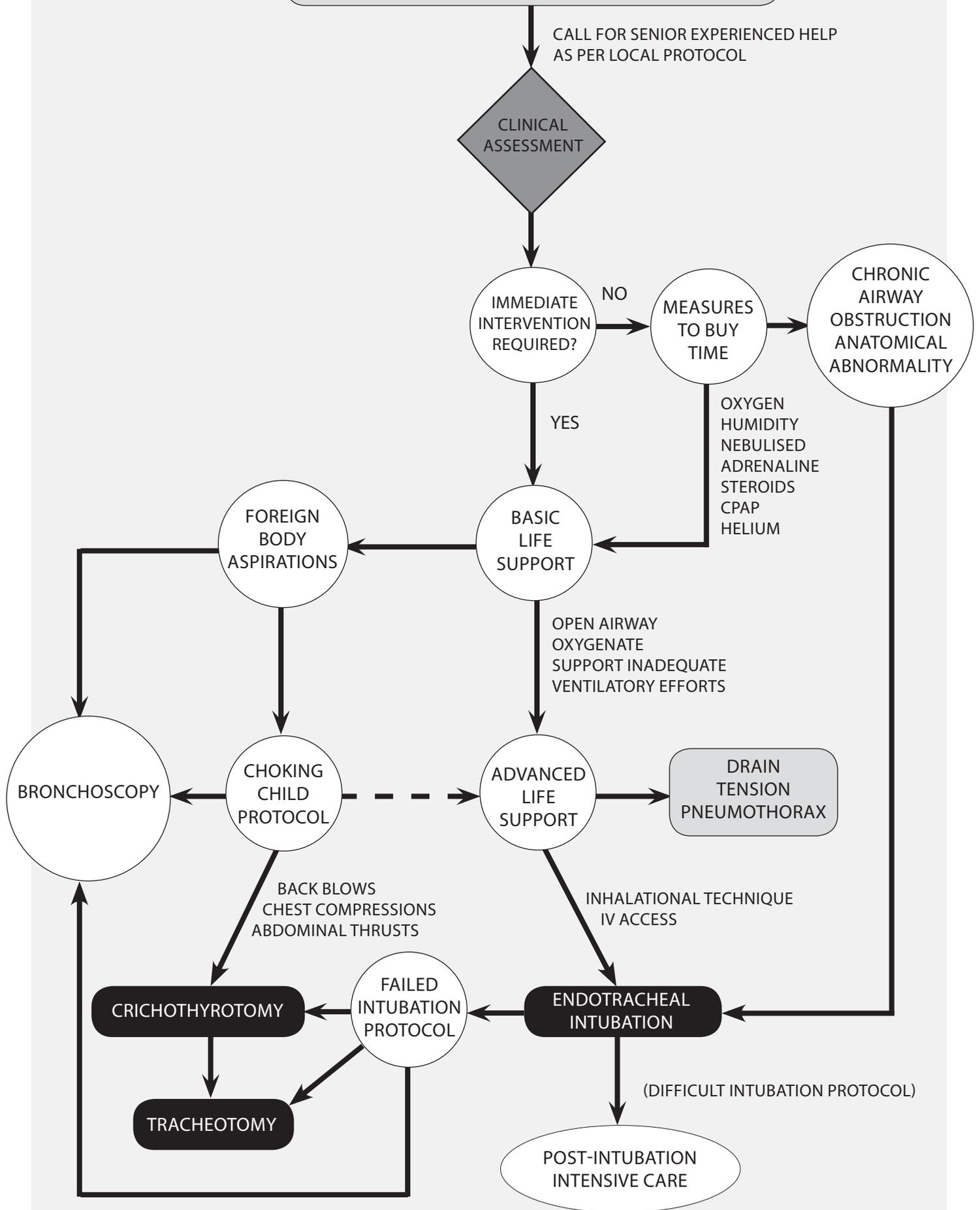
Table 2. Differentiation between croup, tracheitis and epiglottitis

	Croup	Tracheitis	Epiglottitis
Cause	Viral	Staphylococcus aureus Streptococcus	Haemophilus influenzae B
Age	6m – 3y	Any age	2 – 6y
Onset	Gradual	Gradual	Sudden
Pyrexia	Mild	>38°C	>38°C
Abnormal sounds	Barky cough, stridor	Barky cough, stridor	Muffled, guttural cough
Swallowing	Normal	Difficult	Very difficult with drooling
Posture	Recumbent	Sitting	Tripod position
Facies	Normal	Anxious	Anxious, distressed, toxemic

Table 3. Croup score

	0	1	2
Breath sounds	Normal	Harsh, rhonchi	Delayed
Stridor	None	Inspiratory	Insp. + Exp.
Cough	None	Hoarse cry	Bark
Retractions/ flaring	None	Flaring + suprasternal retractions	Flaring + suprasternal + intercostal retractions
Cyanosis	None	In air	In 40% oxygen

Paediatric Large Airways Obstruction



PART 2: MANAGEMENT

Importance of rapid clinical assessment, minimal disturbance and rapid intervention

An assessment from the end of the bed with minimal disturbance should be possible in most cases with the child sitting in the parent's arms and the child should be allowed to adopt the posture in which they are most comfortable. Clinical assessment and a concise history as described in part 1 should allow identification of the need for intervention. A pulse oximeter probe is relatively atraumatic to apply. Gentle physical examination of the chest seeking actively for the important signs described in part 1 should be possible. In some cases, the need for immediate intervention will be obvious. In others, measures to buy time to enable experienced help to arrive may be appropriate. It is often stated that attempts at venous access should not be made as they will upset the child - this is a reasonable view. However, some argue that, in the less ill child and with topical local anaesthesia or ice analgesia of the skin and a skilled paediatric venepuncturist, this is not an issue. Some experienced paediatric anaesthetists are of the view that, in the hypercarbic, obtunded child with a hyperdynamic circulation, establishing venous access is relatively simple and appropriate. Despite this debate, the classical approach of not attempting venepuncture is recommended for the non-specialist anaesthetist.

When is immediate intervention required to open the airway?

Immediate intervention is needed in the choking child or if the child is apnoeic or exhausted and making ineffective respiratory efforts. A silent chest or no stridor (either in the child making maximum effort to breathe or in the exhausted child) are very sinister indications of complete airway obstruction. The simultaneous treatment priorities are oxygenation, opening the airway, improving failing respiratory efforts and relieving tension pneumothorax.

Can I "buy time" prior to intervening to secure the airway?

Steroids improve airway patency in croup, haemangioma, lymphoma and some mediastinal masses. They can be given orally, parenterally or by nebuliser. Prednisolone 4mg.kg⁻¹ orally, or dexamethasone 0.6mg.kg⁻¹ intravenously or intramuscularly, or budesonide 1-2mg by nebuliser are favoured as initial therapy with maintenance by repeated nebuliser therapy or oral prednisolone or parenteral dexamethasone at one quarter of the initial dose every 8-12h for up to 48h. Prompt administration of steroids often pre-empts the need for intubation in most cases of moderate or severe croup.

Nebulised adrenaline 1:1000 standard solution at a dose of 0.5ml.kg⁻¹ (maximum 5ml) diluted if necessary with 0.9% saline to a total volume of 5ml will give a dose of 2-5mg in most cases. It reduces mucosal oedema and acts very rapidly but when stopped may give rise to a rebound worsening of airway obstruction. It is a useful temporising measure. ECG monitoring is recommended although dysrhythmias are seldom a problem.

CPAP - Continuous positive airway pressure acts as an effective splint for the collapsible, compressible paediatric upper airway and can be delivered to infants and children by a well fitting facemask (especially using the Jackson-Rees T-piece circuit) and, in babies, by nasal

cannulae, nasal prongs or a nasopharyngeal airway. It is a very useful measure particularly during the preparation phase prior to intubation and during inhalational induction of anaesthesia. It can be very useful in the management of chronic airway obstruction. A development of CPAP is bi-level CPAP or BIPAP; it is becoming increasingly popular in the management of children.

Prone position +/- nasopharyngeal airway can be useful in babies with congenital upper airway abnormalities where the tongue is relatively large eg hemifacial microsomia, Pierre-Robin syndrome, or Treacher-Collins syndrome. The tongue falls forward from the posterior pharyngeal wall and often improves the airway. In conjunction with nasal CPAP and /or a nasopharyngeal airway, the child may improve markedly with this simple manoeuvre.

Helium is less dense than air or oxygen and gas flow tends to be more laminar which reduces the work of breathing. However, it is not readily available, is expensive and dilutes the inspired oxygen concentration. Breathing a helium-oxygen mixture may be helpful in buying time.

How should I intervene to open and secure the airway?

The algorithm in Figure 1 is a useful guide. If possible, call for expert help.

Basic life support manoeuvres and the choking child

Head tilt, chin lift and jaw thrust are the first basic steps. Physical methods of clearing the airway should only be used if the diagnosis of foreign body aspiration is clear and dyspnoea is increasing rapidly or apnoea has occurred. Do not use finger sweeps as this may push the foreign body further down the airway and may impact it in the laryngeal inlet. Do not try to examine the throat. In infants, immediately carry out five back blows with the heel of the hand with the infant lying prone and head down along your arm which should be resting along your thigh. If the obstruction remains, turn the baby supine and give five chest thrusts as for cardiac compression but more slowly and repeat airway opening manoeuvres, expired air ventilation and cardiac compressions as appropriate. In older infants and children, five back blows with the child prone across the lap and up to five abdominal thrusts exactly in the midline with the child standing, kneeling, sitting or supine should be used.

Advanced life support manoeuvres

Oxygen 100% should be administered by self-inflating bag and mask or anaesthetic T-piece circuit, depending on familiarity. The latter has the advantage that CPAP can easily be applied and the transition from spontaneous to controlled ventilation is simple. It is very important to actively exclude tension pneumothorax and, if present, to intervene with a needle, cannula or drain. This can be inserted under local anaesthesia in the conscious child giving careful attention to technique. Some distress may be caused to the child but this is transient and justifiable in the emergency situation. In the context of a tension pneumothorax secondary to acute airway obstruction, it is very wise to drain the pneumothorax prior to inducing anaesthesia. However, circumstances may dictate simultaneous intervention to drain the pneumothorax and induce anaesthesia to secure the airway if rapid decompensation is occurring.

Endotracheal intubation technique

It is better to have the child in an intensive care or operating theatre

environment. Senior experienced staff only should attempt intubation unless circumstances mandate a life-saving attempt. Some recommend calling for assistance from the ENT team in case a bronchoscopy or tracheostomy is needed. A wide range of paediatric airway and intubation equipment should be to hand and checked.

Inhalational induction of anaesthesia with halothane (increasing gradually to 5%) in oxygen, starting with the child in the position in which they are most comfortable is recommended. Some experts now prefer sevoflurane (starting with 8%) in oxygen but experience in children with critical airway obstruction is limited and sevoflurane is intrinsically less potent making it difficult to gain a sufficient depth of anaesthesia to allow intubation. Remember, alveolar ventilation may be severely compromised and uptake of volatile agents may be very slow. It may take up to 10 minutes to reach a sufficient depth of anaesthesia. As the depth of anaesthesia increases and the patient is moved to the supine position, airway obstruction may occur and it is wise to change position gradually and to add CPAP to splint the airway open. Once the pupils become constricted and central, wait for a further 30 breaths and then perform laryngoscopy and orotracheal intubation. Change to a nasotracheal tube once the child is stabilised and fix securely with tape or a tube holder and fixation system (eg Tunstall, Burtles, Secure-ET).

If intubation is difficult or impossible, a number of techniques using the flexible fiberoptic or rigid bronchoscope are possible and very occasionally blind or retrograde intubation techniques may be employed. The use of a nasopharyngeal airway or the laryngeal mask airway may be helpful in certain cases. All these techniques are for the expert only.

In certain cases of impossible intubation where the child is rapidly deteriorating, the safest option may be to consider an emergency cricothyrotomy using a cannula, 3mm endotracheal tube connector, a T-piece or self inflating bag and oxygen source. Remember that there must also be a patent expiratory pathway to avoid barotrauma. Carbon dioxide levels will tend to rise with this technique. Jet ventilation is not recommended in children due to the risks of overpressure and barotrauma. In other cases, particularly of severe anatomical abnormalities, the safest option may be an emergency tracheostomy under mask anaesthesia and or local infiltration analgesia.

When should I not intervene?

If you are inexperienced with advanced life support measures in children, you should try to maintain oxygenation, airway patency and ventilatory support with basic measures until experienced staff arrive. Advanced life support interventions should be carried out by the most experienced staff present. However, you may have to intervene in the extreme situation to save the child's life. If it is available, it is vital that expert help is called as early as possible to manage children with airway obstruction.

How should I manage the child after the airway is secured?

It is important to ensure that the artificial airway is fixed securely and is correctly positioned. A post-intubation chest X-ray is useful for checking tube position and identifying lower respiratory tract or pulmonary parenchymal changes. Indications for controlling ventilation rather than allowing spontaneous ventilation are:

- Small diameter endotracheal tube
- Child with signs of septicaemia
- Lower respiratory or lung disease
- Child who has sustained a hypoxic insult
- Very abnormal, inflamed or oedematous airway
- Traumatic intubation
- Need to transport the child to another hospital

Sedation, analgesia and muscle relaxation should be given as appropriate. Some children develop post-intubation pulmonary oedema which requires ventilatory support with PEEP and diuretic therapy.

Antibiotic therapy is indicated for likely organisms - the third or fourth generation cephalosporins are favoured with some preferring flucloxacillin for staphylococcal tracheitis. Duration of intubation varies widely from 18-24 hours for acute epiglottitis to days or weeks for those with lung involvement, severe disease and pre-existing congenital anomalies. Some children may require formal investigation of their airway eg endoscopy, foreign body removal or reconstructive surgery.

CONCLUSION

Large airway obstruction in children is a common emergency. If available, senior experienced help should be summoned immediately. Therapy is guided by clinical assessments. All consultants should be competent in the performance of basic life support measures for the choking child and for opening the airway, oxygenating the child and supporting inadequate ventilation. Measures to buy time can be very helpful in croup and where the airway anatomy is abnormal. Advanced life support measures are for experienced staff but you may have to intervene immediately to save a child's life. The results of correct management are excellent.

FURTHER READING

- Morton NS. Large airway obstruction in children. *Update in Anaesthesia* (2004) **18**:44-49. Available from: http://www.wfsahq.org/components/com_virtual_library/media/92df8f6507212855c41926db87562732-9872ec3f9b81c17cad5c78fcbaa5d71c-Large-Airway-Obstruction-in-Children-Update-18-2004-.pdf
1. Kissoon N. Acute respiratory emergencies. In: Duncan A, ed. *Paediatric Intensive Care*. London: BMJ Books, 1998:9-40.
 2. Robinson D. Airway management. In: Morton N, ed. *Paediatric Intensive Care*. Oxford: Oxford University Press, 1997:81-108.
 3. Advanced Life Support Group. *Basic Life Support. Advanced Paediatric Life Support*. 5th ed. London: BMJ Publishing Group [Wiley-Blackwell], 2005:19-33.
 4. Morton NS, Doyle EI. Case Presentations in Paediatric Anaesthesia and Intensive Care. Oxford: Butterworth-Heinemann, 1994.
 5. Morton NS. Large airway obstruction in children. Part 1: Causes and Assessment. *Royal College of Anaesthetists Newsletter* 1999; Issue 47: 159-162.
 6. Kissoon N. Acute respiratory emergencies. In: Duncan A, ed. *Paediatric Intensive Care*. London: BMJ Books, 1998:9-40.
 7. Robinson D. Airway management. In: Morton N, ed. *Paediatric Intensive Care*. Oxford: Oxford University Press, 1997:81-108.
 8. Morton NS, Doyle EI. Case Presentations in Paediatric Anaesthesia and Intensive Care. Oxford: Butterworth-Heinemann, 1994.
 9. Special Issue: The Pediatric Airway. *Pediatric Anesthesia*, July 2009;19s1:III-IV, 1-197. Available at: <http://onlinelibrary.wiley.com/doi/10.1111/pan.2009.19.issue-s1/issuetoc>

Bronchoscopy for a foreign body in a childBased in part on: Dix P. Bronchoscopy for a foreign body in a child. *Update in Anaesthesia* (2003); **17**: 20-21.

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Summary

1. The clinical effect of an inhaled foreign body depends where the foreign body becomes impacted – most small foreign bodies pass into the distal airway, but larger objects may become impacted in the supraglottic area to cause choking, or in the trachea to cause severe airway distress.

2. Children often inhale foodstuff such as peanuts – organic foreign bodies can cause airway oedema and hyperreactivity, which may be worsened by anaesthetic gases. Topical lignocaine to the airway assists smooth anaesthesia.

3. A foreign body may become impacted distally or exert a ball valve effect to allow inflation but not deflation. For this reason, you must maintain spontaneous ventilation, and allow rigid bronchoscopy prior to intubation.

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INTRODUCTION

Inhalation of a foreign body (FB) is a potentially life threatening event, with boys in the age range 1 to 3 years most at risk. Clinical features of FB inhalation vary from acute upper airway obstruction to pneumonia due to distal airway collapse, depending on where the FB becomes impacted in the airway and when the child presents. This article will discuss common presentations of FB inhalation, and a suggested technique to remove the FB safely.

PATHOPHYSIOLOGY

The clinical features of inhaled FB depend on the size and nature of the FB, and where it becomes impacted in the airway. Resistance to gas flow in the airway is related to the fourth power of the radius, so a small reduction in airway radius in a child will result in a large increase in resistance to airflow. Inhalation of an organic FB may result in airway hyperreactivity and mucosal oedema. The occurrence of oedema in addition to the physical presence of the FB results in a rapid increase in airway resistance. Coupled with the high oxygen consumption of infants and small children, hypoxia may occur rapidly if the FB is lodged in a major airway.

PRESENTATION

Presentation may be acute in the case of a supraglottic FB or FB in a major airway, or more insidious if the FB is distal and presentation is delayed. The signs and symptoms depend on the position of the FB in the airway.

The history may help in the diagnosis, for example sudden onset of respiratory distress while playing with small objects. A child who is actively coughing after a witnessed choking event has a supraglottic FB and should be managed according to choking algorithms - encourage the child to cough to clear the obstruction, use alternating back blows and chest thrusts (abdominal thrusts in the older child), or standard CPR if the child becomes unconscious (see Figure 1 and Paediatric Resuscitation article p 265). It is essential to intervene early – children with untreated airway obstruction due to a supraglottic FB do not often survive to reach hospital.



Figure 1. Back blows in a choking infant with a FB above the vocal cords

The vocal cords are the narrowest part of the airway in the child - in the majority of children who inhale a FB and reach hospital, the FB has passed between the vocal cords to a distal main bronchus (usually right main bronchus). Occasionally, the FB becomes impacted in the larynx or in the trachea:

Signs and symptoms of laryngeal or tracheal obstruction:

- Cough
- Choking
- Respiratory distress
- Cyanosis, desaturation
- Stridor
- Tachypnoea.

Sign and symptoms due to obstruction of a main bronchus:

- Respiratory distress
- Tachypnoea
- Wheeze
- Absent breath sounds on the affected side.

If the FB is small and has lodged in a distal airway, there may be no clinical findings during the acute phase, even following a clear history of FB inhalation. Air trapping might be seen on a chest X-ray on expiratory films, due to a “ball valve effect”. Initially an air bronchogram may be seen, with later evidence of atelectasis distal

to the obstruction. The child may present a few weeks later with chronic cough, chest infection, and signs of consolidation affecting one or more lobes.



Figure 2. Hyperlucent appearance of the right lung on this expiratory chest Xray demonstrates air trapping from a foreign body lodged in the right main bronchus. Image reproduced with kind permission of the Department of Radiology, Virginia Commonwealth University Medical Center, from www.pedsradiology.com

You should consider FB aspiration in every child presenting with cough or stridor. The differential diagnosis includes infective causes such as:

- Croup (viral infection – typical barking cough with stridor and respiratory distress as a late complication),
- Acute epiglottitis (*Haemophilus influenzae* type B infection – causes supraglottic cellulitis with severe sore throat, fever, toxic, ‘muffled voice’ and drooling),
- Acute tracheitis (Staphylococcal infection – child toxic, unwell).

Peanut oil is particularly irritant to the airways and can cause local mucosal oedema as well as a chemical pneumonitis picture, which may be the only presenting factor. A FB in the upper part of the oesophagus (hypopharynx) may present with respiratory distress due to external compression of the trachea.

PREPARATION, INVESTIGATION AND EXAMINATION OF THE CHILD

To a large extent this will be dictated by the clinical condition of the child, and intervention should be planned carefully with the ENT surgeon (if available).

You should make a rapid assessment of the child. Pay particular attention to examination of the airway and chest. If the FB has impacted in the airway but is not causing complete obstruction, the clinical signs may help to localize the site of obstruction:

- Inspiratory stridor - glottic or supraglottic FB

- Expiratory wheeze - infraglottic FB.

Measure the oxygen saturation with a pulse oximeter and give high flow oxygen if required.

If the child is stable a chest Xray may be helpful in localising the FB, although the majority of FBs will not be radio-opaque. In the acute situation few other investigations are indicated.

The child should be starved according to the recommended guidelines, but careful judgement of clinical priorities is required in a child with acute respiratory distress. Consider anticholinergic premedication to decrease airway secretions (atropine 20mcg.kg⁻¹ PO). This will also reduce the vagal tone and avoid bradycardia during airway instrumentation. Sedative premedication should not be used.

TYPES OF BRONCHOSCOPE

A rigid bronchoscope should be prepared, and the anaesthetist should be familiar with the equipment available locally.

Rigid bronchoscopy has several clear advantages:

- Complete airway control
- Better view of the bronchial tree
- Larger channels through which to pass instruments and withdraw FBs.

In older children, the rigid bronchoscope only allows limited access to the upper lobes and more distal airways.

Two types of rigid bronchoscope are available. The older Negus bronchoscope is the original rigid bronchoscope, and has a tapered shape.

The Storz ventilating bronchoscope is the most commonly used rigid bronchoscope. It is available in a variety of sizes and lengths from 2.5mm internal diameter. It has a side port to which the anaesthetic breathing circuit can be attached to provide anaesthesia during airway examination (see Figure 2). This allows safe examination of all children, including neonates.



Figure 3. Storz ventilating bronchoscope with anaesthetic T-piece attached

The Hopkins rod lens telescope is inserted through the lumen of the bronchoscope, allowing a clear view of the endobronchial tree. A wide range of instruments including long grasping forceps are available to enable the FB to be retrieved (see Figure 3).

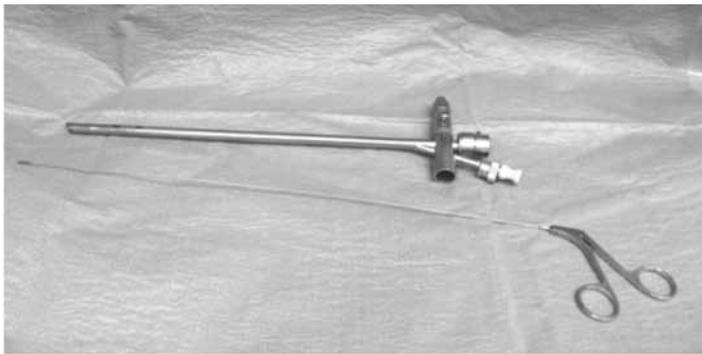


Figure 4. Storz ventilating bronchoscope with long grasping forceps

The anaesthetic T-piece circuit is attached to the sidearm of the bronchoscope to allow delivery of oxygen and anaesthetic gases during the procedure. The presence of the telescope, with the viewing end occluded, results in a closed system, through which either spontaneous or controlled ventilation may occur. When the telescope is removed the system is open, allowing only spontaneous ventilation, unless the operator occludes the port with their thumb. The telescope occupies a significant proportion of the bronchoscope through which expiration must occur. It may be necessary to remove the telescope periodically to allow adequate breathing through the lumen of the bronchoscope, especially when using smaller diameter bronchoscopes.

A Sanders injector can be attached to the sidearm to enable controlled ventilation, when the telescope is not being used. It uses the Venturi effect to entrain oxygen-enriched air. The entrainment of air means that it is not possible to deliver high oxygen concentrations, which may not maintain adequate patient oxygenation. Maintain anaesthesia with IV anaesthetic agents. In smaller children barotrauma can easily occur, so this is not the best technique for them.

ANAESTHESIA

Bronchoscopy is performed under general anaesthesia. The anaesthetic machine and other equipment should be checked, especially suction equipment. A range of sizes of tracheal tubes should be available, in case intubation is required urgently. Airway oedema reduces the tracheal diameter and a smaller tube than usual may be required – prepare a range of sizes.

Monitoring including pulse oximetry, ECG, non-invasive blood pressure, and capnography should be applied. Intravenous access should be secured prior to induction. If the child is distressed this can be performed immediately after induction.

A senior anaesthetist and ENT surgeon should be present at induction, along with the most skilled anaesthetic assistant available. Good communication between all members of staff is vital with an agreed plan for how the case will proceed. We recommend inhalational induction using either sevoflurane or halothane in 100% oxygen. There is much debate about the relative advantages of halothane and sevoflurane:

Advantages of sevoflurane:

- Less airway irritation
- Greater cardiovascular stability. Arrhythmias are a potential

problem with halothane, and are worsened by hypoxia, hypercapnia and high circulating levels of catecholamines

- More rapid onset.

Advantages of halothane:

- Longer lasting anaesthetic effect. This allows more time for airway manipulation without the child becoming too lightly anaesthetised and developing laryngospasm
- Often more readily available
- Lower cost.

The choice will be dictated by personal experience and preference, and also local availability. Ether is very slow in onset and difficult to use for inhalational induction, and is not recommended in this context.

The key to successful bronchoscopy is administration of topical local anaesthetic to the airway – this allows the surgeon to insert the bronchoscope and remove the FB without the child coughing. A safe dose of local anaesthetic must be used ($4\text{mg}\cdot\text{kg}^{-1}$ topical). Draw up the required dose into a syringe prior to the start of anaesthesia; attach a long cannula with the needle removed. This is used to apply the correct dose of local anaesthetic to the cords (see Figure 5)

Remember - 1% lignocaine contains $10\text{mg}\cdot\text{ml}^{-1}$ lignocaine. The safe dose for a 8kg child is 3.2ml.



Figure 5. Use of a cannula to administer topical lignocaine

Spontaneous ventilation should be maintained if possible, although occasionally you might need to use gentle mask ventilation. Aggressive positive ventilation increases the risk of hyperinflation and pneumothorax if the FB is exerting a ball valve effect, allowing inflation but not expiration. Positive pressure ventilation is also likely to dislodge the FB distally. For this reason muscle relaxants are avoided.

After induction, site a cannula if it is not already in place, and discontinue nitrous oxide if used. Wait until the child is deeply anaesthetized before laryngoscopy. This can take a long time due to the reduced airflow. Apply topical lignocaine to the larynx and trachea as described above (maximum dose $4\text{mg}\cdot\text{kg}^{-1}$) – if possible apply the lignocaine to the cords and also between the cords. Rigid bronchoscopy can be performed after a few minutes.

During bronchoscopy, maintain anaesthesia by connecting a T-piece to the sidearm of the Storz rigid bronchoscope. Do not intubate prior to rigid bronchoscopy, due to the risk of dislodging or fragmenting the FB possibly leading to complete airway obstruction. If the child desaturates during bronchoscopy of one lung, the bronchoscope can be withdrawn into the trachea to allow re-ventilation of both lungs, before a further attempt at bronchoscopy is made. The telescope might also need to be removed from the bronchoscope to allow adequate gas flow. Observe chest movements during bronchoscopy and monitor carefully at all times; the child may have little respiratory reserve and will desaturate very quickly. The bronchoscopist and anaesthetist must work closely together.

After removal of the FB the airway can be maintained using a face mask, tracheal tube or laryngeal mask, depending on the condition of the child. If the child has copious distal airway secretions, intubate and suction the airway, with gentle on-table physiotherapy if required.

Discontinue the anaesthetic, administer 100% oxygen, and observe the child carefully until awake and extubated. Address other symptoms at the same time, for example give IV fluids to counter dehydration.

Postoperatively, you must monitor the child for stridor and airway obstruction due to oedema. The child may have a sore throat, which can be managed with simple analgesia such as paracetamol and NSAIDs rather than opioids.

The child may need HDU care depending on their condition, particularly if presentation has been delayed. If the child has a secondary chest infection regular physiotherapy and antibiotics should be given (e.g. co-amoxiclav).

Humidified oxygen should be given for 24 hours if required, particularly if the child has low oxygen saturations and/or stridor. If worsening stridor occurs, nebulised adrenaline 1:1000 may be useful (0.5ml.kg⁻¹, maximum 5ml). Dexamethasone 250mcg.kg⁻¹ IV at induction, followed by 100mcg.kg⁻¹ 6 hourly for 24 hours has also been recommended.

LEARNING POINTS

1. The clinical effect of an inhaled foreign body depends where the foreign body becomes impacted – most small foreign bodies pass into the distal airway, but larger objects may become impacted in the supraglottic area to cause choking, or in the trachea to cause severe airway distress.
2. Children often inhale foodstuff such as peanuts – organic foreign bodies can cause airway oedema and hyperreactivity, which may be worsened by anaesthetic gases. Topical lignocaine to the airway assists smooth anaesthesia.

3. A foreign body may become impacted distally or exert a ball valve effect to allow inflation but not deflation. For this reason, you must maintain spontaneous ventilation, and allow rigid bronchoscopy prior to intubation.

CASE HISTORY

A 4 year-old boy presented to the emergency department with a history of coughing and choking while he was lying on his back playing with some coins. His mother reported that he went blue for a few seconds, and the child said that he had “swallowed” a coin.

On examination he was upset, but well. He was afebrile, and had a respiratory rate of 20min⁻¹ with no recessions. Chest auscultation revealed reduced breath sounds in the right lower lobe. A chest Xray showed a radio-opaque sphere in the area of the right main stem bronchus. Ametop local anaesthetic cream was applied to both hands.

The child was transferred to the operating theatre. The anaesthetic machine, suction equipment, and laryngoscopes were checked. The consultant ENT surgeon was present. A pulse oximeter and ECG leads were applied, and a 22G cannula was sited. Atropine 10mcg.kg⁻¹ and dexamethasone 250mcg.kg⁻¹ were given IV. Anaesthesia was induced using 3% halothane in oxygen breathed spontaneously via a face mask and Ayres T-piece circuit. After several minutes, when deep anaesthesia had been achieved, laryngoscopy was performed and the cords sprayed with 2mls of 4% lignocaine. A 3.5mm rigid bronchoscope was introduced into the trachea. The coin was retrieved uneventfully from the right main bronchus, and the bronchoscope was withdrawn from the airway. The halothane was discontinued, and the airway maintained with a face-mask until the child was fully awake.

Humidified oxygen was given overnight on the ward, and the child was monitored with pulse oximetry. The child was discharged home the next day.

REFERENCE

Dix P. Bronchoscopy for a foreign body in a child. *Update in Anaesthesia* (2003): 17: 20-21. Available from: http://www.wfsahq.org/components/com_virtual_library/media/b27816bcf8b61c6e6fa0cc6a30571d56-424afcbfe58e46b3dc60622590113cad-Bronchoscopy-for-a-Foreign-Body-in-a-Child--Update-17-2003-.pdf

Update in Anaesthesia

Anaesthesia for emergency paediatric general surgery

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Summary

Emergency surgery for bowel obstruction in children presents many challenges for the anaesthesia care provider. Delayed presentation and sepsis have a profound physiological impact on many organ systems. Hospital stays elongated by postoperative complications, and the lack of appropriate paediatric intensive care facilities also contribute to the high morbidity and mortality of this surgical population. Mortality in neonates is even higher.

Children have a great reserve and ability to heal but also the potential for sudden decompensation. Good outcomes rely on meticulous perioperative planning, proper training, equipment, and basic supplies. A team approach involving the nurses, laboratory technicians, paediatricians and surgeons is essential.

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INTRODUCTION

Paediatric emergency general surgery in Sub-Saharan Africa has a high morbidity and mortality with multiple contributing factors. Delayed presentation and diagnosis, hospital stays elongated by postoperative complications, and the lack of appropriate paediatric intensive care facilities all contribute to the overall mortality of this surgical population. The mortality in the neonates (less than 6 months) is even higher.

The following paediatric abdominal surgical emergencies were documented in a case series from a teaching hospital in southeastern Nigeria, and provide a description of the typical paediatric surgical practice in sub-Saharan Africa:¹

	%workload
• Typhoid intestinal perforation (TIP)	19
• Intussusception	17
• Obstructed hernia	15
• Neonatal intestinal obstruction	10
• Appendicitis	10
• Trauma	7
• Ruptured omphalocele/ gastroschisis	7
• Hirschsprung's disease	6
• Adhesive bowel obstruction	6
• Malrotation	4

The incidence of typhoid perforation varies between regions; it follows infection with *Salmonella typhi*, the usual source of infection being contaminated water supplies.

Patients frequently present late, often with

signs of severe illness, and careful preoperative assessment and resuscitation is essential. Severe pre-existing metabolic abnormalities, including acidosis and dehydration, will be a challenge for any anaesthesia care provider, but in an environment with limited anaesthesia, surgery and intensive care resources, the challenges are even greater.

This article will review the pathophysiology of intra-abdominal emergencies in children; how to construct an anaesthesia plan for such patients; intraoperative and postoperative problems; and typical case presentations. We hope to provide a greater understanding of general surgical emergencies in children, and to assist with the management of these challenging patients.

PATHOPHYSIOLOGY OF BOWEL OBSTRUCTION

The normal bowel contains gas and the sum of food and salivary, gastric, biliary, pancreatic, and intestinal secretions. Intrinsic or extrinsic blockage of the small bowel leads to accumulation of secretions that dilate the intestine proximal to the obstruction. Patients with delayed presentation may have a diminished oral intake for many hours, and perhaps even days or weeks, but intestinal secretions continue so that the bowel remains full of fluid. Vomiting is an important sign of obstructed bowel in children; the nature of the fluid vomited will suggest the level of the obstruction. Green coloured 'bilious' vomiting is characteristic of small bowel obstruction.

If the intraluminal pressure of the obstructed bowel exceeds the capillary and venous pressure in the bowel wall, intestinal absorption and lymphatic drainage decreases and the bowel may become ischaemic; this is a dangerous

development that may lead to perforation, but it is difficult to predict when this will occur.

Bacterial translocation

Once the bowel becomes ischaemic, bacteria will pass into the peritoneum by a process known as bacterial translocation, even if the perforation cannot be visualized. The colonized fluid is then transported via the lymphatic channels into the thoracic duct, which drains into the central venous system. This allows bacteria to enter the circulation, which in turn causes septicaemia. Studies have shown that bacteria injected into the peritoneum can be cultivated from peripheral blood only six minutes after the injection into the peritoneum, confirming the extremely rapid flow from the peritoneum into the systemic circulation. Bacterial translocation will produce a cascade of events, which will impact multiple systems. Septicaemia is common in children with delayed presentation due to an abdominal emergency, and a child presenting for emergency paediatric surgery in this condition will provide many challenges to the anaesthetist.

DEFINITION OF PAEDIATRIC SEPSIS

The International Paediatric Sepsis Consensus Conference (2005) attempted to develop specific criteria for an international definition of sepsis. These experts determined that the presence of two of the following four criteria, one of which must be abnormal temperature or leukocyte (white blood cell) count, along with a suspected or proven infection, would constitute the definition of sepsis. The infection could be bacterial, viral, fungal or rickettsial in origin.² In most patients with a surgical abdomen, the infection would be a 'suspected' bacterial infection.

The criteria for the definition of sepsis in children are as follows:

- Core (rectal, oral) temperature of $>38.5^{\circ}\text{C}$ or $<36^{\circ}\text{C}$.
- Tachycardia in the absence of external stimulus, chronic drugs or painful stimuli, or otherwise unexplained elevation, for over a 30 minute to 4 hour period, OR, for children

<1 year, bradycardia in absence of drugs, vagal stimulation or congenital heart disease, or otherwise unexplained bradycardia, for over a 30 minute time period (see Table 1).

- High respiratory rate or mechanical ventilation for an acute problem not related to a neuromuscular disease or for just receiving general anaesthesia (see Table 1).
- Leukocyte count elevated or depressed for age, not related to chemotherapy.

The criteria for definition of severe sepsis in children is 'sepsis' PLUS one of the following:²

- Cardiovascular dysfunction,
- Acute respiratory distress syndrome (proven need for $>50\%$ inspired oxygen ($\text{FiO}_2 >0.5$), to maintain arterial saturation $>91\%$),
- Two or more organ dysfunction (respiratory, renal, neurologic, hematologic, or hepatic),

The presence of organ dysfunction can be determined clinically or with basic lab tests such as platelet count, creatinine level, or bilirubin level. This allows the clinician at the bedside to diagnose sepsis without sophisticated tests, and prompts the appropriate interventions. It is important to understand that the paediatric surgical patients who require emergency surgery could have an assortment of organs that are functioning abnormally; this information will help you to plan the perioperative care for these very ill patients.

Cardiovascular dysfunction

Children compensate for a decrease in circulating volume and may not become hypotensive until long after septic shock develops, unlike in adult shock patients where hypotension is commonly seen.

Some investigators have defined the cardiovascular symptoms of septic shock as tachycardia with signs of decreased perfusion including:²

- Decreased peripheral pulses compared to central pulses
- Altered alertness

Table 1. Criteria for definition of sepsis in children of different ages

Age Group	Heart Rate, Beats/Min	Heart Rate, Beats/Min	Respiratory Rate, Breaths/Min	Leukocyte Count, Leukocytes x $10^3/\text{mm}^3$	Systolic Blood Pressure, mmHg
	Tachycardia	Bradycardia			
0 days to 1 wk	>180	<100	>50	>34	<65
1 wk to 1 mo	>180	<100	>40	>19.5 or < 5	<75
1 mo to 1 yr	>180	<90	>34	>17.5 or < 5	<100
2-5 yrs	>140	N/A	>22	>15.5 or < 6	<94
6-12 yrs	>130	N/A	>18	>13.5 or < 4.5	<105
13 to <18 yrs	>130	N/A	>14	>11 or < 4.5	<117

- Capillary refill >2 seconds
- Cool extremities
- Decreased urine output.

If a child presents with signs above and with hypotension and tachycardia, this indicates that they have lost a significant amount of blood (e.g. from trauma), or they are severely volume depleted (e.g. due to intra-abdominal pathology). A child presenting for surgery with this picture requires significant fluid resuscitation prior to induction of anaesthesia.

Blood volumes in children are small in absolute terms (75-80 ml.kg⁻¹). Every paediatric patient will need to be weighed accurately prior to starting fluid resuscitation. There are many references that discuss assessment of percentage of deficit, but in general, if a patient is dehydrated, a fluid bolus of 10ml.kg⁻¹ normal saline (0.9% saline) or a balanced salt solution such as lactated Ringer's, should be given and repeated as required. A child with a 'surgical' abdomen may typically require repeated IV boluses up to 30-50ml.kg⁻¹ preoperatively to restore adequate intravascular volume. If repeated crystalloid boluses are required, greater than 30ml.kg⁻¹, consider blood transfusion.

Renal system and fluid and electrolyte management

Decreased circulating volume and hypotension also trigger sodium and water conservation by the kidneys through decreased urinary output. Remember that the developing child does not have the same anatomical and functional renal capacity as the adult patient, thus maximum concentrating capacity may not be demonstrated. Increase in sympathetic tone in shock causes constriction of the renal arteries, which reduces blood flow to the kidneys and decreases filtration and urine output. This low flow state causes the kidney to release vasopressin (antidiuretic hormone, ADH), a potent vasoconstrictor, and activates the renin-angiotensin-aldosterone hormone pathway which results in further decrease of blood flow to the kidneys, as well as generalized vasoconstriction. All of these actions are focused on improving systemic blood pressure, blood flow, and maintaining tissue perfusion. Note the above mechanism describes the normal mechanisms that will occur in all patients with fluid loss into the gut; some may have the abnormal 'syndrome of inappropriate anti-diuretic hormone' secretion; SIADH - see below.

Patients with septic shock typically present with acute changes in their fluid status including:

- Decreased oral intake
- Fever
- Fluid shift into the gastrointestinal system
- Electrolyte disturbances
- Pronounced reduction in intravascular volume.

A detailed history will help determine the period of decreased fluid intake and also the type of fluid used for resuscitation by the caregivers or the medical care provider prior to arriving at your institution. A question to the patient's mother (or caregiver) regarding frequency of a wet diaper/nappy in the last 24 hours will help one assess intravascular status and renal function.

'Inappropriate' ADH (vasopressin) secretion is common in the 'sick' child, and children presenting with abdominal pathology are at particular risk of perioperative hyponatraemia. It is important to avoid intravenous fluids with a low sodium content at all times in these patients, especially if they are requiring fluid at more than normal maintenance rate.

Unfortunately, iatrogenic harm is common in paediatric fluid management, particularly if low sodium containing fluids such as 4% dextrose 0.18% saline are used for resuscitation. This may lead to symptomatic hyponatraemia (low plasma sodium), which may complicate diagnosis and delay surgical intervention. For example, if hypotonic fluids are used for fluid resuscitation in a child with an acute abdomen, the plasma sodium may fall to extremely low levels (<125mmol.l⁻¹), and the child may have seizures. This causes diagnostic confusion if the child is investigated for a seizure disorder when in fact the child has dehydration secondary to bowel obstruction. Severe hyponatraemia may lead to irreversible brain damage in children, but can be avoided by using isotonic fluids for resuscitation.

Early symptoms of hyponatraemia include nausea and headache. Later symptoms can include confusion, seizures and even respiratory arrest. A seizure caused by hyponatraemia is very difficult to control with benzodiazepines and barbiturates until the sodium level reaches 120mmol.l⁻¹. This scenario may be extremely dangerous without adequate critical care facilities to manage ventilation and cardiac monitoring since the tendency is overaggressive treatment of the seizures with long-acting benzodiazepines causing respiratory depression. A bolus of hypertonic saline (3% sodium chloride) 1ml.kg⁻¹ should be given, and may be repeated if the seizures continue. Over correction must be avoided; the rate of correction of plasma sodium should not exceed 8mmol.kg.d⁻¹.

The renal system in children is immature, which affects the ability to compensate during periods of decreased oral intake, emesis, and bowel obstruction. For example, in the first year of life, the renal plasma flow and the GFR are approximately one half that of the adult patient. Additionally, the fractional excretion of sodium does not reach adult levels until 6 months of age. Hyponatraemia is common at presentation in children with an acute abdomen, again reinforcing the importance of

only using isotonic fluids (0.9% saline, Ringer's) to expand the circulating blood volume in the dehydrated patient.

Large extracellular fluid deficits due to poor intake, excessive losses and poor renal capability to compensate may require aggressive fluid resuscitation with close monitoring of fluid input, urine output and pulmonary status. An indwelling bladder catheter may be necessary, and the respiratory rate and oxygen saturation (if available) should be monitored closely. Even if a patient receives 50-70ml.kg⁻¹ of intravenous fluids, pulmonary oedema is rare in this patient population with previously normal cardiovascular function.

Haematologic system

Altered production and function of white blood cells (WBC), red blood cells (RBC), and platelets is common in children with bowel obstruction and sepsis. Sepsis may be associated with anaemia, or the haemoglobin level may be artificially high due to severe dehydration. Anaemia in sepsis may develop from haemodilution due to early fluid resuscitation and also anaemia of inflammation (decreased erythropoietin production, decreased bone marrow response to the erythropoietin, and decreased RBC survival).³ Chronic anaemia is also common in this patient population. The platelet count may be reduced in sepsis, or the function may be abnormal, which may lead to excessive bleeding. Coagulation abnormalities frequently occur in sepsis. The more delayed the presentation of bowel obstruction, the greater the chance of disseminated intravascular coagulation (DIC). DIC is characterized by intravascular thrombosis and secondary tissue ischaemia, which leads to abnormal bleeding.⁴

In many hospital settings, the ability to perform a bleeding study in a patient with suspected DIC is impossible, and a clinical diagnosis must be attempted. The clinician should have a low index of suspicion for coagulopathy in a patient with sepsis, particularly if there is prolonged bleeding with simple interventions such as insertion of IV lines. Whole fresh blood, less than 48 hours old, is available in the blood bank in most rural hospitals, which is a good option for children who undergo emergency abdominal surgery as it will improve haemoglobin, platelet count and coagulation factors. A dose of 20ml.kg⁻¹ blood in divided doses is a good starting point for children requiring blood transfusion. The heart rate, blood pressure and respiratory status should be monitored. Blood that is needed for surgery should be given immediately (less than one hour) before the surgery starts, to maximize the function of coagulation factors and platelets. If blood is donated immediately prior to surgery, it can be given without refrigeration, as the warmth of the recently donated blood will assist in keeping the temperature normal during surgery, for instance during an exploratory laparotomy. If possible, blood

should not be warmed by placing into a bath of warm water; water that is too hot may cause haemolysis of the red blood cells, which would produce a massive release of potassium. The hyperkalaemia could result in sudden cardiac arrest immediately after the blood is transfused. Water that is too hot to submerge your hand in for over 5 seconds will be too hot to warm IV fluids and should not be used.

CLINICAL APPLICATION - CASE EXAMPLE

A previously healthy 10-month-old male infant presents to the clinic with a five day history of sudden crying, irritability, and sweating but with periods where the child seems to be acting normally. The child has been able to take fluids during the times where he appears normal but the frequency of pain and sweatiness has increased in the last two days and the intake has significantly decreased. The mother denies any fever until yesterday but over the last 4 days the child has had loose stools described as some blood and mucus with a frequency of 4-6 stools per day.

The mother took the child to a local dispensary where he was given IM antibiotics and some intravenous fluids while being monitored. After 12 hours of fluids and no improvement, the child was transported to the referral hospital (6 hours by taxi) where he presented with abdominal tenderness and distention, decreased peripheral perfusion, and breathing difficulties. Vital signs: HR= 200bpm; RR= 40min⁻¹; BP 50/30 (very weak pulses); oxygen saturation (no monitor available) but patient appears pale. Initial labs: Na⁺= 126mmol.l⁻¹; creatinine

Figure 1. *Intussusception. Upright abdominal Xray: note loops of bowel, flattened diaphragm, and rounded abdomen. The child will be at risk of aspiration on induction of anaesthesia*



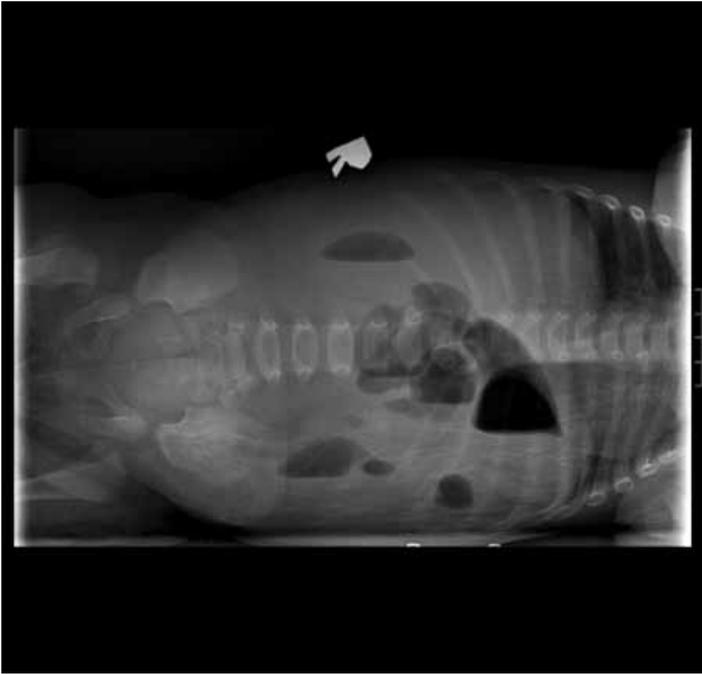


Figure 2. Intussusception. Lateral abdominal Xray: air-fluid levels (intravascular volume status impacted) and no obvious perforation (non-specific)

= 1.1mg.dl⁻¹ (97mcmol.l⁻¹); K⁺= 3.0mmol.l⁻¹; haemoglobin = 8g.dl⁻¹. Weight = 6kg. The surgery team has decided that this patient needs an exploratory laparotomy for bowel obstruction.

In most centres, intussusception is the most common cause of intestinal obstruction in the age group 3 months to 3 years. Delayed presentation is common due to the nature of the disease, partly as the obstruction is intermittent initially, partly due to the lack of appropriate centres for paediatric surgery and anaesthesia. Typically, upper intestinal obstruction causes emesis as the primary symptom and lower intestinal obstruction results in diarrhoea, unless the obstruction is prolonged in which case you will see emesis with diarrhoea. Once the obstruction has become fixed, the pain will get progressively worse and will become constant, which typically causes the care givers to seek urgent surgical care.

Preoperative considerations

Haemodynamic assessment and initial management

This child has low blood pressure, tachycardia, and poor peripheral perfusion and demonstrates the most common form of cardiovascular instability, namely hypovolaemia. The history describes a period of low intake, diarrhoea, bowel obstruction, fever, and possibly sepsis. Each of these factors contributes to an overall picture, that results in low circulating blood volume in a child where the total body water represents 60-70% of body weight. Delayed presentation further amplifies the negative

impact of the pathological state on the loss of intravascular volume. This child is demonstrating clinical signs of over 15% deficit which will require a significant amount of fluid in the form of normal saline. Bloody diarrhoea in this patient may cause anaemia, protein loss, and electrolyte disturbances; a normal haemoglobin in a patient with this clinical picture may be misleading as it may be due to haemoconcentration and severe dehydration with a RBC mass which is actually low. Be aware that significant volume resuscitation may then reveal the true anaemia and low haemoglobin.

This child needs urgent intravenous or intra-osseous access and resuscitation with a normal saline (isotonic) fluid bolus. IV access should not be delayed; the patient is near complete cardiac collapse, and his sympathetic nervous system is working at maximum capacity to maintain mean arterial blood pressure with vasoconstriction and tachycardia. Children at this age can have a significant reduction in the intravascular volume without demonstrating a large drop in blood pressure, but then suddenly have a cardiac arrest due to low intravascular volume as their compensatory mechanisms are overwhelmed. The mother may be able to inform you of the last time the child had any urine output which will help estimate deficit. The normal urine output is 0.5 to 1ml.kg⁻¹.hr⁻¹ but many patients with this clinical picture may be severely dehydrated with present with no urine output and no wet diapers for 24 hours.

This patient should have 20ml.kg⁻¹ IV (IO) 0.9% saline bolus push, repeated until there are clinical signs of improved perfusion and the HR falls; the mental status will also improve as perfusion improves. Don't forget – blood should be considered if 30-50ml.kg⁻¹ volume resuscitation is required, particularly if there is a history of bleeding. The patient may require up to 100ml.kg⁻¹ IV volume resuscitation, ideally a combination of isotonic saline and blood prior to surgery. The fluids need to be warmed to avoid causing hypothermia. Do not give potassium to any volume depleted patient until urine output is established and renal function can be assessed. Place a bladder catheter to help determine fluid status in this situation. The specific gravity (SG) of the urine has been shown to be a rough indicator of intravascular dehydration and sensitivity to the negative inotropic effects of anaesthetic agents. If the SG is less than 1009, i.e. the urine is not very concentrated, then the risk of hypotension with halothane is lower compared to when the SG is greater than 1009. A urine specific gravity can be obtained in most basic laboratories. Note that the patient with decreased level of consciousness will improve after volume resuscitation. The serum glucose level must be measured in any patient with reduced level of consciousness to rule out hypoglycaemia. At times, when the intravascular volume is replaced but due to hypoglycaemia, the

child remains in a less responsive state. When in this clinical situation an intravascular glucose bolus may be required while still attempting to maintain normal serum glucose levels.

Respiratory assessment and initial management

A patient who presents with tachypnoea and/or hypoxia (or low oxygen saturation documented with pulse oximetry) must be assessed carefully prior to the induction of anaesthesia. High respiratory rate may be a secondary compensation to metabolic acidosis due to decreased perfusion. Oxygen delivery to the cells may be inadequate in conditions of poor perfusion for instance due to septic shock, which results in acidosis and excess hydrogen ion production. The body is attempting to remove carbon dioxide to correct the acidosis. Subtle changes in the respiratory system may precede frank respiratory failure if the child becomes exhausted and compensation is overwhelmed. The lungs are not the primary organ of failure but merely required to overwork to balance the metabolic derangement. As with the cardiovascular system, the respiratory reserve is immense but can suddenly fail, resulting in respiratory arrest. Any sudden slowing of the respiratory rate should prompt immediate airway assistance such as tracheal intubation. The treatment of this metabolic process is not bicarbonate but resuscitation fluids (normal saline), titrated as described. When the patient's fluid status improves, the patient will become more alert, the perfusion will improve and the respiratory rate will become more normal. Note that fever will also elevate the respiratory rate and could confuse the clinical picture. Metabolic acidosis should be primarily treated in this scenario with isotonic fluids.

An alternative cause of tachypnoea in this patient may be primary respiratory pathology, such as pneumonia caused by the aspiration of gastric contents. Gastro-oesophageal reflux is common in the child less than 1 year and when a child has a distended abdomen, reflux of gastric contents occurs readily. Gentle insertion of a nasogastric tube should be considered to drain some gastric fluid, although this procedure can prompt emesis and aspiration in the child with a decreased mental status. The nasogastric tube (size 10-12 French) should be softened in warm water prior to placement to help avoid trauma and the patient should be placed in the lateral position to avoid aspiration in case they vomit. Auscultation of chest sounds will help to differentiate between metabolic acidosis (non-pulmonary) and aspiration pneumonia. If the chest sounds are clear the chance of aspiration pneumonia is lowered but if you hear crackles or wheezing particularly on the right side of the chest, this could help diagnose pneumonia. Pneumonia in this setting should not delay surgery in a child with compromised gut perfusion, but will indicate that the child is at greater risk postoperatively.

Electrolytes

A child presenting late with bowel obstruction may have significant electrolyte abnormalities. In patients with altered mental status, the sodium and glucose levels must be measured and corrected. Hypoglycemia can be a result of decreased intake over many days, especially in paediatric patients who are at baseline malnourished and then get sepsis. If the blood glucose level is low, give a bolus of 50% dextrose 2ml.kg⁻¹ and recheck with a glucometer before starting surgery. If the surgery lasts longer than 1 hour, recheck the blood sugar again as undetected hypoglycaemia under anaesthesia can have serious neurological implications.

Hyponatremia (low sodium) is common in the surgical paediatric patient and could be caused by diarrhoea, vomiting, low salt oral fluids, and use of incorrect fluids such as 4% dextrose 0.18% saline for resuscitation at a medical care facility. Paediatric patients who have been resuscitated using 5% dextrose (without added sodium) due to concerns about blood glucose levels can present with an altered mental status and even seizure activity due to iatrogenic hyponatremia. Hyponatraemic seizures are very difficult to manage with anti-seizure medications such as diazepam, and even phenobarbital, if the sodium level is below 120mmol.l⁻¹. These patients should be resuscitated with 0.9% saline; their seizure activity and alertness will improve as the plasma sodium rises. The anaesthesia care provider should not delay emergency surgery until the plasma sodium level is normal if the intravascular fluid status has been corrected. A documented rise in plasma sodium towards normal is reassuring, and shows that resuscitation measures are having a positive impact.

When can you start the surgery?

The case described is a surgical emergency, particularly if there is vascular compromise to the intestines. Bowel ischaemia, necrosis then perforation (with ensuing sepsis) increases the morbidity and mortality of surgery dramatically. If the patient continues to demonstrate metabolic acidosis after resuscitation with isotonic fluids, removal of a necrotic section of intestine may be the only intervention that improves the clinical condition of this child. With the history of delayed presentation, the child does not need to go to theatre in a few minutes, rather in a period of less than 2 hours after IV fluid resuscitation has commenced. The heart rate, respiratory rate and peripheral perfusion should begin to normalize prior to the induction of anaesthesia. Establishing some urine output is a useful sign and indicates improved renal preload following resuscitation efforts.

The following laboratory measurements need to be obtained if possible, but do not delay surgery for the results to normalize:

- Full blood count

- Type and cross match for one unit of blood (typically 400ml of whole blood in a rural hospital).
- Plasma sodium
- Glucose

If you are working in a hospital that cannot measure electrolytes, do not attempt to get a plasma sodium level at another hospital, but rather give volume resuscitation with 0.9% saline as indicated, and proceed with the surgery.

Prepare the theatre with a warmer if the environment is cool, as the child will have extensive exposure and opportunity for heat loss. Prepare all the equipment required to anaesthetise a child. Do not attempt to give the anaesthetic alone but find an assistant. Explain about airway management, the aspiration risk, and the need for cricoid pressure.

Consider the plans for postoperative care well in advance:

- Staffing level on the ward
- Oxygen
- Suction
- Monitoring
- Location on the ward relative to the nursing station
- Light.

Intraoperative considerations

Induction concerns

This patient has a distended abdomen due to obstruction and is considered to have a “full-stomach” so must have a rapid sequence induction with cricoid pressure. The anaesthetist must have an able assistant. A child with intestinal obstruction



Figure 3. Child with bowel obstruction. The high intraperitoneal pressure will decrease the lung capacity, FRC and oxygen reserve prompting a rapid drop in oxygen saturation during intubation

will need full precautions to prevent aspiration on induction, irrespective of the length of time they have been starved, or the fact they have a nasogastric tube (NGT). Cricoid pressure needs to be applied so that it does not distort the airway, as this will make the intubation more difficult. Ask the assistant to direct the trachea backwards, upwards and gently to the right. Preoxygenation prior to intubation is extremely important in children with intestinal obstruction as the functional residual capacity (FRC) is reduced and the child will desaturate rapidly when apnoeic. However, preoxygenation may be difficult if the child is crying, but you can achieve some form of preoxygenation if you waft high flow oxygen through a mask directed towards the child's face. If the patient is obtunded and less responsive, a good mask fit may be achieved, which will allow preoxygenation with 100% oxygen, thus reducing the risk of desaturation during intubation. As you can see from figure 3, if the abdomen is very distended it has the potential to restrict diaphragmatic movement and lung volumes, particularly in the supine position. Both of these factors will cause a rapid drop in the oxygen saturation once the patient stops breathing spontaneously.



Figure 4. Intubation hand positioning: notice that the trachea is being displaced posterior and midline

If the patient has an NGT to help drain the stomach, it should be suctioned prior to the induction of anaesthesia. Some suggest it should be removed immediately prior to induction to ensure a good seal with the facemask if you need to ventilate the patient with cricoid pressure, should more than one attempt at intubation be required. Check the position of the endotracheal tube by auscultation prior to removal of cricoid pressure. Early removal of the cricoid pressure can result in aspiration if the endotracheal tube is placed in the oesophagus. A cuffed tube of an appropriate size can be considered in children of all ages, but the cuff pressure should be measured and the cuff should not be overinflated. Uncuffed tubes are still routinely used in many institutions. Clinical evaluation of the

position of the tube in the trachea is essential after intubation, ideally confirmed by capnography, if available.

The choice of induction agent includes thiopentone (2-4mg.kg⁻¹), propofol (1-2mg.kg⁻¹), or ketamine (1-2mg.kg⁻¹) IV. Ketamine is an ideal agent if the child is critically unwell. An even lower dose of induction agent should be used if the patient is in shock not responding to fluid. If the child has been sick for some time, the blood pressure may drop even with ketamine as the sympathetic nervous system is overwhelmed. Never perform an inhalation induction in these patients. Succinylcholine (2mg.kg⁻¹ IV) should be used as the muscle relaxant. Rocuronium could be considered as an alternative, if available. Once the endotracheal tube position is confirmed, the NGT should be replaced if it has been removed to decompress the stomach during the remainder of the intraoperative and postoperative course.

Maintenance concerns

After induction of anaesthesia and intubation with succinylcholine, monitor the haemodynamic status closely. Induction may reveal hypovolaemia and cause the blood pressure to fall. If this happens, give a fluid bolus of normal saline or blood in 10ml.kg⁻¹ increments. Two peripheral IV lines should be inserted (ideally 22G), with access to an injection port during surgery. Place a three-way stop-cock in line so that blood or normal saline can be pushed with a 20-60ml syringe. This helps to deliver fluids accurately, also rapidly if required. It is important that fluid balance is documented as accurately as possible; pulmonary oedema can be a complication of excessive fluid administration in the operative setting. Blood should be given based upon blood loss, with the goal of improving oxygen delivery dictated by cardiac output, oxygen saturation, and haemoglobin level. Studies have shown that in "stable sepsis" in the paediatric population that a haemoglobin level above 7g.dl⁻¹ should be safe.⁵ In a rural hospital setting, this figure may need to be higher due to the weak medical infrastructure and support systems.

Inotropes will need to be started if blood pressure remains low despite fluid administration. Opening the abdominal fascia may reduce venous return if the abdomen is tense, for instance where there is bowel ischaemia and necrosis, which will cause a fall in the cardiac output (BP). In addition, movement of bacteria from the obstructed, and possibly necrotic intestines to the blood stream may release mediators and hydrogen ions (producing acidosis), resulting in more cardiovascular instability during surgical manipulation and repair of the damaged intestines. A bolus of adrenaline 1 mcg.kg⁻¹ IV may be useful whilst an infusion of adrenaline is prepared (dilute 1 mg adrenaline in 1000ml saline to give a solution of 1mcg.ml⁻¹ adrenaline).

To prepare a solution of dopamine for use without a syringe infusion pump, place 200mg dopamine into 100ml of normal saline in a paediatric microdrop giving set. Titrate this infusion to maintain the blood pressure in the normal range. The infusion may be required for a few days in severe cases. Monitor urine output as an indirect measurements to assess adequate organ perfusion and keep the patient warm in the perioperative period with the means which you have available to you in your hospital setting.

The use of inhalation agents, ketamine, opioids or any combination will be determined by the patient's response to induction and surgery. Inhalation agents will all cause some degree of vasodilation and cardiac depression, which may not be tolerated in the seriously ill patient, thus prompting the use of a ketamine infusion. You will need to control the ventilation, either by manual ventilation or using a ventilator with settings for a paediatric patient. If the patient is acidotic (determined clinically or by measurement of the venous or arterial blood gas), they will not tolerate spontaneous ventilation with low tidal volumes, particularly if the abdomen is compressed by the surgical team during laparotomy. Consider using a non-depolarising muscle relaxant to assist the surgeon and expedite the procedure, but the child must be fully reversed at the end of the operation. Beware sudden cardiac arrest in theatre due to hypovolaemia, myocardial depression, or associated with CO₂ in the absence of end-tidal CO₂ monitoring. If the monitors stop recording, do not automatically consider monitor malfunction but immediately evaluate the patient for cardiovascular collapse.

At the end of surgery, consider the options for extubation carefully. If a stoma is placed, the surgeon may manually decompress the bowel and the abdominal compartment may not be too tight. Alternatively, if there is a primary anastomosis, abdominal distension may persist postoperatively. In severe cases of obstruction and sepsis, primary anastomosis would not be the primary option due to potential failure and leakage. In either case, the child needs to be fully awake, breathing well and adequately reversed, indicated clinically by flexion of the legs.

Postoperative considerations

The two most important factors for safe postoperative care are the location in the hospital and the nurse: patient ratio. The ideal location should have oxygen, suction, good lighting, be close to the nursing station; the room should be warm, the head of the bed elevated, and there should be, one paediatric nurse assigned to 1-2 children. In many hospitals the nurse: patient ratio is 1:15, with very ill children, and this will not be safe for this child for the 72 hour period when the risk of morbidity and mortality will be greatest. The location of

postoperative care must be considered early so that the child has the best chance of survival.

Regular paracetamol 20-30mg.kg⁻¹ PR should be given every 6-8 hours around the clock for 3 days, with very small doses of opioids titrated as required. Good nursing care is essential. Many of these patients will have an oxygen requirement for a few days while the sepsis and any pneumonia resolves. The respiratory status, respiratory rate, should be monitored carefully, particularly if opioids are given to a child receiving oxygen. A fall in saturation is a late finding and narcotics should only be used in the setting of a 1:2 nurse:patient ratio. Diligent monitoring of the vital signs is essential, and the surgical or ICU team must respond rapidly to concerns raised by the nursing team.

CONCLUSION

Emergency surgery for bowel obstruction in children presents many challenges for the anaesthesia care provider. Delayed presentation and sepsis have a profound physiological impact on many organ systems. Children have a great reserve and ability to heal but may also hide the seriousness of their illness, and have the potential for sudden decompensation. Good outcomes rely on meticulous perioperative planning, proper training, equipment, and basic supplies. A team approach

involving the nurses, laboratory technicians, paediatricians and surgeons is essential. The anaesthesia care provider faced with this challenge needs to be cautious, ask for assistance, and be extremely diligent in monitoring the patient during the entire perioperative course.

REFERENCES

1. Ekenze SO, Anyanwu PA, Ezomike UO. Profile of pediatric abdominal surgical emergencies in a developing countries. *Int Surg*. 2010; Oct-Dec; **95**(4): 319-24.
2. Goldstein B, Giroir B, Randolph A. International pediatric sepsis consensus conference: Definition for sepsis and organ dysfunction in pediatrics. *PediatrCrit Care Med* 2005; 6 No. :pp 2-8.
3. Aird, WC. The hematologic system as a marker of organ dysfunction in sepsis. *Mayo Clin Proc* 2003; **29**: 605-606, vii.
4. Levi, M. The coagulant response in sepsis. *Clin Chest Med* 2008; **29**: 627-642, viii.
5. Karam, O, et al. Red blood cell transfusion thresholds in pediatric patients with sepsis. *PediatrCrit Care Med* 2011; 12, No. 5. (free online link via <http://www.researchgate.net/>)

Head injury in paediatrics

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INTRODUCTION

Trauma is one of the leading causes of death and disability in children. The aetiology of injury varies with age and children with serious head trauma often have multiple injuries. The most common mechanism for head injuries globally is from motor vehicle collisions, and in the developing world the incidence is increasing dramatically. The presentation of head injury varies with the severity of the insult ranging from an altered level of consciousness to deep coma. Early identification and proper management of these patients greatly affects the outcome. Survivors of severe traumatic brain injury in childhood unfortunately are frequently left with significant behavioural, cognitive, emotional and physical challenges.

PATHOPHYSIOLOGY

Anatomy

Children have a disproportionately larger and heavier head and relatively weak neck

muscles, this makes them vulnerable to head injury following trauma. The open fontanelles and sutures also predispose infants to a higher incidence of subdural haematoma. Primarily for social reasons the causation varies with age; toddlers frequently suffer head injuries from falls, while older children suffer head injuries from road traffic collisions and sports related injuries. Non-accidental injury must always be considered, particularly in infants.

Cerebral blood flow

Normally, cerebral blood flow is maintained at a constant level to meet the metabolic demands of the brain over a wide range of blood pressure by the process of autoregulation. The autoregulation range is not known in infants and children, but is likely to be around 40-90mmHg.

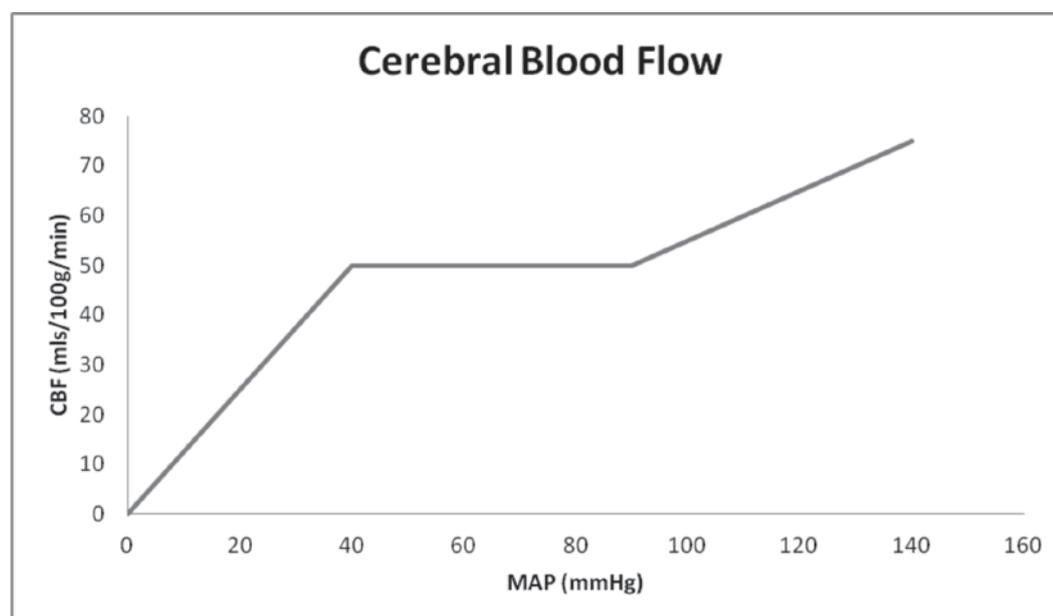
Cerebral autoregulation is impaired by acute brain injury; in this situation, cerebral blood flow follows cerebral perfusion pressure passively.

SUMMARY

Trauma is one of the leading causes of death and disability in children. Children with serious head trauma often have multiple injuries. Early identification and proper management of these patients greatly affects the outcome.

This article covers pathophysiology of head injury in children; assessment and immediate management; anaesthesia for neurosurgical management; and principles of postoperative care in PICU.

Figure 1. Cerebral blood flow



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As cerebral perfusion pressure is an easier figure to derive clinically, it is this parameter that is usually used as a treatment target.

Cerebral perfusion pressure

Cerebral perfusion pressure (CPP) is the blood pressure that perfuses the brain and is defined as the difference between the mean arterial pressure (MAP) and the intra-cranial pressure (ICP). The central venous pressure (CVP) also contributes negatively to the perfusion pressure:

$$\text{CPP} = \text{MAP} - (\text{ICP} + \text{CVP})$$

In head injury patients the intracranial pressure, in the absence of invasive monitoring, is often assumed to be 20mmHg, therefore a MAP 70-90mmHg is targeted to achieve a cerebral perfusion pressure of 50-70mmHg.

Mean arterial pressure (MAP)

The MAP may be calculated from systolic (SBP) and diastolic (DBP) pressures:

$$\text{MAP} = \text{DBP} + (\text{SBP} - \text{DBP})/3$$

Cerebral perfusion pressure less than 50mmHg has been demonstrated to be a predictor of poor outcome in severe traumatic brain injury in children and adults. Extreme hypertension should also be avoided, as it will result in increased cerebral blood flow and cerebral oedema.

The MAP and therefore CPP vary with age, as does the ICP. There is a wide discrepancy between different organisations as to the acceptable blood pressures in paediatric traumatic head injury. In one of the few evidence based papers Haque and Zaritsky suggested in children aged one to eighteen years the MAP target should be calculated as follows:

$$\text{MAP target} = 1.5 \times (\text{age in years}) + 55$$

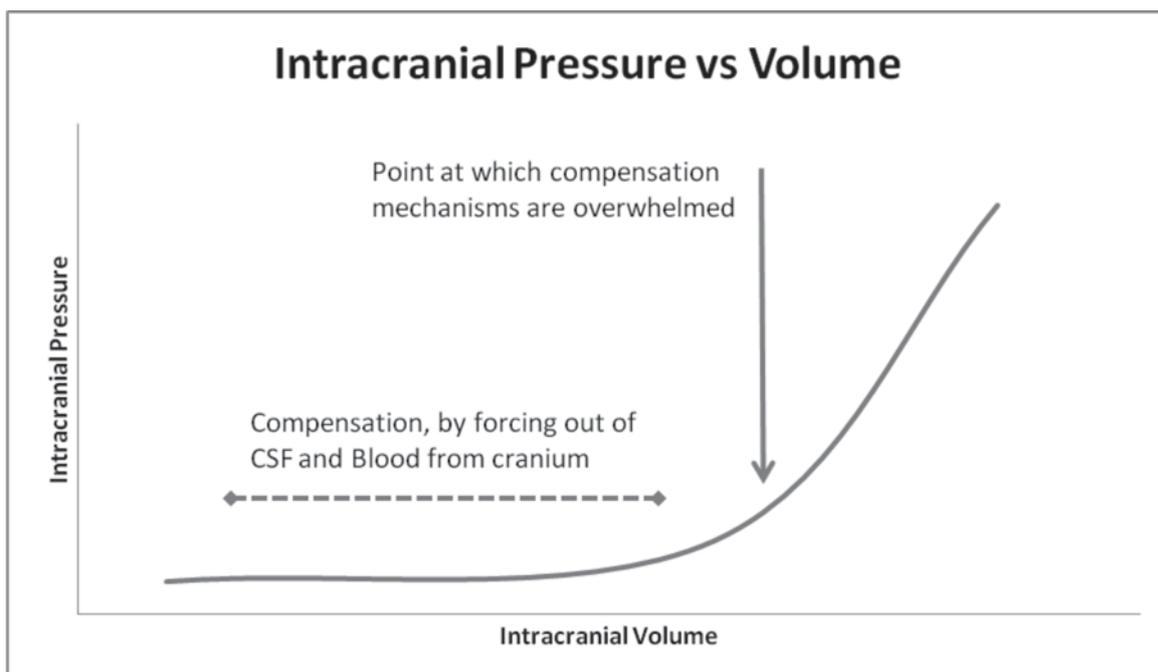
Cautious fluid resuscitation and infusion of vasoconstrictors such as noradrenaline may be required to maintain achieve the target MAP in a child with head injury.

Intracranial pressure

The intracranial pressure (ICP) is determined by the volume of the brain tissue, the cerebrospinal fluid (CSF) volume, and the cerebral blood volume. An increase in any of these volumes increases the ICP. For example an increase in brain tissue may result from swelling of the brain parenchyma, which is referred to as brain oedema.

Small increases in ICP can be compensated by an increase in CSF absorption and reduction in intracranial blood volume. Infants can further compensate due to their open fontanelles and suture lines. Sudden acute changes in intracranial pressure are not well tolerated at any age. If compensatory mechanisms are overwhelmed, intracranial pressure will increase rapidly and the brain will herniate through the structures within the skull or the foramen magnum (coning) to cause coma and death.

Figure 2. Intracranial pressure vs volume



Typical intracranial pressures are lower in children compared to adults (2–10mmHg vs. 8–18mmHg respectively). In health the central venous pressure does not impact upon the CPP, however in polytrauma, thoracic injuries (e.g. tension pneumothorax or cardiac tamponade) may increase the CVP to such an extent that ICP is impaired.

An increase in the cerebral blood volume could result from the following causes of intracranial bleeding:

- Epidural haemorrhage usually results from rupture of the meningeal arteries between the dura mater and the skull. It occurs quickly and may lead to death if not identified and quickly treated. Treatment is usually surgical drainage through a burr hole or craniotomy.
- Subdural haemorrhage usually results from rupture of the bridging veins between the dura and the arachnoid mater. It can be self limiting but if large can raise the intracranial pressure and will need to be evacuated.
- Intracerebral haematoma arise when there is bleeding from the blood vessels within the brain tissue. The figure below shows the three types of intracranial haemorrhage as would be seen on CT scans (note CT scans are usually shown as if looking up from the feet). (See Figure 3).
- Traumatic subarachnoid haemorrhage occurs when blood accumulates below the arachnoid mater where cerebral spinal fluid normally resides. This type of bleed is more classically associated with spontaneous rupture of cerebral aneurysms but is also common in head injury where it frequently communicates with intraventricular blood.

ASSESSMENT AND IMMEDIATE MANAGEMENT OF CHILDREN WITH HEAD INJURY

Assess the conscious level and pupil responses at the same time as attending to 'ABCDE' as follows:

- Airway with cervical spine immobilisation
- Breathing and ventilatory control
- Circulation and control of obvious external bleeding
- Disability and neurological status, including pupil responses
- Exposure - secondary survey with top to toe examination to detect associate injuries (consider non-accidental injury).

Airway and cervical spine control and immobilisation

Establish a patent airway with jaw thrust, making sure to keep the cervical spine immobilised. Suspect cervical spine injuries whenever there is high-energy mechanism of injury, reduced level of consciousness, and tenderness or bruising of the cervical spine. Foreign objects in the mouth and pharynx should be scooped out with a finger and secretions gently suctioned. Immediately provide oxygen by mask as soon as the airway is patent.

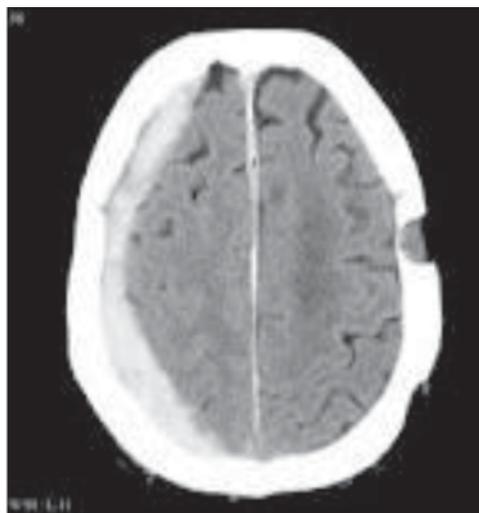
Breathing and ventilatory control

Assess the respiratory rate. Look for chest expansion while assessing symmetry and adequacy. Listen for presence/absence of breath sounds. Palpate for emphysema and broken ribs. Assess the percussion note. Pathologies to look out for include pneumo-haemothoraces, flail chest, surgical emphysema. If

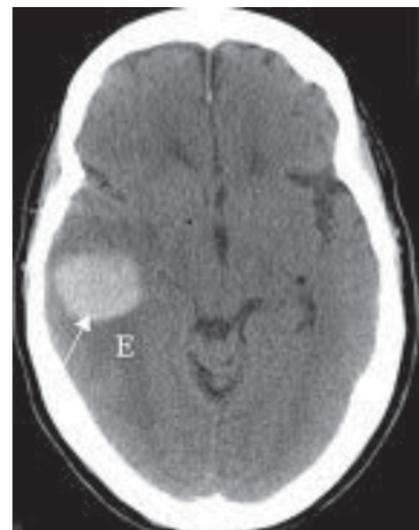
Figure 3. CT scans of different types of intracranial haemorrhage



Left extradural haematoma



Right subdural haematoma



Right intracerebral haematoma

any of these are present they should be managed immediately:

- Give rescue breaths if the child is not breathing
- Decompress a tension pneumothorax by a large bore cannula in the second intercostal space midclavicular line on the affected side
- Chest drain insertion for a haemothorax and also as definitive management for the tension pneumothorax.

Consider advanced airway management in unconscious patients who cannot protect their airway and also in children with hypo- or hyper- ventilation and significant injuries to the head, neck and thorax.

Use arterial blood gas analysis to assess the oxygenation, ventilation and acid base, and electrolyte status.

Circulation and control of obvious external bleeding

Assess the haemodynamic status of the child first by feeling for arterial pulsation. Use the brachial artery in small children and the carotid in older children. Avoid palpating both carotid arteries at once as this may cause cerebral hypoperfusion.

Treat hypotension actively, obtain venous access as an early priority and collect blood samples for full blood count, electrolytes and group and save. Rule out any potential sources of external bleeding - an isolated head injury will not cause hypotension.

As discussed above, a higher than normal blood pressure should be maintained to ensure adequate cerebral perfusion. Use normal saline or Ringers for initial resuscitation. Give an initial bolus of 10ml.kg⁻¹, repeat if hypotension persists. Avoid over hydration and anaemia. Ensure a haemoglobin >7g.dl⁻¹ for adequate oxygen delivery. Look out for active bleeding and prevent any further blood losses.

Disability

Assess the neurological status of the patient quickly using the AVPU scale.

Alert

Responds to Verbal stimuli

Responds to Painful stimuli

Unresponsive

Assess pupil size, equality and responses to light alongside the AVPU as this can give clues that an intracranial haemorrhage is present, and can help guide surgical management.

The pupils should be equal in size and round. Abnormal shapes may indicate cerebral damage; oval shape could indicate intracranial hypertension.

The normal pupil reaction to light is brisk. After removal of the light source, the pupil should return to its original size. There should also be a consensual reaction to the light-source - that is the opposite pupil also constricts when the light source is applied to one eye. Unreactive pupils can be caused by an expanding mass compressing the third cranial nerve. A fixed dilated pupil may be due to herniation of the medial temporal lobe.

It is important to consider factors that may affect the assessment of pupils:

- Any pre-existing irregularity with the pupils, for example cataracts, false eye or previous eye injury
- Any other factors that can cause pupillary dilation, for example medications including atropine and sympathomimetic drugs (adrenaline) and direct trauma (traumatic mydriasis)
- Any pre-existing factors that can cause pupillary constriction, for example medications including narcotics and topical beta-blockers.

The best guide to the severity of head injury is the conscious state and the Glasgow Coma Scale (GCS), which allows the conscious state to be quantified. The score is decided on the patient's best responses. Note that:

- The GCS may be falsely low if one of the following is present: shock, hypoxia, hypothermia, intoxication, post-ictal state or sedative drug administration
- The GCS may be impossible to evaluate accurately if the patient is agitated, uncooperative, dysphasic, intubated or has significant facial or spinal cord injuries
- It should be repeated regularly every 15 minutes, as rapid deterioration may occur
- The GCS is an important tool, but should be used in conjunction with a full neurological assessment to assess the child's neurological state.

The modified paediatric GCS is the standard tool for the assessment of a child's neurological status over time. The trend in the level of the consciousness is more important than a single value. (See Table 1 below). A GCS of 15 indicates no neurological disability and GCS of 3 means a deeply unconscious patient. All children with GCS less than 8 should be intubated and ventilated (see below).

Exposure

Expose the child to allow head to toe and back examination. Do this cautiously to avoid hypothermia, i.e. in a cool environment, keep the child covered when possible. Treat

Table 1: Modified Glasgow Coma Scale for children

Score	Score	Infants	Older children
4	<i>Eye opening</i>	Spontaneously	Spontaneously
3		To verbal	To verbal
2		To pain	To pain
1		No response	No response
5	<i>Speech</i>	Babbles and coos normally	Oriented
4		Spontaneous irritable cries	Confused
3		Cries to pain	Inappropriate words
2		Moans to pain	Incomprehensible sounds
1		No response	No response
6	<i>Best motor response</i>	Normal spontaneous movement	Follows commands
5		Withdraws to touch	Localises pain
4		Withdraws to pain	Withdraws from pain
3		Abnormal flexion to pain	Abnormal flexion to
2		Extension to pain	Extension to pain
1		No response	No response

any injuries immediately they are discovered. Note that scalp lacerations may result in significant blood loss. If the child remains cardiovascularly unstable and requires volume resuscitation, consider other sites of blood loss, for instance, chest, abdomen, pelvis or major limb fracture. Specifically look for:

- Lacerations, bruising and deformity of the face and scalp. Be aware that scalp lacerations may result in significant blood loss
- Signs of base of skull fractures which include:
 - Bleeding or leakage of cerebrospinal fluid from ear or haemotympanum
 - Periorbital bruising (“raccoon eyes”) and bruising around the mastoid (Battle’s sign)
 - Rhinorrhea (CSF leak from the nose).

History

It is important to gain as much information as possible regarding the incident and specifically to determine:

- Time, mechanism and circumstances of the injury; speed of the vehicle; any restraints? (seat belt, car seat)
- Loss or impairment of consciousness with duration (may be inconsistent or unreliable)
- Seizures or fits

- Nausea and vomiting (children may vomit 2 or 3 times, even after a minor head injury)
- Clinical course prior to consultation - stable, deteriorating, improving
- Other injuries sustained.

Even in the heat of a major trauma, always seek a history. Especially in children, it could be that the injury might be not the reason for the coma, but vice versa. A child with cerebral malaria or meningitis may become unconscious and then subsequently fall.

Radiological investigations

Consider the following investigations in patients presenting with traumatic head injury (see Table 2):

Skull Xray

This is useful in areas where CT or MRI scans are not available. Lateral and antero-posterior (AP) views of the skull should be taken. Look for depressed fracture, diastasis and other bony abnormalities. Skull Xray is no better than clinical examination at identifying intracerebral pathology.

Computed Tomography (CT) scan

This is the imaging of choice. It reveals bony pathologies, haematoma (appears hyperdense when compared to brain parenchyma), evidence of cerebral oedema (hypodense

compared to normal brain parenchyma and isodense compared to CSF) and also mass effect (midline shift).

Magnetic resonance imaging (MRI)

MRI can be used in the acute setting to determine the presence and extent of injury and to guide surgical planning and minimally invasive interventions. It is also important in the long-term management of traumatic brain injury to identify chronic sequelae, determine prognosis, and guide rehabilitation. It is however expensive and time consuming.

Cervical spine Xray

A lateral view of the cervical spine is indicated to rule out cervical bony injury or dislocation. However, if CT or MRI is available, do a CT/MRI of the cervical spine at the same time as the head scan as these are more precise and save time.

NEUROSURGICAL MANAGEMENT

Neurosurgical management includes operative removal of epidural or subdural haematomas, or insertion of an extraventricular drain (EVD) as soon as possible after the diagnosis is made. An intra-cranial pressure monitor can be placed for ICP and CPP monitoring and further management in the paediatric intensive care unit.

When there is intractable raised ICP, a decompressive craniectomy to remove part of the frontal lobes may be indicated.

Anaesthesia for evacuation of intracranial haematoma

A small percentage of children with head injury may require surgery to evacuate intracranial haematoma. Bear in mind that:

- Volatile anaesthetic agents reduce cerebral metabolic rate but increase cerebral blood flow and ICP. Halothane increases ICP more than isoflurane and should be avoided if possible.
- Intravenous anaesthetic agents reduce the cerebral metabolic rate, and also reduce cerebral blood flow and ICP.
- Use adequate doses of opioids to obtund the reflex cardiovascular responses to intubation.
- Suxamethonium is indicated for rapid sequence induction in head injured patients even though it causes transient increase in ICP.
- Although ketamine has beneficial effects on blood pressure and is an analgesic, it raises ICP when used as a single agent in induction doses so is usually avoided.

Table 2. Indications for CT or MRI scanning in children

Age over 1 year: Glasgow coma score <14 on assessment on initial assessment
Age under 1 year: Glasgow coma score paediatric <15 on assessment on initial management
Age under 1 year and presence of bruise, swelling, or laceration (>5 cm) on the head
Dangerous mechanism of injury
Clinical suspicion of non-accidental injury
Loss of consciousness lasting more than five minutes (witnessed)
Post-traumatic seizure but no history of epilepsy
Abnormal drowsiness
Confusion or aggression
Signs of a basal or depressed skull fracture
Worsening level of consciousness
Failure of the mental status to improve over time
Focal neurological findings
Penetrating skull injuries
Amnesia

MANAGEMENT IN THE PAEDIATRIC INTENSIVE CARE UNIT (PICU)

The management is mainly supportive, to contain the primary injury and prevent secondary insult to the brain. It is important to maintain cerebral perfusion pressure by control of ICP and haemodynamic status.

General principles of PICU management

Management of intracranial hypertension and cerebral perfusion pressure (CPP).

Intracranial hypertension is defined as ICP >20mmHg for >5 min. Recommendations for adults include maintaining an ICP of less than 20mmHg and perfusion pressure (CPP) of >60mmHg. These may be applicable for older children. Although no normal data is available through clinical evidence, in infants with severe traumatic brain injury it would be logical to suggest targeting for an ICP of <15mmHg and a CPP of >45 to 50mmHg. Careful use of inotropic agents such as dopamine or noradrenaline may be necessary to maintain these parameters. Induced hypertension with phenylephrine is not recommended since cerebral autoregulation is lost and further increases in cerebral blood flow may exacerbate oedema formation by hydrostatic effect. Some units advocate the insertion of a jugular venous bulb catheter, to allow measurement of the SjO_2 . This allows identification of global mismatch between oxygen delivery to the brain and cerebral metabolic rate.

CSF drainage by ventriculostomy has been shown to be as effective as mannitol therapy in reducing ICP.

Ventilation

As both low oxygen (hypoxia) and carbon dioxide retention (hypercarbia) cause cerebral vasodilatation and increase cerebral blood flow, it is key to maintain these in the normal range (PaO_2 8-12kPa and $PaCO_2$ 4-5kPa). Over-ventilation to hypocarbia causes damaging cerebral vasoconstriction. Hyperventilation is occasionally employed as part of salvage therapy if the ICP is very high. This may be as a bridge to neurosurgical intervention, or more long term if jugular venous oxygenation is monitored.

It is assumed that all children with GCS less than 8 will not be able to protect their airway or maintain ventilation adequately; intubation and mechanical ventilation is required. Keep the head in the midline and at 30 degrees elevated position. Positive end expiratory pressure (PEEP) in the range of 4 to 5 cm of H_2O has minimal impact on ICP, and should be used to prevent atelectasis.

Patients with severe head injuries often develop neurogenic pulmonary oedema. This may require further elevation of the PEEP if adequate oxygenation cannot be achieved.

Sedation and muscle relaxation

Sedation and initially muscle relaxation are recommended for adequate control of ICP. An opioid such as morphine, midazolam and a non-depolarising neuromuscular blocker are good in combination as long as hypotension can be avoided. Ketamine was previously avoided but new data suggest that it produces minimal effects on ICP when used in combination with midazolam, and it may help to maintain the blood pressure. Pentobarbitone, phenobarbitone, or propofol may be used, subject to availability. In severely raised ICP multiple agents may be required, but it is essential to avoid hypotension.

Pain and anxiety increase cerebral metabolic demands and should be treated promptly. Intermittent thiopentone and IV lidocaine are recommended to blunt raised ICP response while suctioning the endotracheal tube. Alternatively instillation of lidocaine into the endotracheal tube may be as effective.

Seizure control

Seizures, both convulsive and non-convulsive, are extremely common after head injury. They increase both cerebral metabolic rate and ICP and must be avoided or treated promptly. Deep sedation should reduce the rate of seizure activity but may not abolish it totally.

Midazolam boluses may be used to control seizures. Propofol or thiopentone may also be used. Treat hypotension associated with the use of these agents with fluid therapy. Start Phenytoin patients with posttraumatic seizures. Use of prophylactic phenytoin in head injured children without seizures is not supported by clinical evidence.

If available, EEG monitoring is extremely useful in identifying non-convulsive seizures, which should be controlled to reduce the cerebral oxygen consumption.

Fluid therapy

Initial resuscitation should be with normal saline. Hypotonic fluids must be avoided in children with head injury as a fall in plasma sodium will exacerbate cerebral oedema. Ringer's lactate is slightly hypotonic and many authors suggest it should not be used. Maintain plasma sodium and plasma osmolality within the high normal range (aim for plasma sodium 150mmol.l^{-1} in severe head injury). Place a central line to guide fluid therapy by central venous pressure monitoring.

Maintain glucose within normal levels. Due to stress of the head injury, serum glucose is commonly high, so avoid glucose containing fluids initially.

Children with traumatic brain injury are susceptible to a variety of abnormalities of plasma sodium for example:

- Syndrome of inappropriate antidiuretic hormone (SIADH). This is characterised by hyponatraemia,

low plasma osmolality, high urinary osmolality, normo- or hypervolaemia. Patients are at risk of developing cerebral oedema. Manage promptly with fluid restriction if asymptomatic or hypertonic saline if symptomatic (1-2 ml.kg⁻¹ 3% saline iv bolus).

- Cerebral salt wasting. This typically presents with hyponatraemia, high urinary osmolality, hypovolaemia and hypotension. Manage with an IV bolus of normal saline.
- Diabetes insipidus. This presents with hypernatraemia, high plasma osmolality, low urinary osmolality, hypovolaemia and hypotension. Diabetes insipidus occurs as a result of failure of blood supply to the posterior pituitary with loss of ADH production in the posterior pituitary. Treat it by administration of DDAVP (Desmopressin). Note that diabetes insipidus is often a late sign in head injury and often heralds brain stem death.

Mannitol

Mannitol reduces ICP by two mechanisms, osmotic and reduction in viscosity. Its osmotic effect is the more effective mechanism. Reduction of viscosity is transient, and depends upon autoregulation being intact.

Mannitol in doses of 0.5 to 1g.kg⁻¹ may be used intravenously at 6 hourly intervals with monitoring of serum osmolality (aim to keep serum osmolality under 320mOsmol.kg⁻¹). Mannitol is contraindicated if any of the following is present:

- Serum osmolality is >330mOsmol.kg⁻¹
- The patient is hypotensive
- The patient is known to be in renal failure.

Rapid boluses of mannitol can transiently increase ICP by causing transient systemic hypertension and should be avoided. Mannitol also has a theoretical risk of enlarging a haematoma by rapid shrinkage of brain and tearing of bridging veins.

Temperature control

Body temperature has an important effect on cerebral blood flow. For every 1°C increase in body temperature, there is a 5% increase in cerebral metabolic rate leading to an increase in cerebral blood flow and intracranial pressure. In children with head injury, avoid pyrexia (>37.6°C), and aim for normothermia or moderate hypothermia. Excessive hypothermia (< 33°C) has been shown to increase mortality. In patients with refractory ICP induced hypothermia may be used as a last resort, however clinical studies have shown this to be of no benefit.

Steroids

There is no clinical evidence supporting the use of steroids in cerebral oedema due to head injury. Reserve steroids for patients with brain tumours.

Nutrition

Early institution of enteral feeds is recommended if there is no associated intra-abdominal injury to major organs such as liver, spleen, or duodenal hematoma.

Positioning

Nurse intubated children at 30 degrees head up to reduce ICP and reduce the risk of ventilator-associated pneumonia (VAP). Regular turning and the use of splints will reduce bedsores and contractures.

Rehabilitation

Tracheostomy may be indicated in patients in prolonged comatose state who cannot protect their airway or who require long-term ventilation.

Once the patient has recovered from acute injury in the PICU, early physiotherapy for prevention of deep vein thrombosis and prevention of contractures may be necessary. Arm, leg, hand and feet splints should be used as indicated.

Table 3. Outcomes after paediatric head injury

Early complications

- Transient cortical blindness
- Seizures
- Cranial Nerve palsy
- Diabetes insipidus
- Syndrome of inappropriate secretion of ADH
- Cortical venous occlusion
- Hemiparesis

Late complications

- Post traumatic epilepsy
 - Post traumatic aneurysm
 - Meningitis
 - Hydrocephalus
 - Memory loss
 - Disability
 - Muscle contractures
-

OUTCOMES OF HEAD INJURY IN CHILDREN

Complications of traumatic head injury can be grouped into early and late categories (see Table 3).

Several outcome scores have been described. The King's Outcome Score for Childhood Head Injury, which was derived from the adult Glasgow Outcome Score is described in Table 4.

ICU mortality in paediatric severe traumatic brain injury is slightly lower than in adult practice. Children younger than 4 years of age have been reported to have poor prognosis similar to adults while better outcomes have been reported in 5 to 15 year age group. Unfortunately in many survivors there are significant neurocognitive, educational and social consequences. The GCS at 24hrs remains the strongest predictor of outcome.

CONCLUSION

The improvement in outcomes from traumatic head injury over the past decade has been achieved by strict attention to physiology, a protocolised approach to treatment and underlying improvements in critical care. In well-resourced countries the focus is on centralised care allowing more invasive monitoring particularly of ICP. Unfortunately in less well-resourced countries the burden of road traffic collisions is increasing, but with the application of applied physiology one would still expect to make an impact on outcomes.

REFERENCES

1. Raghavan K, Waddington R. Management of Paediatric traumatic brain injury. *Anaesthesia Tutorial of the Week* 127 (2009). Available at: http://www.wfsahq.org/components/com_virtual_library/media/f1fa2673a7be4eb43f4f85980290b135-4fa20962568cbe51f494b34724e63b32-127---management-of-paediatric-traumatic-brain-injury.pdf
2. Khilnani P. Management of paediatric head injury. *India J Crit Care Med* 2004; **8**: 85-92.
3. Dykes E. Paediatric trauma. *Br J Anaesth* 1999; **83**: 130-8.
4. Haque I, Zaritsky A. *Pediatr Crit Care Med* 2007; **8**:138 –144.
5. Verive M et al. Paediatric head trauma. Medscape 2012 <http://emedicine.medscape.com/article/907273-overview> (accessed 4th October 2014).
6. Alterman D et al. Considerations in paediatric trauma. Medscape 2011. <http://emedicine.medscape.com/article/435031-overview> (accessed 4th October 2014).
7. Clayton TJ et al. Reduction in mortality from severe head injury following introduction of a protocol for intensive care management *Br. J. Anaesth.* 2004; **93**: 761-67.
8. Crouchman M et al. A practical outcome scale for paediatric head injury. *Arch Dis Child* 2001; **84**: 120–4.

Table 4. The King's Outcome Score for Childhood Head Injury

1	Death
2	Vegetative The child is breathing spontaneously and may have sleep/wake cycles. He may have non-purposeful or reflex movements of limbs or eyes. There is no evidence of ability to communicate verbally or non-verbally or to respond to commands.
3	Severe disability (a) The child is at least intermittently able to move part of the body/eyes to command or make purposeful spontaneous (b) Implies a continuing high level of dependency.
4	Moderate disability (a) The child is mostly independent but needs a degree of supervision/actual help for physical or behavioural problems (b) The child is age-appropriately independent but has residual problems with learning/behaviour or neurological sequelae affecting function.
5	Good recovery (a) This should only be assigned if the head injury has resulted in a new condition that does not interfere with the child's well being and/or functioning.

Major haemorrhage in paediatric surgery

Stephen Bree* and Isabeau Walker

*Correspondence Email: stevebree@gmail.com**SUMMARY**

Major haemorrhage is the commonest cause of cardiac arrest during surgery in children. Problems include:

- underestimate of blood loss
- inadequate venous access
- insufficient help
- electrolyte imbalance (hyperkalaemia, hypocalcaemia)
- coagulopathy
- hypothermia

We consider a practical approach to management of major haemorrhage in a child from both a clinical and logistical point of view.

Effective teamwork and communication is essential.

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INTRODUCTION

Major haemorrhage is the single most common cause of cardiac arrest during surgery in children.¹ Problems arise as blood loss is often underestimated, venous access is inadequate, there is not enough help, or the child develops electrolyte imbalance (hyperkalaemia, hypocalcaemia) or coagulopathy.¹ Hypothermia is a particular challenge in major haemorrhage in children.

This article will consider a practical approach to management of major haemorrhage in a child from both a clinical and logistical point of view. Effective teamwork and communication is an essential part of this process.²

CLINICAL ASPECTS

Major haemorrhage may be seen in children in the following situations:^{1,2,3,4}

- Road traffic accidents
- Civil conflict or war (penetrating injuries, typically gunshot wounds)
- Major surgery (neurosurgery, spinal, cardiac or tumour surgery)
- Associated with an underlying disorder of coagulation.

The magnitude of blood loss is typically underestimated in smaller children. All sources of blood loss must be estimated by direct observation, also from the clinical signs and symptoms displayed by the child.

There are a number of definitions of major haemorrhage; a pragmatic definition is when 1-1.5 times the blood volume needs to be infused acutely, or within a 24-hour period.¹ The estimated blood volume of the child must be calculated.

The blood volume of a child can be estimated using the following formulae:

- Neonate 90 ml.kg⁻¹
- Child 80 ml.kg⁻¹
- Adult 70 ml.kg⁻¹

The weight of the child will be known already in a child undergoing elective surgery; in the case of trauma the weight of the child can be estimated using a Broselow tape, paediatric growth chart (use the 50th centile for age); or using the following formula:

- Weight = (age +4) x 2

It is recommended that each hospital develop a major haemorrhage protocol for adults and for children; as soon as a diagnosis of major haemorrhage has been made, the child should be treated according to the major haemorrhage protocol as outlined below:^{2,4}

Immediate actions for a child presenting to the hospital with a diagnosis of major haemorrhage after trauma:

- Stop the bleeding by applying direct pressure, a tourniquet or by packing the wound.
- Give oxygen.
- Obtain IV access; ideally two large bore IV cannulae, the size depending on the age of the child, or central access (femoral vein or internal jugular). Intra-osseous access is effective and can be used to help establish perfusion but the IO needle is likely to become dislodged, so should only be used as a temporary measure. Do not repeat IO access at the same site, as there is a risk of extravasation. Do not attempt sternal IO access in children.

- Take blood for cross match. Call for group compatible, non-cross matched blood if required.
- Avoid crystalloid infusions if the need for early blood transfusion is obvious.
- Order 1 unit of packed red cells and 1 unit of fresh frozen plasma (FFP) per 20kg weight of the child. A 25kg child will need 2 units of blood and 2 units of FFP.
- 4ml.kg^{-1} of packed cells in a child is equivalent to one unit of blood in an adult, and will raise the haemoglobin by approximately 1g.dl^{-1}
- Volumes of blood products may seem high to those not experienced in major haemorrhage in children, but be reassured that cardiovascular parameters provide an excellent guide to effectiveness of ongoing resuscitation.
- Once the bleeding has been controlled, it is important to aim for a normal blood pressure, treat acidosis and to keep the patient warm. Avoid vasopressors if possible. Be aware of over transfusion once stability has been achieved.

Assess the child

- Baseline observations – heart rate, blood pressure, capillary refill, CVP.
- Blood tests – haemoglobin, platelet count; blood gases, coagulation profile (PT, APTT, fibrinogen), lactate if available.
- Near patient coagulation testing if available (TEG/ROTEM).

On-going management

- Transfuse blood – use blood Group O negative if immediate transfusion is required; or group specific blood if there is time. Full cross match may take up to 45 minutes. O Positive blood may be given to males if there are insufficient stocks of O Negative blood.
- Transfuse 5ml.kg^{-1} packed red cells alternating with 5ml.kg^{-1} FFP, to a total of 15ml.kg^{-1} of each if required. Ongoing transfusion will be required if there is continued bleeding and/or the cardiovascular targets are not achieved.
- Monitor the temperature. The blood and fluids MUST be warmed. The patient must be covered, and actively warmed if possible.
- Monitor the ECG. Watch for hypothermia, hyperkalaemia, hypocalcaemia, hypomagnesaemia.
- Give tranexamic acid as early as possible (but do not give more than 3 hours after the acute injury).
- Consider imaging/definitive management of the bleeding (surgery). Alert the theatre teams.

On-going assessment

- Continuous assessment and reassessment of the clinical signs (HR, BP, capillary refill; blood gasses if possible). Is the blood pressure adequate? If the child is in the emergency department and is talking and you can feel a peripheral pulse, the blood pressure is adequate. It is not necessary to achieve a normal blood pressure at this stage.

Coagulation problems

- Dilutional coagulopathy is likely if packed cells are used without FFP; whole blood contains platelets and coagulation factors, so coagulopathy is not so much of a problem if whole blood is used.
- If blood loss of one blood volume is expected, give FFP early to prevent dilutional coagulopathy, rather than waiting for coagulopathy to occur.
- Continue transfusion as required, alternating 5ml.kg^{-1} packed red cells with 5ml.kg^{-1} FFP, to a total of 15ml.kg^{-1} of each if required. Keep an accurate record of blood products and volume given.
- Consider platelets 5ml.kg^{-1} and cryoprecipitate 5ml.kg^{-1} if there is on-going bleeding or signs of coagulopathy.
- Recheck blood tests – haemoglobin, platelet count; blood gases, coagulation profile (PT, APTT, fibrinogen), lactate if possible.
- Near patient coagulation testing if possible (TEG/ROTEM).
- Coagulation factor concentrates may be required for patients with inherited abnormalities of clotting (haemophilia, von Willibrands disease).

Targets

- Aim to maintain fibrinogen $>1.5\text{g.l}^{-1}$ and platelet count $>75 \times 10^9.\text{l}^{-1}$
- Aim for ionised calcium greater than 1mmol.l^{-1} . If measurement of calcium is difficult, consider giving 10% CaCl 0.2ml.kg^{-1} slow IV bolus for every 20ml.kg^{-1} of blood given.
- Monitor potassium levels. If there are 'tented' T waves on the ECG, or K^+ is greater than 5.5mmol.l^{-1} , consider 10% CaCl 0.2ml.kg^{-1} or insulin/dextrose infusion.
- Aim for haematocrit greater than 0.3 ($\text{Hb} >10\text{g.dl}^{-1}$) once a steady state is achieved.
- PT and APTT are not very sensitive in the context of bleeding (the values may be relatively normal, but the patient is still bleeding). Aim to maintain PT and APTT $<1.5\text{x}$ normal value.

- Repeat coagulations tests, if possible, every hour.
- Repeat this cycle if necessary.

Tranexamic acid

- There is good evidence to support the use of tranexamic acid in trauma. It is also frequently used in major haemorrhage after elective surgery. It should be given within 3 hours of the haemorrhagic insult.
- Give a loading dose of 15mg.kg⁻¹ slow IV bolus (consider an additional infusion of 15 mg.kg⁻¹ over 8 hours).
- Tranexamic acid is excreted in the kidney; it should be used with caution in patients with renal failure.

ORGANISATIONAL ASPECTS

Organisational aspects of management of major haemorrhage are important to consider. Good leadership is essential, with clear allocation of tasks. Make sure the phone numbers of key personnel are readily available in the event of a major haemorrhage being called.

- Identify a team leader to be 'in charge' and to coordinate care. This is usually the senior surgeon or anaesthetist, and ideally someone who can stand back and direct as a non-hands on leader.
- Communicate early with the laboratory so that they understand the gravity of the situation.
- Identify someone to take blood samples to the lab or to collect the blood.
- Make sure all blood is checked properly before it is given; put two name bands on the patient (in case one is taken off); use this to check the blood. It is the responsibility of the person administering the blood to check that the correct blood is being given to the correct patient.
- All blood should be given through a blood giving set with filter; a special filter is not required for platelets, but they should be given through a clean giving set to avoid the platelets sticking to the blood in the giving set.
- A "scribe" should be designated early on to record all interventions and products / drugs given until the tempo has settled down.

CONCLUSION

The management of major blood loss in children can be a daunting prospect in any facility due to the limited physiological reserve of the patients and the technical difficulties of dealing with a small child in shock. The emotional component can also add to the stresses involved but it is essential that simple basic resuscitation principles are applied, as they would be for an adult. Calm strong leadership adds to positive outcomes in these circumstances.

Recent experiences in conflict zones have emphasised the importance of applying the above principles to the most severely injured and shocked children.⁴ Early blood gas results can demonstrate extremely deranged physiology, which corrects rapidly when resuscitation is adequate. Volumes of products used may often seem excessive for those new to these circumstances but careful monitoring of clinical parameters, bedside clotting, Hb, electrolytes and blood gases will support and aid in decision making with respect to ongoing blood product requirements. Acute transfusion of many multiples of the patient's blood volume is not unusual.

The switch from the unstable to stable patient will be achieved once surgeons have gained control of the bleeding and coagulopathy is managed. Until this is achieved, the outcome will be determined by the ability to follow and apply the above guidelines in a methodical way, as well as clear leadership and teamwork.

REFERENCES

1. Bhananker SM, Ramamoorthy C, Geiduschek JM et al. *Anesth Analg* 2007; **105**: 344-50.
2. Association of Anaesthetists of Great Britain and Ireland, Thomas D, Wee M, Clyburn P et al. Blood transfusion and the anaesthetist: management of massive haemorrhage. *Anaesthesia*. 2010; **65**: 1153-61.
3. Gonzalez LP, Braz JRC, Modolo MP et al. Pediatric Perioperative Cardiac Arrest and Mortality: A Study From a Tertiary Teaching Hospital. *Pediatr Crit Care Med* 2014; **15**: (e-publish before print).
4. Bree S, Wood K, Nordmann GR, McNichols J. The Paediatric Transfusion Challenge on Deployed Operations. *J R Army Med Corps* 2010; **156** 4: S361-364.

Paediatric burns and associated injuries

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CASE SCENARIO

A 5-year-old boy is brought to your hospital at midnight from a housefire. He had been rescued from his upstairs bedroom by a neighbour who had subsequently jumped to the ground with him. His rescuer, who suffered a fractured ankle, tells you that when he found him in his smoke-filled room, he was 'deeply asleep' and his sheets were smouldering.

On examination he is sleepy but rouseable, and cries when disturbed. His pyjamas are charred across the chest and left arm. His respiratory rate is 25 breaths per minute, pulse is 130 beats per minute, blood pressure is 75/40, and capillary refill time is 4 seconds. A pulse oximeter reads 99%. You notice soot around his nostrils.

EPIDEMIOLOGY

Cutaneous burns, or thermal injury, can be conveniently divided into scalds and flame burns. Flame burns are frequently associated with flammable liquids spilt onto clothing. Scalds are burns specifically caused by contact with hot liquids. They also often involve clothing, which prolongs the duration of harmful contact between hot liquid and skin. The World Health Organization (WHO) estimates there are 265,000 deaths per year worldwide attributed to burns, with greater than 95% occurring in low and middle-income countries (LMIC). Most fatalities occur in house fires, where death by smoke inhalation is the usual cause.

Burns are the 5th most common cause of non-fatal childhood injury, and the 11th leading cause of death in children aged 1-9, with comparable incidence between males and females. Children are particularly prone to burns due to the inability to recognise danger in the younger age groups and the risk taking behaviours of the older child. As well, children have thinner skin, lose proportionately more fluid, are more prone to hypothermia and mount a greater Systemic Inflammatory Response than adults. Through prevention mechanisms and better treatment modalities, high-income countries have made significant advancements in lowering the rate of

paediatric deaths due to burns. The same is not true for LMIC children, where prevention and treatment advances have been incompletely applied, and where mortality from burns remains more than 7 times higher.

Mortality following a burn occurs as a bimodal distribution. Early deaths occur due to airway obstruction (eg smoke inhalation and associated oedema), carbon monoxide poisoning, refractory shock or co-existing trauma. With good resuscitation techniques, this early mortality can be reduced to 5%. Late mortality often occurs as a result of wound sepsis and multi-organ failure. A coordinated multidisciplinary approach to patient care can also reduce this late mortality.

PATHOPHYSIOLOGY

Two factors determine the severity of a burn – its temperature and the duration of contact with it. Cell death occurs exponentially quickly as temperature rises. Temperatures as low as 40 degrees centigrade can rapidly inflict significant injury in young children.

Scalds caused by water below its boiling point in brief contact (the majority of such injuries) are not surprisingly less severe than scalds caused by liquids e.g. cooking oil at higher temperature, or liquids which are in skin contact for a longer time. Infants or those children physically unable to move themselves away from the burning agent are susceptible to this latter mechanism.

Flame burns have a higher temperature and cause the most severe injuries. Histologically, the burnt skin consists of a central coagulated, necrotic area surrounded by zones of venous stasis and hyperaemia. The capillary leakage that occurs in these outer two areas is the result of both direct heating and secondary inflammation. Burnt epidermis permits large evaporative fluid losses of up to 200ml.m⁻².h⁻¹. These and other fluid losses cause hypoalbuminaemia and the clinical picture of shock. Nevertheless, in the first hour after a burn, the commonest cause of death is smoke inhalation. The burning contents of enclosed rooms contain a lethal mixture of hot, noxious gases, soot particles and depleted oxygen level.

Summary

Despite the temptation to immediately attend to the burn, it is vital to remember the ABCDE approach to a victim of trauma. Severely burnt children may well have other major injuries not obviously apparent.

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EARLY MANAGEMENT OF SIGNIFICANT BURNS

Despite the temptation to immediately attend to the burn, it is vital to remember the ABCDE approach to a victim of trauma. Severely burnt children may well have other major injuries not obviously apparent, either as a result of an associated blast injury or in their efforts to escape the fire e.g. by jumping from a window.

Airway and inhalational injury

The airway, as well as breathing and circulatory systems should be rapidly assessed whilst you administer high-flow oxygen and establishing monitoring. You may need to immobilise the cervical spine with collar, sandbags and tape or manual inline stabilisation (with an assistant). Obtain further history. The patient's airway may be affected either by inhalational injury (which if present increases mortality by up to 50%) or by thermal injury to the face. Whilst the latter is usually obvious, indicators of the former may be more subtle. Environmental factors that can contribute to inhalational injury include fire in an enclosed space or fire associated with explosion. Loss of consciousness is also associated with inhalational injury. The following features on examination also point to an inhalational injury:

- Soot around the nose/mouth/in sputum, burning of eyelash/brow & nasal hairs
- Drooling, dysphonia or dysphagia, stridor.

Approximately 60% of flame facial burns will have an associated smoke inhalational injury. Inhalation of hot dry gases tends to cause supraglottic injury to the lungs, whereas steam inhalation also results in deeper, parenchymal injury. The shockwave from a blast can cause a mixture of chest injuries: barotrauma and contusion to the lung and blunt trauma. Any suspicion that an inhalational injury has occurred should make you consider immediate, elective intubation. Airway oedema can develop very quickly and make later attempts at securing the airway extremely difficult.

There are several considerations about endotracheal intubation that are particular to burns patients:

- Expect to use a smaller endotracheal tube than calculated due to airway oedema
- Do not cut the endotracheal tube, as facial swelling can be dramatic and 'bury' the end of an inappropriately short tube somewhere within the mouth
- The tube should also be inserted sufficiently far that the distal end is not 'drawn' out of the trachea by the effects of proximal mucosal oedema
- A tracheostomy might not be feasible for a prolonged period if the tissues of the neck are injured, but may help prevent long-term complications such as tracheomalacia and subglottic stenosis if done earlier during hospital stay
- It is useful to site a nasogastric tube at the same time to allow gastric decompression and early feeding, which is important in any burn affecting >10% body surface area.

Breathing

Tachypnoea, hypoxaemia, and eventual cyanosis suggest a lung parenchymal thermal injury, which will almost certainly require

ventilatory support. Invasive arterial monitoring, if available, prevents the need for repeated blood sampling and provides important metabolic, respiratory and haemodynamic information. An Acute Respiratory Distress Syndrome frequently supervenes which requires specialist critical care, including protective ventilation strategies, regular bronchoalveolar lavage, and timely tracheostomy insertion if required. Circumferential burns to the chest (or abdomen in infants) can restrict chest wall excursion and require urgent excision – escharotomy. This procedure is not without difficulties; the potential for severe blood loss, hypothermia, and difficult positioning are all significant considerations.

Carbon monoxide poisoning

Fire produces carbon monoxide (CO) when organic chemicals burn in low oxygen environments. Inhaled CO combines with haemoglobin with 200 times the affinity of oxygen

to form carboxyhaemoglobin. Thus, less oxygen is delivered to the tissues. Pulse oximeters cannot distinguish oxyhaemoglobin from carboxyhaemoglobin, giving erroneously high (or falsely 'normal') readings. If possible, carboxyhaemoglobin level should be measured (some blood gas analysers do this) and if between 5-20%, the child should remain on high-flow oxygen, which speeds the dissociation of carboxyhaemoglobin.

Metabolic acidosis and coma are associated with levels >20%. Whilst hyperbaric oxygen therapy is the treatment of choice where available, use intubation and 100% oxygen where it is not to treat carboxyhaemoglobin levels over 20%.

Other poisons e.g. cyanide can sometimes accompany the incomplete combustion of common household plastics. Cyanide poisoning presents as metabolic acidosis, coma and unusually high venous oxygen saturation (cyanide prevents cells from using oxygen).

Circulation

Hypovolaemic shock occurring in the first few hours after a significant burn must be assumed to be due to another injury, which must be sought. Obtain the largest venous access possible, preferably in two unaffected sites. Burnt skin can be cannulated if necessary, although the skin creases over the femoral vessels are often spared. Finding unaffected sites for non-invasive monitoring can be problematic; non-invasive blood pressure measurement may be difficult with extensive limb burns and non-adhesive ECG electrodes can be stapled directly to burnt skin. Urine output measurement, ideally via urinary catheter is essential to monitor haemodynamic status and guide ongoing fluid resuscitation in the shocked burnt victim. Send initial blood samples should be sent for complete blood count, electrolytes, glucose and cross-match. Haematocrit rises initially as a result of plasma loss but anaemia may then follow due to haemolysis, recurring surgical blood loss and sepsis-induced marrow suppression. A significant catabolism develops; early excision and skin cover of the burn can reduce this hypermetabolic state, although protracted protein loss can occur up to a year after burn injury.

Fluid calculation

Shocked children should initially be managed as would any shocked child: warmed IV saline 0.9% boluses of 20ml.kg⁻¹. Non-shocked children with burns of greater than 10% will require IV fluid

resuscitation, which can be calculated according to total body surface area (TBSA) burnt, with the following (Parkland) formula:

$$\% \text{ TBSA} \times \text{weight in kg} \times 4 \text{ ml}$$

TBSA involvement can be calculated in adolescents using the 'rule of nines' (Figure 1A), and can be calculated with Lund-Browder modification in infants and children (Figure 1B). Give half of this volume over the first 8 hours since the burn occurred and the rest over the next 16 hours; this is in addition to the normal daily fluid requirements of the child. The type of fluid used is generally a crystalloid such as compound sodium lactate (Hartmanns) or 0.9% saline. Colloid has not been shown to confer any additional benefit in the early stages of burn management.

There are criticisms of the Parkland formula, especially with the extremes of injury, in which the Parkland formula tends to underestimate fluid resuscitative needs with large

TBSA burns, and overestimate resuscitative needs with small TBSA involvement. Other formulae exist that can assist in guiding fluid resuscitation, which can alternatively be used, especially in these extremes of injury. These formulae are guides only, for you to use in conjunction with regular clinical assessment and measurement

of urine output, which should be maintained at $>2\text{ml.kg}^{-1}.\text{hour}^{-1}$ in children. Of note, significant over-resuscitation with fluids can result in abdominal or extremity compartment syndrome or severe pulmonary insult causing worsening gas exchange.

There has been renewed interest in resuscitation using oral rehydration, and while this may be appropriate for minor burns $<10\text{-}15\%$, you must be cautious in the patient with more extensive burns due to the high incidence of ileus.

(A) Rule of 'nines'

(B) Lund-Browder diagram for estimating extent of burns

Adapted from U.S. Department of Health and Human Services [Public domain] (<http://www.remm.nlm.gov/burns.htm>)

Neurological Assessment

A rapid assessment of conscious state is part of the trauma primary survey. Reduced conscious state could have many causes such as head injury, hypoxaemia, inhalation of toxins, shock or pre-existing disease states (e.g. diabetes).

Exposure and secondary survey

It is important to fully undress the child to estimate the full extent of the burn, but once done ensure the child is covered again as rapid heat loss occurs. Warm the room if necessary. If taking the child to the operating room, the temperature should be raised, aiming for a warm "thermoneutral" environment. This reduces the patient's energy expenditure as well as helping to prevent hypothermia.

Although beyond the remit of this article, it is important to think of significantly burnt children as trauma victims and conduct a thorough head to toe examination once the initial stabilisation has been completed.

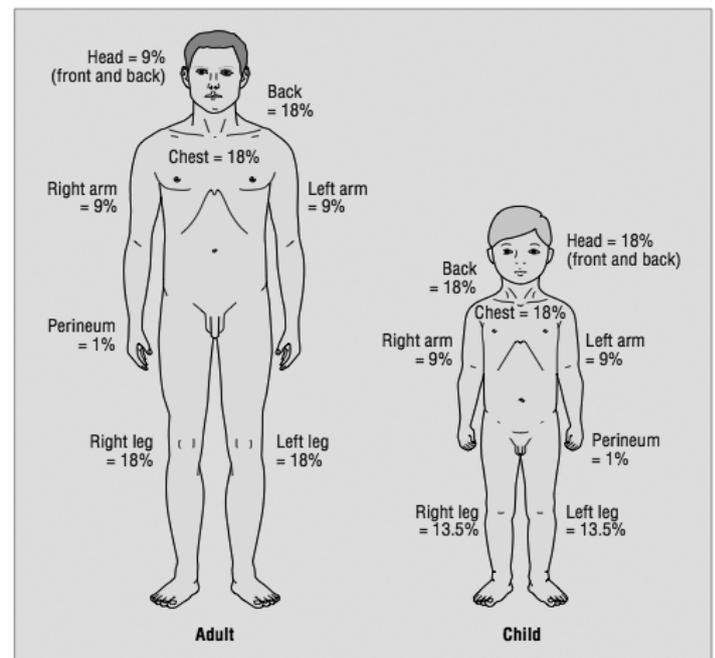
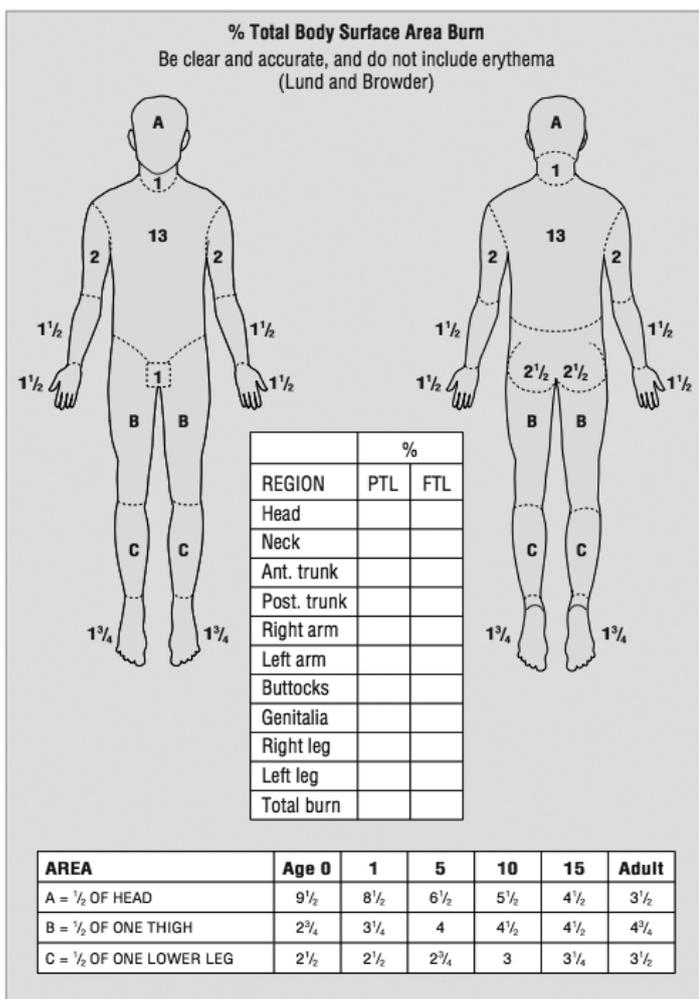


Figure 1: Estimating Percentage Total Body Surface Area in Children Affected by Burns

FURTHER SUPPORTIVE MEASURES

Optimal dietary support is extremely important in view of the hypermetabolic state; a patient with a burn of 40% Body Surface Area can lose up to 25% of body weight in 3 weeks if not optimally supported. Starting enteral feeds as soon as practically possible can help to prevent gastroparesis and meet the high metabolic demand.

Some centres are using partial beta blockade (e.g. propranolol to decrease heart rate by 20%) in children who remain tachycardic after the initial resuscitation phase despite appropriate fluid replacement and adequate analgesia. This further reduces the hypermetabolic state. This has been associated with an increased net protein balance and reduced energy expenditure.

BURN ASSESSMENT

Severity is related to surface area affected and depth. Also, burns to certain areas of the body warrant specialist attention e.g. hands, feet and perineum in order to achieve a good functional outcome.

Area

Burn area is often assessed with the aid of charts (Figure 1). The familiar 'rule of 9' and adult burn charts are not suitable for use in children younger than 14 years because of the variation in the relative size of their heads and limbs with age. A useful rule states that the area of the child's hand (palm and adducted fingers) is approximately equal to 1% of their total body surface area, whatever their age. Alternatively, a Lund-Browder diagram can be used.

Depth

Burns are classified into groups dependent upon depth:

- Superficial - epidermis only; skin appears red, no blisters
- Partial thickness - epidermis/dermis; skin appears pink/mottled, some blisters
- Full thickness - dermis/deeper layers, appears white/charred, painless (although marginal areas may still be painful).

Superficial burns are painful but heal rapidly. Partial thickness burns come in 2 varieties – those in which epidermal 'islands' persist around sweat glands and hair follicles and those more severe burns in which these 'islands' are destroyed. The former are painful and take up to 2 weeks to spontaneously heal. The latter may be less painful (due to destruction of nerves) but take longer to heal and may require surgical excision and grafting in order to do so. Full thickness burns require early excision and grafting to reduce infection and improve morbidity and mortality. In practice, the exact depth of many partial thickness burns are difficult to determine with accuracy on initial presentation, however surgical management often comprises early cleaning and covering in theatre under anaesthesia.

FURTHER MANAGEMENT

Analgesia

Burnt children are likely to experience significant pain – titrate boluses of IV morphine 0.1mg.kg⁻¹ until comfortable, carefully observing their level of sedation. Ketamine, if used for anaesthesia/dressing changes, provides excellent analgesia. Later, oral analgesics are preferred – though beware non-steroidal anti-inflammatory drugs because of the

observed predisposition to gastric stress-ulceration.

Wound care

Leave blisters intact. Cold irrigation and compresses can be soothing but run the risk of excessive heat loss and should only be used for <10 minutes in situations with superficial/partial thickness burns of <10-15%. Cling film if available may be applied loosely to protect and prevent fluid loss. Cover burns with sterile towels and avoid repeated exposure.

Wound infection is a major cause of late fatality following a burn; gram-negative bacterial colonisation occurs early, despite aseptic techniques in cleaning the wound. Suitable dressings should be applied early both to reduce this colonisation and to provide some analgesia. A variety of silver (Ag) based dressings are commonly used due to their antimicrobial gram-negative, gram-positive and anti-fungal activity.

Antibiotics should not be used prophylactically. Take care to diagnose sepsis and wound infection; this can be difficult as frequently the patient's temperature increases due to the raised metabolism. If a patient is unwell, particularly with fever, rash, drowsiness, low serum sodium concentration and lymphopaenia, you must exclude other causes of sepsis (eg line-related infection) and consider the diagnosis of toxic shock syndrome. This is potentially fatal and should be treated with antibiotics and fluids. Consider further dressing change and possibly fresh frozen plasma.

OTHER ANAESTHETIC CONSIDERATIONS

Suxamethonium is safe to use, if indicated, in the first 24 hours after a burn. Following this, concerns about causing acute severe hyperkalemia precludes its use for up to a year. To complicate matters, resistance to non-depolarising muscle relaxants is often seen for up to 3 months.

In the acute situation, ketamine is the intravenous induction agent of choice (2mg.kg⁻¹), particularly in cases of hypovolemia. Clearly, it is very important to try to fluid resuscitate any hypovolemic patient prior to anaesthetizing them, just as in any other emergency situation. If you suspect a significant inhalational injury with impending upper airway obstruction, treat the patient as you would any such case: either a gaseous induction, maintaining spontaneous breathing, or if you have one and are confident of your skills (and the child!), an awake fiberoptic technique (impossible in non-cooperative children). In the extreme and catastrophic case of complete, sudden upper airway obstruction you should be prepared to perform an emergency surgical airway. Having an appropriately skilled (i.e. ENT) surgical colleague scrubbed and standing by with the necessary instruments to hand can be immensely reassuring in such cases.

FINAL POINTS

The most effective strategy for burns management involves prevention. Widespread education, safety procedures and devices such as smoke alarms can help to prevent many thousands of deaths per year.

Finally, it is important to remember that occasionally burns are caused by non-accidental injury. If you suspect this due to pattern of injury, delay in presentation or inconsistencies in the history then you must investigate with experienced colleagues to prevent further future injury to the child.

CASE SCENARIO DISCUSSION

The history has several clues as to the likely type and extent of the injuries. He was found 'deeply asleep', probably unconscious, in an enclosed burning room (his sheets were smouldering). The fact that the rescuer sustained a broken ankle suggests he may also have traumatic injury. The pattern of charring to his pyjamas raises the possibility of circumferential chest burn. His initial vital signs indicate that he is shocked. The reduced level of consciousness in the context of soot around the nostrils strongly suggests an inhalational injury, despite the normal pulse oximeter reading. He will require early definitive airway management.

After giving high inspired oxygen and applying an immobilising hard cervical collar resuscitation proceeds according to the familiar ABCDE approach. Upon removing his pyjamas, he is seen to have an extensive area of pink-blistered skin across his chest and left arm. Unfortunately, no burns chart is available so the extent of the burnt area is estimated using the 'child's palm + adducted fingers = 1%' rule. Using this method the burn, which has partial thickness characteristics, is estimated at 20%.

Using the Parkland formula (and assuming a weight of $[\text{age}+4] \times 2$ i.e. $[5+4] \times 2 = 18\text{kg}$), the fluid bolus required over the ensuing 24 hours is: $20 \times 18 \times 4 = 1440\text{ml}$. 720ml should be given over the first 8 hours since the burn and the rest over the next 16 hours. This is in addition to the normal daily maintenance requirements. Estimated weight enables calculation of drug doses e.g. morphine bolus $0.1\text{mg}\cdot\text{kg}^{-1} = 1.8\text{mg}$.

Endotracheal tube size is estimated in the usual way: $\text{age}/4+4$ i.e. $5/4+4=5$. It is prudent to have smaller tube sizes available than the estimated size in case of airway oedema.

REFERENCES

- Pittaway AJ. Managing Paediatric Burns. *ATOTW* (2007) **78**. Available from: http://www.wfsahq.org/components/com_virtual_library/media/833fed4a0395f87316973c2fc9fdf31cf7f1848f0519358f530a9b575414659b-78-Managing-paediatric-burns.pdf
- Barret JP, Desai MH, Herndon DN. Effects of tracheostomies on infection and airway complications in pediatric burn patients. *Burns* **2000**; **26**: 190-2.
- Black RG, Kinsella J. Anaesthetic management for burns patients. *BJA CEACCP* **2001**; **26**: 177-80.
- Cockcroft S, Way C. Burns and smoke inhalation. *Anaesth Int Care Med* **2002**; **38**: 279-82.
- Fuzaylov, G. and Fidkowski, CW. Anesthetic considerations for major burn injury in pediatric patients. *Paediatric Anaesthesia* **2009**; **19**: 202-11.
- Hilton PJ, Hepp M. Immediate care of the burned patient. *BJA CEACCP* **2001**; **1**: 113-6.
- Fraser JF, Venkatesh BS. Recent Advances in the Management of Burns. In: *Australasian Anaesthesia 2005*; 23-32. Australia and New Zealand College of Anaesthetists.
- Fenlon S, Nene S. Burns in Children. *BJA CEACCP*, May 2007; **7**: 76-80
Instructor Manual: *Advanced Paediatric Life Support*, 4th Ed. 2004
- White MC, Thornton K, Young AER. Early Diagnosis and Treatment of Toxic Shock Syndrome in Paediatric Burns. *Burns* **2005**; **31**: 193-197.

Stabilisation and preparation for transfer in paediatric trauma patients

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INTRODUCTION

The principles of trauma management in paediatric patients are the same as those that apply to adults who have been involved in trauma. These principles are those taught in courses such as Advanced Trauma Life Support (ATLS), European Trauma Course and Primary Trauma Care.

This article will focus on recent developments in trauma management and in the stabilisation of the traumatised patient. Many of these come from experience gained from the military. It is important to note that many of these developments have occurred in relation to patients with penetrating trauma and the lessons may not apply in the same way to blunt trauma.

STABILISATION

Patients who have suffered significant trauma are likely to require stabilisation as part of their treatment. This may either be as part of their definitive treatment or, in smaller units, prior to their transfer to a larger trauma centre or hospital with specialist facilities. Patients should be as stable as possible prior to transfer in order to reduce the risks of a complication occurring during transit.

In order to stabilise the patient it is important to consider what is happening at a physiological level.

Massive haemorrhage caused by trauma is associated with poor tissue perfusion, which leads to multi-organ dysfunction and death. Stabilisation of a trauma patient will involve restoration of normal physiology as soon as possible in order to maintain adequate organ perfusion and so minimize damage and improve outcome.

Coagulopathy, acidosis and hypothermia are termed 'the lethal triad of trauma' (Figure 1). This term is used to describe factors in major trauma that, if present, are associated with increased risk of death.

Coagulopathy

Approximately one third of trauma patients have a coagulopathy on admission to hospital, and these patients have a worse outcome than patients without a coagulopathy.¹ Therefore treatment of major trauma should involve identification of any coagulopathy.

Summary

Stabilisation of paediatric trauma patients and preparation for transfer of these patients, should follow the same principles as when dealing with adult patients in similar circumstances. Attempting to return physiology to normal as soon as possible and transferring in a safe and timely fashion to a hospital with the appropriate facilities to provide the best possible care, will allow for the best outcomes after paediatric trauma.

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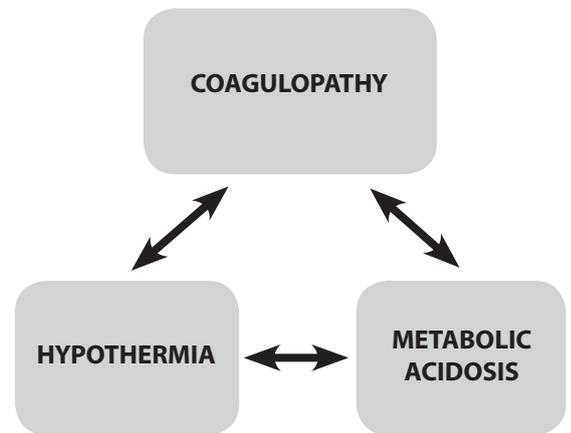


Figure 1. The lethal triad of trauma

Management of coagulopathy, as part of management of massive transfusion, will be discussed in a separate article in this journal.

Acidosis

Acidosis is associated with poor tissue perfusion and will be associated with a low pH, high base deficit (negative base excess) and raised lactate on blood gases. If blood gases are not easily available, then other factors associated with poor tissue perfusion should be considered. These clinical markers of poor tissue perfusion include factors such as prolonged central capillary refill time, tachycardia and reduced urine output. Treatment of acidosis in these circumstances involves adequate fluid resuscitation. Where injuries are life-threatening or where fluid requirements are expected to be more than 20ml.kg⁻¹ it is best to start fluid replacement using blood products wherever available. Restoration of circulating volume to provide adequate tissue perfusion and thus allow resolution of the acidosis is the aim here, so if blood is not available, use crystalloids instead. Where hypotension is present due to loss of blood volume from haemorrhage, it should be treated by replacing circulating volume in the first instance, not with inotropes or vasoconstrictors, which will only serve to worsen tissue acidosis. Once the patient has adequate tissue perfusion and has become haemodynamically stable the acidosis should begin to resolve.

Hypothermia

Hypothermia is likely to be seen in trauma patients, even in warm environments, and has negative effects on many bodily functions including, for example, causing coagulopathy. It should be actively avoided in major trauma patients and simple treatment solutions adopted. Minimise the exposure of the patient where possible using blankets and covers in the pre-hospital environment and continue this in the emergency department. Use of active warmers, such as forced warm air devices will reduce hypothermia further and can be used to actively warm children if they have become cold despite best efforts. If the operating theatre has temperature control, this should be raised to ensure a warm environment for surgery and if fluid warmers are available these should be used as well. Monitoring of the patient's temperature is important because once children start to warm up it can be easy to overshoot and for the patient to become hyperthermic. This is particularly important in head trauma where hyperthermia worsens secondary brain injury, so every attempt should be made to maintain normothermia. Regular temperature checks should form part of the standard management of these patients.

All parts of the lethal triad should be considered and appropriately managed when treating trauma patients in order to reduce trauma related deaths.

Recent developments in trauma management have considered how to correct the physiological disturbance associated with trauma as quickly as possible, in order to try to reduce multi-organ dysfunction. Whilst many of these involve use of new and technical solutions, which may not be easily available worldwide, the principles behind them can be applied in many situations.

Catastrophic haemorrhage control

Any patient who presents with major trauma has the potential to be unstable, particularly from a haemodynamic perspective. In recent years, it has been recognised that in situations of overwhelming haemorrhage, in order to save life, quick interventions to stop further blood loss need to be undertaken before airway measures. This has led to the development of a new paradigm, changing Airway, Breathing, Circulation (ABC) to <C>ABC with <C> being catastrophic haemorrhage control.²

Catastrophic haemorrhage control involves the use of tourniquets to control massive haemorrhage (applied ideally at the point of wounding). Tourniquets are used for any traumatic limb amputation or limb injuries where there is blood loss which would be significant enough to threaten life and which is not controlled by direct pressure. Tourniquets cannot be used in head and neck trauma or to control chest or abdominal bleeding. Other methods of haemorrhage control include direct pressure and the use of novel haemostatic agents such as Quiclot[®], Haemcon[®] and Celox[®]. Quiclot[®] is a kaolin based product which initiates the intrinsic coagulation pathway, whilst Haemcon[®] and Celox[®] are both polysaccharide based haemostatic agents, which are non-exothermic. If none of these novel agents is available, then direct pressure should be used and can be applied in areas where a tourniquet would not work. If possible use a sterile dressing and if bleeding occurs through the dressing then apply a second dressing over the top of the first, rather than removing the first and disrupting

any clot that may be forming. This approach should be used both in the pre-hospital setting and also within the Emergency Department (ED). Controlling catastrophic haemorrhage has resulted in improved numbers of patients surviving who previously may have exsanguinated in the pre-hospital setting.³

When using tourniquets, try to apply the tourniquet as close as is feasible to the proximal edge of the wound in order to preserve as much tissue as possible, in particular saving joints where practicable. Ideally tourniquets should be applied directly onto exposed skin to avoid slipping. If one tourniquet adequately tightened does not control the haemorrhage then a second tourniquet can be applied proximal to the first. It is important to remember that a tourniquet which is tight enough to control haemorrhage is going to be painful and that the patient may try to remove it. Reassuring the patient is very important and in paediatric patients parental presence in this situation may be vital. Always bear in mind that that permanent lethal damage can be caused to the tissues distal to the tourniquet, so tourniquets should only be used where the distal limb is not viable or the haemorrhage is life threatening.

In paediatric patients, control of blood loss is vitally important due to the reduced total blood volume in comparison to that of an adult. Whilst commercially available tourniquets may be too large for the paediatric patient, tourniquets can be improvised or adapted to ensure haemorrhage control. Improvised tourniquets may be created by the use of belts or similar devices with cloth used under the tourniquet to act as padding. The same principles apply when using an improvised tourniquet as when using a commercial device. Experience shows that hospital surgical tourniquets are more effective than field tourniquets, even those that are purpose designed. It is therefore appropriate to change to surgical tourniquets as soon after arrival as is felt safe to do so, which will usually be once large bore venous access or an intra-osseous needle is sited and fluids/blood products are available for resuscitation.

Damage control resuscitation (DCR)

This is a term that has been used to describe a systematic approach to trauma management, combining <C>ABC with attempts to minimise blood loss, maximise tissue oxygenation and optimise outcome.⁴

DCR begins with haemorrhage control at point of wounding and continues with use of advanced resuscitation techniques during casualty evacuation. On arrival in hospital this involves the use of a consultant led trauma team and aggressive management of the lethal triad of trauma.

Early use of diagnostic imaging including chest Xray, pelvic Xray, Focused Abdominal Sonography for Trauma (FAST) and CT allows decisions to be made in a timely manner. DCR may involve an early decision to transfer to theatre for 'damage control surgery' if haemorrhage control is difficult, and surgical haemorrhage control is required.

Diagnostic peritoneal lavage (DPL) could be considered if diagnostic imaging is not available, although this is no longer used in areas where the technique of DCR has been developed. DPL has a significant false positive rate and so will result in some unnecessary laparotomies being performed. It is also invasive and there are risks of organ perforation associated with it. However, a positive DPL in a haemodynamically

unstable child with abdominal trauma would be an indication for laparotomy. If there is life-threatening haemorrhage and clinical examination identifies the abdomen as the obvious source, it is appropriate to proceed straight to laparotomy as part of the damage control resuscitation strategy and not waste time preceding this with DPL.

Fluid resuscitation is carried out using blood and blood products instead of crystalloid or colloid. This, as well as correction of coagulopathy as part of management of paediatric massive transfusion, will be discussed in another article within this journal and so will not be covered here. Early use of intra-osseous access, if venous access is not available, should also be considered.

Right turn resuscitation (RTR)

This phrase refers to the movement of the trauma team from the ED into the operating theatre in order to carry out DCR at the same time as surgical haemorrhage control.⁵ The intent is to reduce the time needed to restore normal physiology by carrying out resuscitation in the operating theatre at the same time as surgical damage control.

This approach to the most critically injured relies upon having a full trauma team on standby and also an immediately available operating theatre. It may not be possible to replicate this in all hospitals, but the principles can still be used. If a patient is determined to be critically injured, and in need of immediate surgery to control the haemorrhage at the same time as resuscitation, then the decision is made to run the resuscitation in the operating theatre. The entire team from the ED move into the theatre, provide immediate airway control and anaesthesia and continue trauma resuscitation as they would have done in the ED at the same time as the surgeon is obtaining haemorrhage control as quickly as possible. This may include clamping large vessels or performing a thoracotomy. Whilst this is ongoing, the ED consultant and ED team will be present and continuing to run the trauma until stability is achieved.

The decision to begin RTR may be made before the patient arrives in the hospital if reports from any pre-hospital team suggest it is necessary. Alternatively an early decision made by the trauma team leader very shortly after the arrival of the patient ED will optimise the usefulness of this strategy for the most seriously injured trauma casualties.

A team working together to reduce blood loss, particularly of non-compressible haemorrhage, use of blood products to resuscitate the patient and early airway control and ventilation will give the patient the best chance of survival. These new developments when used in combination with the standard principles of trauma management will allow the best possible care to be delivered to the severely injured patient.

PREPARATION FOR TRANSFER

Introduction

The need to transfer children who have been involved in trauma between hospitals is well recognised. Children may need to be moved urgently to specialist centres for treatment, to be moved to larger hospitals for definitive treatment or may need to be repatriated for care closer to home once the need for specialist care is finished. This part of the article will look at the safe management of the transfer of children for emergency or urgent care.

Decision to transfer

When considering the need for transfer a number of questions need to be considered in order to ensure that the right patient is moved at the right time to the right place and with the right team to carry out that transfer safely.

These questions can be considered as a list:

Why?

Where?

When?

How?

Who?

Is it safe?

Why?

This question should focus on “Why are we transferring this child?”

Is this move for urgent care – for example a child with an extra-dural haematoma being transferred for urgent neurosurgery, or could this urgent care be provided in the current location? This can also be thought of as a risk/benefit balance: What are the benefits to the child of being moved at this time and what are the risks associated with the transfer.

It may be that the risks for transferring an unstable child are greater than the benefits of movement to a specialist centre and so the child may need to undergo damage control surgery in order to be made stable enough for the transfer.

Where?

This question should focus on “Where are we moving this child to?”

Is the closest specialist centre the most appropriate to look after this child or may it be more appropriate to move the child to a more distant hospital, but one that has the facilities to provide better care? For example, not all neurosurgical facilities can provide paediatric care and so the local neurosurgical unit may not be the best place to transfer the child to, it may be better to move further to definitive care, if the child is stable enough, rather than transfer to the closest unit and then have a second transfer within a short period of time.

Answering this question requires local knowledge and understanding of the facilities and capabilities of the local units and good communication between departments will allow a critically injured child to be moved to the appropriate location for treatment.

When?

This question should focus on “When is the appropriate time to transfer this child?”

Answering this question will depend on the injuries the child has sustained. A child with a traumatic amputation or significant penetrating abdominal injury is likely to require damage control surgery before they are stable enough to transfer to a specialist centre. This is likely to involve haemorrhage control, airway management, ventilation, massive blood transfusion and warming, as they are being stabilised. Alternatively, a child with a head injury who requires

neurosurgery that cannot be carried out in the local hospital, may have to be transferred before they are “stable” because the only way to stabilise them is at the specialist centre.

How?

This question should focus on “How are we going to carry out the transfer?”

The answer to this question will depend on the methods of transport available. The main alternatives are road ambulance, rotary aircraft (helicopter) or fixed wing aircraft (aeroplane). Access to aircraft may be limited both by availability and also by weather conditions. If all the alternatives are available, then timelines for transfer need to be considered. It may actually take longer to task an aircraft, move the patient to the airport/helipad, load the patient, fly between points, move the patient out of the aircraft and transfer to the destination hospital, than it would to carry out a road transfer between the two points. Again, knowledge of the local area and what alternatives are available will dictate the method of transfer between locations. Any vehicles used should be well maintained and appropriately equipped. Ability of transferring staff to access the patient and ability to control temperature and light should be considered.

Who?

This question should focus on “Who is going to carry out this transfer?”

Again the answer to this question will vary depending on the situation and the type of transport to be used. Staff who transfer children should ideally have completed a training course on transfer. Those staff carrying out the transfer should be competent in the management of the paediatric airway, including intubation, and in the management of the acutely unwell child. Ideally a doctor and nurse should carry out a transfer, in addition to the usual crew of the vehicle. Which staff go will depend on individuals’ levels of experience, the need to provide appropriate cover at the hospital and the injuries sustained by the child. Parental presence also needs to be considered at this point.

Is it safe?

This question covers two areas: “Is it safe for the child and is it safe for the team?”

Is it safe for the child? This means, is the child in a safe condition to transfer? Are they stable enough, or do they require further treatment prior to the journey? Is the airway and are all lines secured, or should further access be obtained first? Is there anything that can be done to make this child safer for transfer? Intubation during transfer can be difficult and even dangerous. Consider if it is safer to secure the airway with an endotracheal tube prior to transfer than to attempt this during the transfer.

Is it safe for the team? This means, are there external factors, such as poor weather, that make carrying out the transfer a risk to the medical staff, and therefore the patient? If this is the case, the risk of carrying out the transfer and the potential loss of medical staff, needs to be balanced with the need to carry out the transfer.

Preparation for transfer

Once the decision to carry out the transfer has been made, then preparation for transfer needs to begin. Preparing to transfer a child

requires communication, preparation of equipment and preparation of documentation.

Communication

Communication is one of the most important parts of carrying out a transfer. Communication allows all those involved in the child’s care to work together to ensure the best outcome for the child. This will involve communication by a lot of people, both external and internal to the team carrying out the transfer.

Communication with the receiving hospital

Before a transfer can be made, the receiving hospital needs to have accepted the patient. Often the team taking over the care will have been contacted, but this may not mean that a bed has been arranged or that there is space in the intensive care unit. All of this should be verified before starting the transfer and the patient’s destination within the hospital should be confirmed. Communication with the receiving unit should also include patient information such as whether they are intubated and ventilated, what lines are in situ and what drugs are running. This allows the receiving unit to prepare in advance of the patient’s arrival.

Communication within the hospital transfer team

Once the decision has been made about who is going on the transfer then the members of the transfer team need to discuss the plans for the transfer. This will include discussion about what equipment and drugs need to be prepared and drawn up and what else may be required. “Actions on” plans may also need to be discussed. This means planning in advance what roles people will carry out in an emergency scenario. This may include planning who will take what role in the event of a cardiac arrest, an accidental extubation or need for intubation. Discussion about what to do in the event of oxygen failure or power failure should also be considered.

Communication with the vehicle team

Whatever type of transport is used, the team on the vehicle will all need to know the destination of the patient, any concerns about what may occur during the transport and how urgent the transfer is. Further details required will depend on the type of transport being used. Good communication will enable the transfer to go as smoothly as possible and will help to minimise the risk to the patient.

Communication with the patient and their relatives

The patient and their relatives will also need to know about the transfer. This will include the reasons for transfer, the destination and arrangements for transfer. If a parent is to travel with the child this will need to be arranged, or if the parents are to travel independently, then they will require information about how to get to the receiving hospital. Contact details for the parents should also be obtained and given to the receiving hospital and contact details of the receiving hospital should be given to the parents.

Equipment

The exact equipment needed on a transfer will vary from location to location. Many units have transfer bags pre-prepared and stocked with a full range of equipment to carry out a transfer. Often there

will be different bags for neonatal or infant transfer and transfer of an older child.

This reduces the need to carry all the possible sizes of equipment on every transfer and may also mean taking a different ventilator dependent on the child's age. If these bags are used it is important to ensure those carrying out the transfer are familiar with the equipment and where it is stored in the bag. Table 1 (following page) lists some essential requirements: please note this is not an exhaustive list and other equipment may be needed depending on the individual situation.

Equipment used for transfer should be durable, lightweight and be able to work on battery supply for prolonged periods of time, and replacement batteries should be available. For this reason gas driven ventilators are often used for transport, provided there is an adequate supply of compressed gases. A self-inflating bag may be preferable, and is essential as back-up. The equipment must be able to be adequately secured within the chosen transport, either using brackets or onto a special transport bridge. This could be manufacture locally to suit specific needs. Equipment alarms need to be pre-set appropriately to each child and should be visible as well as audible as conditions during transfer are often noisy. Sufficient equipment should be left at the departing hospital to allow treatment of any further emergencies that occur whilst the transfer team are away.

Equipment should be checked prior to transfer to ensure it is fully working and there is nothing missing. Drugs should be checked to ensure they are in date.

If the patient is receiving drug infusions, then these should be made up in advance of the transfer and spare syringes prepared. The time taken for the transfer should always be considered, allowing for significant delays, and consideration should also be given to the possibility of needing to increase the dose during the transfer. It is better to make up too many spare syringes than to try and draw up further spares in the back of a moving vehicle. Emergency drugs should also be drawn up, labelled and capped off so they are easily accessible during the transfer. Drugs that may need to be bolused during transfer should also be prepared, labelled and capped off for easy use. Consider having two syringes of any emergency or bolus drugs in case one is dropped in the vehicle.

Enough fluid should be taken to provide maintenance fluid as well as boluses during the transfer. In the traumatised child this may include taking blood or blood products and this will need to be arranged prior to transfer and suitable storage organised to keep the products at the correct temperature during the transfer.

Any electrical equipment must be fully charged prior to transfer. If the equipment can be plugged in during the transfer then power cables should be taken. If this is not possible then enough spare batteries should be taken for the equipment to last for the longest possible duration of the transfer.

The amount of oxygen required for the transfer should be calculated. This applies to both ventilated and non-ventilated patients. How much is needed will depend on the FiO_2 of the patient, their minute

ventilation, whether the ventilator is driven by compressed oxygen and the availability of oxygen supplies in the vehicle. It is always worth taking double the calculated requirement to allow for delays and equipment problems. A suitable calculation is:

Minute volume x estimated journey time (in minutes) x 2

- Round this up to the nearest cylinder size. The contents of various cylinders are as follows: D cylinder 340L, E cylinder 680L, F cylinder 1360L.

Documentation

Ensuring the correct documentation is taken on the transfer is important. Copies of all notes and all imaging such as Xrays and CT's should be sent with the patient. Blood test results should also be included. If there is specific documentation for the transfer then this should be taken and completed during the transfer. If there is no specific paperwork then a chart such as an anaesthetic chart where observations, drugs and any interventions required can be completed should be used and filled in for the transfer.

Packaging for transfer

Once the decision has been made to transfer the patient will need to be prepared. This will include transferring onto the appropriate transfer trolley, and ensuring all IV access lines, the endotracheal tube and any other lines such as chest drains, urinary catheter and Nasogastric tubes are inserted if required and are secured appropriately. If there is any question about cervical spine injury then the cervical spine will need to be appropriately protected. When packaging the patient for transfer consideration needs to be given to protecting the patient's pressure areas. Once packaged the child's clinical condition should be re-assessed to ensure they remain appropriately stable at point of departure.

Once the team are ready to depart, the receiving hospital should be contacted again to let them know that the patient is on the way. During transfer vital signs should be observed and documented and any interventions carried out recorded, so that the receiving team have a complete picture on handover.

REFERENCES

- 1 Brohi, K., Singh, J., Heron, M. & Coats, T. (2003) Acute traumatic coagulopathy. *The Journal of Trauma Injury, Infection and Critical Care*; **54**: 1127-30.
- 2 Hodgetts T, Mahoney PF, Byers M, Russell MQ. ABC to <C>ABC: Redefining the military trauma paradigm. *Emerg Med J* 2006; **23**: 745-46.
- 3 Kragh JF, Walters TJ, Baer DG, Fox CJ, Wade CE, Salinas J, Holcomb JB. Survival with Emergency Tourniquet Use to Stop Bleeding in Major Limb Trauma. *Annals of Surgery* 2009; **249**: 1-7.
- 4 Hodgetts TJ, Mahoney PF, Kirkman E. Damage Control Resuscitation. *J R Army Med Corps*; **153**(4): 299-300.
- 5 Tai NRM, Russell R. Right Turn Resuscitation: Frequently Asked Questions. *J R Army Med Corps*; **157** (3 Suppl 1): S310-S314.

Table 1. Suggested equipment for transfer of a critically injured child

Airway equipment	<p>Oro-pharyngeal airway Naso-pharyngeal airway Facemask Ambu-bag (self-inflating bag) and Ayre's T-piece Endotracheal tubes Bougie Laryngoscope + blades + batteries Ties / tape to secure endotracheal tube Suction device Yankauer suction catheter Endotracheal suction catheter End tidal CO₂ monitoring</p>
Breathing equipment	<p>Ventilator appropriate to child's weight Oxygen Spare batteries / power adaptors for ventilator</p>
Circulation equipment	<p>IV fluids Pressure bags Syringe drivers + spare batteries Cannulae Intra-osseous device Invasive monitoring equipment Defibrillator</p>
Drugs	<p>Sedatives Muscle relaxants Epinephrine Atropine Inotropes Anti-emetics Mannitol / hypertonic saline</p>
Other	<p>Portable monitor giving SpO₂, ECG, Non-invasive blood pressure. Ideally with facility to monitor CVP and arterial BP. Blankets Thermometer Urinary catheter + bag Pen torch Blood glucose monitor Medical + nursing notes Radiology images Transfer documentation</p>
Personal Equipment	<p>Money Mobile phone + contact numbers Protective clothing and footwear Personal protective equipment - gloves</p>

Paediatric intensive care in resource-limited countries

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SUMMARY

This article provides guidance on setting up a PICU in a low-income country, based on the personal experience of the authors. We also describe clinical management of specific conditions.

The leading causes of death in the under-five age group are pneumonia, diarrhoea, malaria, complications of prematurity and birth asphyxia. Acute febrile illness is the most common cause of hospital admission, and the largest proportion of admissions are due to malaria and/or invasive bacterial disease. Malnutrition contributes to approximately one third of all childhood deaths. The demand for intensive care services for children and the global burden of critical illness is hard to estimate, but 10 to 20% of children who present to primary care are severely ill and need referral for hospital care. Mortality rates of severely ill children admitted to hospital vary between 10% and 30%.

Critical care is defined as care given in hospital to patients with sudden and reversible critical illness. Paediatric critical care is often thought of as a luxury, but the underlying principles of rapid recognition and targeted interventions using a simple ABC approach, appropriate fluid resuscitation, oxygen and antibiotic therapy are not expensive and do not depend on complex equipment. Younger patients with reversible processes represent the majority of critical illness in low income countries; therefore, simple timely intervention can save lives.

This article provides guidance on setting up a PICU in a low-income country, based on the personal experience of the authors from working in Bangladesh, Nepal and Uganda. We will also consider clinical management of children presenting with shock, convulsions, fever, gastroenteritis and malnutrition; the management of children with pneumonia is considered in detail on page 251.

(LIC) as those having a per capita gross national income of US \$1,025 or less. This describes the situation in 36 countries, mainly in sub-Saharan Africa.

The WHO recommends that every hospital performing surgery and anaesthesia should have an ICU, but a survey from Zambia showed that only 7% of hospitals do.¹ Where an ICU does exist, many admissions are for children under five years old (nearly 30% of admissions to an urban ICU in Uganda (RH personal experience)).

The challenges to provision of intensive care for children are similar in many LIC. There are few trained staff, and few trained in care of the critically ill child; in one study, 73% of doctors and 91% of nursing staff displayed insufficient knowledge in at least one of the medical conditions assessed.¹ Nurses provide most of the care, but there are often few nurses on duty overnight. Basic resources to support intensive care may not always be available; the majority of hospitals have access to supplies of water and electricity, but power surges and intermittent water shortages are common. In a study of 21 hospitals in Bangladesh, Dominican Republic, Ethiopia, Indonesia, Philippines, Tanzania and Uganda, supplementary oxygen was present in 87% of city teaching hospitals but only in 47% of district hospitals.¹ Compressed air, pressurized oxygen or basic monitors such as pulse oximeters may not be available, and there are few biomedical engineers to maintain sophisticated equipment. Mechanical ventilation is not a viable option for children in these situations, and alternative approaches to respiratory support must be used.

SETTING UP A PICU

Children requiring intensive monitoring and support include those with a 'medical' diagnosis, such as severe pneumonia, acute dehydration or

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PAEDIATRIC CRITICAL CARE IN LOW-INCOME COUNTRIES

The World Bank defines low-income countries

sepsis, or after traumatic injury or burns, or after major surgery such as laparotomy.

Much can be achieved through structured education and training of existing staff and reorganization of available resources. Substantial financial input is not required and setting up a PICU is thus a realistic goal. Establishing an intensive care unit may also motivate professional development more widely, and help to foster a culture of education, training and clinical improvement in a hospital.

An initial needs assessment is essential to identify and prioritize key issues; these depend on the institution, the patient volume, rates and causes of mortality, and available resources. The WHO has developed a useful tool for assessing the quality of hospital care for children that scores various aspects of hospital care against a standard applicable to LIC.¹

The following are useful questions to consider:

Why do you want to establish a PICU?

- What are your rates of admission of children?
- What are the leading causes of mortality and morbidity amongst children in your hospital?

What physical space do you have available?

- How many bed spaces?
- Do you have the actual beds?
- Are isolation rooms available?

Is the infrastructure reliable?

- Is the water supply reliable? Do you need a water purification system?
- Is the electricity supply reliable? Do you need an upstream voltage stabiliser to protect your equipment?
- Is there a temporary automatic emergency power supply in case of power failure?

What equipment is available?

There are many pieces of equipment to consider, but we think these are the most important:

- Patient cardio-respiratory monitors, including pulse oximeters, one per patient bed space
- Patient charts to record observations
- An Ambu bag (self-inflating bag), adult, child and infant sizes
- Airway equipment, including facemasks, oral airways, tracheal tubes

- A reliable oxygen source. Oxygen concentrators cost up to \$1200, but are reliable, cheap to maintain, and can provide oxygen up to four patients simultaneously using a flowsplitter. Oxygen cylinders are cheap to buy but expensive and cumbersome to maintain. A supply of compressed air and oxygen will be needed for invasive ventilation, but this is by no means an essential part of setting up a PICU
- Further equipment is needed for an advanced PICU such as patient ventilators, a blood gas machine, and IV infusion pumps
- Is this equipment available from a local vendor or agent?

Are essential and emergency drugs available?

- Antibiotics, fluids, resuscitation drugs, inotropic agents, anti-epileptic agents, bronchodilators, anaesthetic agents, analgesics and sedatives should be available in the PICU
- Drugs should be stored in a locked drug cupboard, but must be easily accessible when needed
- Access to blood and blood products

Is there access to basic laboratory studies and diagnostics?

- A laboratory that can run a basic chemistry panel, a complete blood count, cultures, and check for malaria should be available
- Diagnostics capabilities such as X-ray should also be available within the institution. Although a CT-scan is not essential, it will be of benefit in a location with multiple trauma patients

Personnel

- Who will be responsible for the management of children in the PICU?
- How many nurses do you have available to work on the PICU?
- Are they local staff? Are they trained? Do you have links with any other institutions to assist with training?
- Are the staff dedicated to the PICU, or are they required to cross cover for another area? A nurse is required to look after the child at the bedside, and another is required as a 'runner'

Management

- Does your hospital management support this initiative?
- Who will be the key people in the hospital/community/country that are going to move this project forward?

Finances

- How much money is available for this PICU to be established?
- Will this be adequate to make changes to the physical space and buy the equipment that is required?
- Is someone assigned to take care of the finances?
- Is there a plan to maintain the PICU once opened, for instance to support on-going costs of staff, maintenance of equipment, drugs and supplies of disposable equipment?

Time-line

- When do you expect to open the PICU?
- Is this realistic?

Hurdles

- What are the main hurdles you and other key personnel see?
- This process is useful to identify other issues that may not have been considered previously

It is possible to start small, with a few 'high dependency' beds identified on the children's ward or in the adult intensive care unit. The ideal is to have specific areas identified for children's critical care, both for neonates and for infants and older children.

GENERAL PRINCIPLES OF PICU MANAGEMENT

The WHO 'Pocket book for hospital care of children'¹ is a valuable resource, which should be available to all clinicians caring for children in these settings. The WHO Emergency Triage Assessment and Treatment (ETAT) course has been developed by the WHO and is planned for widespread implementation in LIC. Staff retention and training and a change in culture are often the key to sustain improvements.

Triage

'Triage' is the process of rapid screening of children on arrival to hospital in order to identify those with emergency signs who require immediate life-saving treatment to avert death, and those with priority signs who need to be treated before those deemed non-urgent. It is an essential part of care for the critically ill child, and is described in detail on page 223. Initial management of the critically ill child is also described there.

In practice, we have found that triage is frequently not present, or is of variable quality. Parents queue with their children at the hospital or clinic, and do not undergo medical evaluation until they reach the front of the line, which may be hours later.

Children who are 'marginal' may decompensate while waiting to be seen. In one study, triage in 14 of 21 hospitals was judged to be 'poor' due to avoidable delays, poor organization of the facilities or inadequate assessment of patients.¹

An estimated 50 to 87% of children who die in hospital do so in the first 24 hours.¹ Effective triage and emergency care can lead to impressive reductions in in-patient mortality, and should be an early focus of training when setting up a paediatric critical care unit.

Diagnostic considerations

Precise diagnosis of the cause of severe illness, in particular, differentiation between severe malaria, sepsis, pneumonia, and meningitis, is often not possible at the time of admission as most sick children present with signs and symptoms related to more than one of these conditions. Striving to make a single diagnosis may not be possible or appropriate, and may lead to incorrect or delayed management.

The initial management of critically ill children should therefore be largely independent of the underlying diagnosis, and should focus on addressing life threatening conditions such as hypoxia, hypovolaemia, hypoglycaemia, and convulsions. The WHO advocates the use of integrated management of childhood illnesses (IMCI) approach whereby children are treated according to clinical symptoms rather than focusing on a specific diagnosis. This approach is illustrated in the following case study and has been described in detail on page 224.

Case study

Adebola is a 16-month-old girl in Nigeria. She weighs 10 kg. Her temperature is 38°C. Her mother says "Adebola has been coughing for 6 days, and she is having trouble breathing." The health worker checks Adebola for general danger signs. The mother says that Adebola is able to drink. She has not been vomiting. She has not had convulsions during this illness. The health worker asks, "Does Adebola seem unusually sleepy?" The mother says, "Yes." The health worker claps his hands. He asks the mother to shake the child. Adebola opens her eyes, but does not look around. The health worker talks to Adebola, but she does not watch his face. She stares blankly and appears not to notice what is going on around her.

The health worker asks the mother to lift Adebola's shirt. He then counts the number of breaths the child takes in a minute. He counts 60 breaths per minute. The health worker sees lower chest wall in-drawing, but does not hear stridor.

The health worker asks, "Does the child have diarrhoea?" The mother says, "No".

Due to presence of danger signs, the child is admitted. Oxygen and anti-pyretics are given. IV antibiotics are given due to presence of a clinical condition requiring antibiotics (pneumonia). It is high season for malaria so IV anti-malarials are also given.

Adebola makes good progress and is discharged 5 days later.

Treatment of hypoxaemia

Hypoxaemia is common in children with pneumonia, and is associated with high mortality.

- Hypoxaemia is defined as peripheral arterial oxygen saturation $<90\%$ in room air at sea level as recorded by pulse oximetry
- The treatment target SpO_2 is $>92\%$

Severity of hypoxaemia is defined as follows

- Mild: SpO_2 85-90%
- Moderate: SpO_2 80-85%
- Severe: SpO_2 $<80\%$

Indications for oxygen therapy:

- SpO_2 $<90\%$

In the absence of pulse oximetry:

- Central cyanosis
- Severe lower chest-wall in-drawing
- Grunting respiration
- Restlessness (due to hypoxaemia)
- Inability to drink or feed
- Respiratory rate >70 breaths.min⁻¹
- Head nodding.

Bubble CPAP

Continuous Positive Airway Pressure (CPAP) is commonly used to provide non-invasive mechanical support for children in ICU, either using conventional mechanical ventilators or increasingly using specifically designed CPAP devices.^{2,3}

CPAP may be given by facemask or nasal cannulae. Advantages of nasal CPAP (nCPAP) are as follows:

- Effective treatment for hypoxaemia
- Reduces the number of children requiring endo-tracheal intubation and mechanical ventilation

- Helps stent airways open and decreases the work of breathing, often with minimal oxygen requirement; particularly effective as children are prone to small airway disease.

Bubble-CPAP is a low cost, but effective method of providing continuous positive airway pressure (CPAP) oxygenation in neonates, infants and children and is widely used in LIC for conditions such as pneumonia and respiratory distress of the newborn. The following are required (see figure 1 and 2):

- Source of gas flow (typically 5–10L.minute⁻¹; start at low flow rates in neonates)
- An air-oxygen blender
- A humidifier
- ‘T-piece’ connector, for instance nasal prongs with inspiratory and expiratory limbs (see Figure 1).

The long expiratory limb of the T-piece breathing tube is inserted into a bottle of water: the level of CPAP delivered is equivalent to the length of the expiratory tubing that remains under water. Modern equipment is now available at a fraction of the cost of mechanical ventilators.

Bubble CPAP has been proposed as an inexpensive method of delivering CPAP in developing countries. Bubble CPAP is used as a ‘step up’ treatment from facemask or nasal prong oxygen.

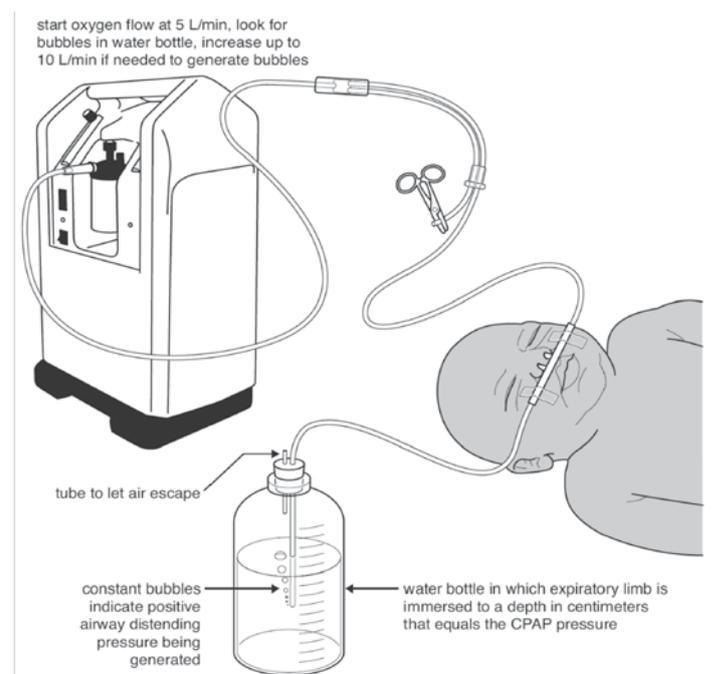


Figure 1. Figure of bubble CPAP. Reproduced, with permission of the publisher, from *The clinical use of oxygen*. Geneva, World Health Organisation, 2011 (Fig 6.1, page 38)²



Figure 2. Bubble CPAP in the ICU of the Dhaka Hospital of ICDDR,B

The advantages of Bubble-CPAP over invasive mechanical ventilation are as follows:

- Low cost
- Easy to use, minimal training required
- Sedation not required to tolerate treatment
- No need for a physician to intubate the patient
- No need for respiratory therapist to operate/maintain a ventilator
- Lower risk of complications e.g. pneumothorax
- Greater hemodynamic stability.

Use of antibiotics in PICU

There is an emerging problem with antibiotic resistance, and 'antimicrobial stewardship' is being encouraged to assure that antibiotics are used appropriately, and for the correct length of time. The basic tenet of antimicrobial stewardship is 'Start smart, then focus':

'Start smart' is:

- Do not start antibiotics in the absence of clinical evidence of bacterial infection
- If there is evidence/suspicion of bacterial infection, use local guidelines to initiate prompt effective antibiotic treatment
- Document on a drug chart and in the medical notes: clinical indication, duration or review date, route and dose of antibiotics

- Obtain cultures first wherever possible.

'Then Focus' is:

- Review the clinical diagnosis and the continuing need for antibiotics by 48 hours and make a clear plan of action - the "Antimicrobial Prescribing Decision"
- The four Antimicrobial Prescribing Decision options are:
 1. Stop
 2. Switch IV to Oral
 3. Change
 4. Continue and Outpatient Parenteral Antibiotic Therapy (OPAT)
- It is essential that the review and subsequent decision is clearly documented in the medical notes.

MANAGEMENT OF SPECIFIC CONDITIONS

Management of children presenting with shock, convulsions, fever, gastroenteritis and malnutrition will be considered below. Detailed management of children presenting with respiratory infections is considered on page 251.

Child presenting with shock

The WHO definition of shock is as follows¹:

- Cold peripheries
- Capillary refill time greater than 3 seconds
- Weak pulse with tachycardia.
- Narrow pulse pressure.

Blood pressure is not commonly recorded in low resource settings and a fall in blood pressure is a very late sign. Hypotension is associated with poor outcomes (See Table 1).

Types of shock:

- Haemorrhagic shock
- Cardiac shock
- Septic shock
- Hypovolemic shock
- Specific infectious outbreak: e.g. Dengue shock syndrome
- A combination of several of the above.

A careful history, examination and investigations should be performed (see Table 2).

Table 1. Cardiorespiratory parameters of shock in children

	<2 months	2- 12 months	1-5 years	>5 years
Blood pressure (mmHg)	<50	<50	<60	<70
Heart rate (beats.min⁻¹)	>180	>180	>160	>140
Respiratory rate (breaths.min⁻¹)	60	50	40	30

Septic shock in children in LIC

Septic shock is common in LIC, and treatment guidelines formulated for high-income countries should be adopted with care.

Pathophysiology

Septic shock occurs when a severe infection in any anatomic location within the body leads to Systemic Inflammatory Response Syndrome (SIRS). When uncontrolled, this leads to hypotension, multi-system organ dysfunction, failure, and eventual death. SIRS triggers both pro- and anti-inflammatory mediators; the vascular endothelium is the primary culprit in being both a source and a target of injury to multiple organs. The three basic changes that occur are vasodilation, ‘third spacing’ due to capillary leak, and myocardial dysfunction.

Diagnosis

The following may be present:

- High fever

- Hypothermia, particularly neonates and malnourished patients
- Hypotension (late finding)
- Prolonged capillary refill time of >3 seconds (cold shock)
- Flash capillary refill (warm shock)
- Altered mental status
- Decreased urine output.

Children with concerning findings on physical examination should receive an IM dose of antibiotics immediately, and should be referred to the hospital if in a clinic setting. Concerning signs include convulsions, lethargy or unconscious, tachypnea, severe chest indrawing, nasal flaring or grunting, bulging fontanelle, umbilical redness, fever or hypothermia, severe skin pustules.

2007 American College of Critical Care Medicine algorithm for management of septic shock in children is as follows⁴:

Table 2. Shock in children

History	Examination	Investigation
Nature of onset?	As per rapid initial assessment plus:	Haemoglobin
Any trauma?	Fever?	
Bleeding?	Any bruises or bleeding?	Malaria blood smear
History of congenital or rheumatic heart disease?	Heart murmur?	
History of diarrhoea?	Distended neck veins?	Blood cultures if fever
Any febrile illness?	Enlarged liver?	
Any known regional outbreaks of infectious diseases eg: meningitis/dengue fever?	Petechiae?	Microscopy of CSF and urine
Are they able to feed?	Purpura?	
	Level of consciousness?	HIV testing

- Prompt recognition
- Initial resuscitation
- Diagnosis
- Appropriate antibiotics
- Source identification and control
- Early access to critical care if available, but delays in access to a critical care bed should not delay these interventions
- Institution of inotropic support
- Management of metabolic derangements
- Frequent, repeated assessment of the response to therapy

Early aggressive fluid therapy should be used with caution in LIC

- The recent FEAST trial results⁵ suggest the traditional recommendations of aggressive fluid resuscitation for patients with septic shock may not be applicable in the LIC environment (see page 81). Use IV fluids with caution when resuscitating patients in septic shock in LIC, particularly if there is concern that the patient has a malarial infection. Start with isotonic fluids (Ringer's) at normal maintenance rate initially. 5% dextrose must NOT be used in this setting
- Malnourished children require slower rehydration and careful observation (every 5-10 minutes) as they are at greater risk of congestive heart failure from over-hydration
- Anaemia is common in LIC; consider blood transfusion early

Other interventions in septic shock are as follows

- Within the first hour (ideally within the first 15 minutes), give the first dose of antibiotics
- Correct any abnormalities in calcium and glucose
- If venous access is difficult, place an intraosseous line to administer fluids, antibiotics, blood products and inotropes if required
- If available, place a central venous catheter with central venous pressure (CVP) monitoring if more than 60ml.kg⁻¹ fluid boluses are required
- Consider starting inotropes if a patient continues in shock despite fluid resuscitation: use dopamine (5-9mcg.kg⁻¹.min⁻¹) or adrenaline (0.05-0.3mcg.kg⁻¹.min⁻¹) for cold shock and norepinephrine (0.05-0.3mcg.kg⁻¹.min⁻¹) for warm shock

- If the patient shows signs of respiratory distress and non-invasive or invasive mechanical support is available, use it. Otherwise, give facemask oxygen
- If an infusion pump is not available to administer inotropes, a solution of dopamine can be made by placing 200mg dopamine into 100ml normal saline in the burette of a paediatric microdrop IV set. Titrate this infusion to maintain the blood pressure in the normal range. Similarly, adrenaline or noradrenaline 1mg can be added to 100ml saline and titrated to effect in a microdrop burette
- Inotropes are always best administered through an infusion pump when available.

Child presenting with convulsions

The differential diagnosis for a child presenting with convulsions is as follows:

Age less than 2 months

- Birth asphyxia/birth trauma
- Hypoxic ischaemic encephalopathy
- Intracranial haemorrhage
- Haemolytic disease of the newborn
- Neonatal tetanus
- Meningitis
- Sepsis.

Age more than 2 months

- Meningitis
- Encephalitis
- Cerebral or severe malaria
- Febrile convulsion
- Hypoglycaemia
- Head injury (including Non Accidental Injury)
- Poisoning
- Shock (not likely to cause convulsions)
- Diabetic ketoacidosis
- Acute glomerulonephritis with encephalopathy.

A careful history, examination and special investigations is required (see Table 3).

Table 3. Convulsions in children

History	Examination	Investigations
Fever	General: (as per rapid initial assessment +) Fever	Malaria blood smear
Head injury	Jaundice	Glucose
Drug overdose or toxins	Palmar pallor	
History of convulsions: how many, how long did they last?	Peripheral oedema	
Known epilepsy?	Level of consciousness (AVPU)	Lumbar puncture and CSF microscopy if no signs of raised intracranial pressure (unequal pupils, paralysis, irregular breathing, rigid posturing)
If < 1 week: birth asphyxia or trauma	Petechial rash	
	Deep breathing (sign of acidosis)	
	Head/neck:	
	Stiff neck	
	Signs of trauma	
	Pupil size and reaction to light	
	Tense or bulging fontanelle	
	Abnormal posture	

Child presenting with fever

A febrile illness is the commonest cause of children presenting to hospital in LICs. Some causes are only found in particular regions (e.g. dengue fever); some are seasonal or occur in epidemics.

There are three main diagnostic categories:

- *Fever without localizing signs*

- Malaria
- Septicaemia
- Typhoid
- Urine tract infection
- HIV related.

- *Fever with localizing signs*

- Meningitis
- Pneumonia
- Otitis/mastoiditis/sinusitis
- Septic arthritis

- Skin/soft tissue infection
- Viral URTI
- Throat abscess
- Dengue fever.

- *Fever with a rash*

- Measles
- Viral infection
- Meningococcal infection
- Relapsing fever
- Typhus
- Dengue haemorrhagic fever.

A careful history, examination and special investigations are required (see Table 4).

Child with gastroenteritis

Gastroenteritis is a common problem in developing countries, presenting with diarrhoea and vomiting. Pre-existing malnutrition can cause the diarrhoea to be more

Table 4. A child with a fever

History	Examination	Investigation
Duration of fever	As for rapid initial assessment +	Malaria blood smear
Are they in a malarious region?	Stiff neck?	LP if suggestion of meningitis
Skin rash? Headache?	Bulging fontanelle	
Pain on passing urine	Mastoid region tenderness	Urine microscopy
Cough or difficulty breathing?	Rash?	Blood culture
Ear ache	Skin sepsis: pustules, purpura, petechiae	
	Discharge/redness in ear	
	Refusal to move joint or limb	
	Tachypnoea	

severe, prolonged and frequent compared to diarrhoea in the non-malnourished child. Close monitoring in PICU may be necessary to treat life-threatening consequences of gastroenteritis such as severe dehydration and electrolyte abnormalities, especially in children with severe acute malnutrition (SAM).

Differential diagnosis of children who present severely unwell with diarrhoea:

Acute watery diarrhoea:

- Cholera
- Dysentery e.g. shigella.

Persistent diarrhoea >14 days.

- Diarrhoea with severe malnutrition
- Diarrhoea secondary to recent antibiotic use
- Intussusception.

Take a careful history, including the frequency and number of days of diarrhoea, the presence of blood, and any relevant infectious history. Assessment and management of dehydration is described in Table 5.

Pneumonia in children with dehydrating diarrhoea

The clinical classification of pneumonia based on the diagnostic criteria according to WHO¹ should be carefully evaluated in children presenting with dehydrating diarrhoea caused by *Vibrio cholerae*, Enterotoxigenic E Coli (ETEC) or rotavirus.

A child who presents with dehydration is likely to be acidotic, which can cause tachypnoea. Under these circumstances, it can be difficult to differentiate increased respiratory rate due to pneumonia, or acidosis, or both. The following approach is suggested:

- Rehydrate the child in the first 4-6 hours using IV/oral fluids according to the type of dehydration present
- Then continue respiratory rate monitoring according to the standard WHO pneumonia guidelines¹
- Perform a chest radiograph after full hydration to confirm/exclude the diagnosis of pneumonia
- Note: Do not delay antibiotics during the rehydration process in a child with suspected sepsis.

Pneumonia in children with severe acute malnutrition (SAM)

Severe malnutrition significantly increases the risk of death from pneumonia. Clinical signs are relatively poor predictors of pneumonia in malnourished children, so you may fail to diagnose pneumonia if using standard WHO criteria.

- Common bacterial pathogens: *S. aureus*, enteric Gram negative bacilli, particularly *K. pneumoniae* and *E. coli*; Gram positive bacteria such as *S. pneumoniae* and *H. influenza*
- Give routine broad-spectrum antibiotics in children with severe malnutrition.

Table 5. Recognition and management of dehydration in children

Level of dehydration	Examination	Management
Severe dehydration > 10%	≥2 of the following: lethargy/unconscious, sunken eyes, unable to drink, skin pinch returns very slowly (≥2 seconds).	WHO treatment plan C Admit to hospital Rapid IV hydration* (100ml.kg ⁻¹ Ringers lactate**) Frequent reassessment (every 15-30 minutes) Switch to ORS when able to drink Give antibiotics if appropriate Note: 5% dextrose is NOT effective and must never be used for IV rehydration.
Some dehydration 5-10%	≥2 of the following: restless/irritable, sunken eyes, thirsty, skin pinch returns slowly.	WHO treatment plan B Give food and fluid Then as for no dehydration
No dehydration 0-4%	Not enough of the above signs	WHO treatment plan A Treat at home Oral rehydration solution (ORS) Advice mother when to return Follow up in 5 days if not improved

*<12 month old: 30ml.kg⁻¹ in first 1 hour, then 70 ml.kg⁻¹ over next 5 hours

*>12 months old: 30ml.kg⁻¹ in first 30mins, then 70 ml.kg⁻¹ over next 2.5 hours

**If Ringer's lactate/Hartmann's is not available, normal saline (0.9% NaCl) can be used.

Electrolyte abnormalities in children with gastroenteritis

Sodium

Children with gastroenteritis may present with dehydration associated with hyponatraemia (Na<135mmol.l⁻¹) or hypernatraemia (Na > 150mmol.l⁻¹); it is important to measure plasma electrolytes so that these children can be managed appropriately. This is a particular challenge in situations where measurement of electrolytes is not routine. Hyponatraemia is more common in older children, and in patients with malnutrition, whereas hypernatraemia is more common in younger children. The case fatality rate from gastroenteritis is

2.5 times greater in children with hyponatraemia compared to those with normal plasma sodium.

Hyponatraemic dehydration

The following signs and symptoms are suggestive of acute hyponatraemic dehydration in a child with diarrhoea:

- Reduced activity
- Lethargy
- Hypotonia
- Convulsion and coma

- History of invasive diarrhoea
- Diminished deep tendon reflexes.

Treatment of hyponatraemic dehydration

Correct hyponatremia slowly to prevent osmotic demyelination syndrome:

If serum sodium $<120\text{mmol.l}^{-1}$, child asymptomatic:

- Add sodium to the diet
- If the child needs IV fluid for other indications (such as paralytic ileus, dehydration with persistent vomiting), give 0.9% NaCl

If serum sodium $<120\text{mmol.l}^{-1}$, child symptomatic (i.e. convulsions):

- Give 3% NaCl (12ml.kg^{-1} over 4 hours IV, maximum 500ml)

If serum sodium $<110\text{mmol.l}^{-1}$, irrespective of presence or absence of symptoms:

- Give 3% NaCl (12ml.kg^{-1} over 4 hours IV, maximum 500ml)

Hypernatraemic dehydration

The following signs and symptoms are suggestive of acute hypernatraemia in children with diarrhoea:

- Irritability
- Excess thirst
- Presentation during winter months (association with rotavirus diarrhoea)
- History of intake of several packets of ORS or of inappropriately concentrated ORS
- Exaggerated deep tendon reflexes
- Fever.

Treatment of hypernatraemic dehydration

The serum sodium concentration should be reduced slowly at a rate of 0.5mmol.l^{-1} per hour to prevent cerebral oedema and convulsions. Estimate the fluid deficit and correct the deficit using reduced osmolarity Oral Rehydration Solution (i.e. WHO recommended ORS, $\text{Na}^+ 75\text{mmol.l}^{-1}$):

- Give ORS PO to replace the fluid deficit over 24-48 hours
- If the child cannot take ORS by mouth, then rehydrate via the nasogastric route

- Give fluids IV in the following circumstances only:
 - Circulatory collapse (hypovolaemic or septic shock)
 - Persistent vomiting (3 or more episodes per hour)
 - High purging (stool output $>15\text{ml.kg}^{-1}.\text{hour}^{-1}$)
 - Paralytic ileus

If IV fluids are required for above conditions, give 0.9% saline 20ml.kg^{-1} IV over 2 hours, then continue with ORS.

Potassium

The normal plasma potassium is $3.5-5.3\text{mmol.l}^{-1}$. The symptoms of hypokalaemia and hyperkalemia are similar:

- Lethargy
- Abdominal distension with ileus
- Bradycardia.

They may be distinguished by ECG morphology:

- Hypokalaemia
 - Narrowing QRS complex
 - Flat T waves
 - Prolonged ST segment
 - U waves
- Hyperkalaemia
 - Tall, peaked T wave (mild hyperkalaemia)
 - Prolonged PR interval
 - ST segment depression
 - Loss of P wave
 - Widening QRS complex, gradually leading to 'sine wave' in severe hyperkalaemia, then asystole

Treatment of hypokalemia

Hypokalemia is common in children with SAM, and fluid overload a great concern if hypokalemia is treated with IV fluids. Oral potassium supplementation is usually adequate:

- Give potassium 4mmol.kg^{-1} per day in divided doses 8 hourly PO for 5 days.
- If a child requires IV fluids for severe dehydration, or has a large stool output that cannot be managed with ORS,

then add potassium to IV fluids as follows:

- If serum potassium is $<2\text{mmol.l}^{-1}$, increase potassium in IV fluids to 40mmol.l^{-1}
- If serum potassium is between 2 and 2.5mmol.l^{-1} , increase potassium in IV fluids to 30mmol.l^{-1}

Treatment of hyperkalemia

- Check potassium not being given in IV fluids
- Rehydrate; may be sufficient to treat mild hyperkalemia ($\text{K}^+ < 6\text{mmol.l}^{-1}$)

If serum K^+ is $>6.0\text{mmol.l}^{-1}$:

- **10% Calcium gluconate:** 0.5ml to 1.0ml.kg^{-1} over 2 to 5 minutes IV
 - Reduces cardiac toxicity
 - Protective effect within minutes, but effective only for an hour
- **Salbutamol (nebulised or inhaler): 2.5 to 5mg inhaled:**
 - Increases potassium movement into the cells by increasing the activity of Na-K-ATPase
 - Very effective, for up to 2 to 4 hours
- **8.4% Sodium bicarbonate: 1 to 2mmol.kg^{-1} over 3 - 5 minutes IV:**
 - Increases pH and shifts potassium into the cells
 - Effect begins in 5 - 15 minutes, lasts for 1 to 2 hours
 - Bicarbonate causes precipitation of calcium; flush the IV line between drugs
- **Insulin and glucose:** dextrose 0.5g.kg^{-1} IV + insulin 0.3 unit per gram of dextrose over 30 minutes. Be sure to check a blood sugar in 30 minutes to assure the patient does not develop hypoglycemia.
 - Use if other measures fail
 - E.g. for a 5.0kg child: 2.5g of glucose (25ml 10% or 10ml 25% dextrose) + 1.5 unit (5×0.3) insulin
- **Calcium resonium:** 1g.kg^{-1} PO or PR
 - Binds potassium in the gut and permanently removes
 - Contraindicated in patients with diarrhoea, hypovolaemia or uraemia since it may precipitate colonic necrosis
- **Furosemide:** $1\text{-}2\text{mg.kg}^{-1}$ – effective in only those patients that have renal function and native urine output

- Increases potassium wasting in urine

- **Dialysis:** If available, when other measures fail.

Typhoid fever

Typhoid fever affects more than 21 million people globally each year. It is due to ingestion of food or water contaminated with *Salmonella typhi*, and is most common in school-aged children or young adults in areas of over-crowding with poor sanitation. Transmission is usually from a chronic carrier and it is endemic in many low-income countries, most commonly in Central and South-East Asia.

The WHO recommendation is to consider typhoid if a child presents with fever lasting 7 or more days with the following signs (and malaria has been excluded):

- Constipation followed by diarrhoea ('pea soup')
- Vomiting
- Abdominal pain
- Headache
- Cough.

The main diagnostic features of typhoid are:

- Fever with no obvious focus of infection
- No features of meningitis
- Inability to feed
- Convulsions
- Lethargy and disorientation
- Persistent vomiting
- Rose spots on the abdominal wall
- Hepatosplenomegaly and a distended and tender abdomen.

Complications

- Mild hyponatremia and hypokalaemia are common
- Enteric encephalopathy is seen in 10-30% of cases of severe enteric fever, and presents with altered consciousness, disorientation, confusion and delirium, mainly in children and young adults. The case fatality from enteric encephalopathy is high. A positive Widal test for typhoid fever, leucopenia, and severe dehydration are predictors of encephalopathy

- Perforation of an ulcer in the small bowel. The child presents critically unwell in the second to third week of untreated illness.

Treatment of Salmonella typhi

- IV fluids (blood transfusion)
- Antibiotics (antibiotic resistance is common)
 - Ciprofloxacin IV (Africa)
 - Ceftriaxone or cefotaxime IV (Asia)
 - Add metronidazole and gentamicin in cases of bowel perforation
- Surgery for bowel perforation; high mortality. If there is general peritonitis the child will require a laparotomy for peritoneal washout and oversewing of the typhoid perforation. Bowel resection may be required for multiple perforations. A localised mass may be treated conservatively (as for an appendix mass)
- Dexamethasone 3 mg.kg⁻¹ loading dose then 1 mg.kg⁻¹ per dose IV 6 hourly for 48 hours – this is recommended mainly for enteric encephalopathy with or without multi drug resistant (MDR) enteric fever, and improves outcomes
- Exclude hypoglycaemia
- Treat electrolyte abnormalities.

The child with malnutrition

Malnutrition is a major independent risk factor for death in children in developing countries, and increases the risk of mortality from other conditions such as septic shock, acute respiratory infection, diarrhoea, malaria, or measles.

Severe Acute malnutrition (SAM) is defined by the WHO as a weight-for length/height < 3SD below normal, a mid-upper arm circumference < 115mm, or oedema of both feet (sign of kwashiorkor).

The cause of the high mortality among malnourished children is unclear, but may relate to both changes in physiology and poor case management. All aspects of physiology are affected including cardiovascular, GI, renal, endocrine and immune function.

A standardised management protocol for severely malnourished children has been developed in the Dhaka hospital at the International Centre for Diarrhoeal Disease Research, Bangladesh (ICDDR,B), which has resulted in a 47% reduction in death rates among severely malnourished children. The management of diarrhoeal disease in children with malnutrition is divided into three phases:

Acute phase

- Identify and treat problems that endanger life e.g. hypoglycaemia and infection
- Initiate feeding according to standard feeding schedule
- Correct micronutrient deficiencies
- Give broad-spectrum antibiotics
- Vitamin and mineral supplementation
- Slow rehydration using oral rehydration therapy
- Early recognition of complications

Mortality is highest during this phase of management, the principal causes being hypoglycaemia, hypothermia, infection, and water-electrolyte imbalance.

Nutritional rehabilitation phase

- Recover lost weight - intensive feeding.
- Stimulate the child emotionally and physically
- Train the mother to continue care at home

Follow up

Follow up prevents relapse of severe malnutrition, and ensures proper physical growth and development of the child

A number of critical problems may be seen during the acute stage of treatment of malnutrition:

Hypoglycaemia

Children with severe malnutrition and hypoglycaemia may die within minutes. Test the child for hypoglycaemia on admission or whenever you find lethargy, convulsions or hypothermia. If blood glucose cannot be measured, suspect all children with SAM to have hypoglycaemia and treat accordingly. Hypoglycaemia can also be a sign of infection.

- If the child is conscious and blood glucose is <3mmol.l⁻¹ (54mg.dl⁻¹): give 50ml of 10% glucose or 10% sucrose solution PO or NG (1 rounded teaspoon of sugar in 3.5 tablespoons water). The 'starter diet' (F-75) is given every 30 minutes for two hours (giving one quarter of the two-hourly feed each time). Thereafter, two-hourly feeds are continued for first 24-48 hours
- If the child is unconscious, lethargic or convulsing, give 10% glucose 5ml.kg⁻¹ IV, followed by 50ml of 10% glucose or sucrose by NG tube. Then give the starter diet F-75 as above.

Septic shock

- Defined as weak or absent radial pulse, delayed CRT (> 3sec), cold peripheries, or hypoglycaemia
- Give fluid bolus 15ml.kg⁻¹ IV over one hour using 5% dextrose 0.9% saline
- 'Cholera saline' with 5% dextrose is preferred if there is a history of watery diarrhoea (Na+133 mmol.l⁻¹, K+ 13mmol.l⁻¹, Cl- 98mmol.l⁻¹, acetate 48mmol.l⁻¹)
- Repeat rescue therapy once if signs of shock remain
- Provide broad-spectrum antibiotics: IV ceftriaxone 100mg.kg⁻¹ once daily and gentamicin 5mg.kg⁻¹.day⁻¹ in divided doses 12 hourly
- Supportive measures include oxygen therapy, correction of hypoglycaemia, hypothermia or acidosis.

Hypothermia

- Wrap the child in blankets if axillary temperature is <35°C
- Place an electric lamp close to the body but sufficiently away to avoid burns
- Measure temperature every 30 minutes during re-warming with a lamp, as the child may become hyperthermic
- Measure rectal temperature with a rectal thermometer; never use an oral thermometer for this purpose
- Start feeding (hypothermia can co-exist with hypoglycaemia).

SUMMARY

In summary, a basic PICU is an opportunity to focus and concentrate resources in terms of personnel, drugs and equipment such that the most critically unwell children have a better standard of medical and nursing care, are observed 24 hours a day and can have vital services available such as oxygen to maximize their chance of survival.

REFERENCES

1. WHO pocket book of hospital care for children, 2nd edition. Geneva, World Health Organisation, 2013. Available from http://www.who.int/maternal_child_adolescent/documents/child_hospital_care/en/ (accessed 22/10/14).
2. The clinical use of oxygen. Geneva, World Health Organisation, 2011 (Fig 6.1, page 38, available from http://video.rch.org.au/cich/The_Clinical_Use_of_Oxygen_November_2011.pdf accessed 20/01/15).
3. Duke T. CPAP: a guide for clinicians in developing countries. *Paediatr Int Child Health* **34**(2014): 3-11.
4. Brierley J et al. Clinical practice parameters for hemodynamic support of pediatric and neonatal septic shock: 2007 update from the American College of Critical Care Medicine. *Crit Care Med.* 2009 Feb;37(2):666-88. doi: 10.1097/CCM.0b013e31819323c6.
5. <http://www.feast-trial.org/> (accessed 20/01/15). Now available as: Maitland K, Kiguli S, Opoka RO et al. Mortality after fluid bolus in African Children with severe infection. *NEJM* 2011; **364**: 2483-95

Recognising the seriously ill child

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INTRODUCTION

Each year approximately 11 million children die before reaching the age of five, 99% from low- and middle-income countries (LMIC).¹⁻³ Three-quarters of deaths are from preventable or treatable causes such as pneumonia, diarrhoea, malaria, and measles. Children can become unwell very quickly, and the outcome from cardiac arrest in a child is poor, so early recognition and treatment of the seriously ill child is vital. In the developed world the recognition, assessment and management of the seriously ill child has improved following the introduction of courses such as Advanced Paediatric Life Support⁴ (APLS) in the UK and Paediatric Advanced Life Support⁵ (PALS) in the US, and these courses are now often mandatory for clinicians working with children.

Studies have shown that many health workers in emergency facilities in resource-poor countries have no standardised assessment or treatment protocols for severely ill children, but that improved training, assessment and emergency care could improve outcomes.^{6,7} In response, the WHO has developed the Emergency Triage Assessment and Treatment (ETAT) system to reduce childhood mortality, particularly within the first 24 hours of admission.⁸ This course has been shown to significantly improve care for children presenting with common serious illnesses (e.g. dehydration, pneumonia and severe malnutrition).⁹ Approximately 50% of children who die after admission to hospital do so in the first 24 hours.¹⁰ ETAT+ has been developed to include admission care for the first 24 hours.¹¹

This article will focus on the assessment, recognition and initial management of the seriously ill child and is based on ETAT and APLS principles; the topics of paediatric life support, trauma and neonatal resuscitation

are also included in this edition of *Update in Anaesthesia* (pages 264 and 269).

PRINCIPLES OF MANAGEMENT OF THE SERIOUSLY ILL CHILD

In order to recognise the child who is unwell it is important to know the normal physiological values for different age groups, signs of critical illness, and how children compensate for serious illness. It is important to be prepared to receive a critically ill child to your facility, to understand the principles of triage and the 'ABC' approach to assessment and treatment.

The normal values of heart rate, respiratory rate and systolic blood pressure are shown in Table 1.¹²

Physiological compensation

Children can compensate effectively during the early stages of serious illness, which may mask how unwell they really are. 'Compensation' refers to the ability to maintain perfusion of 'vital' organs such as the brain and heart at the expense of 'non-vital' organs such as skeletal muscle and gut. When compensatory mechanisms fail, decompensation occurs, which if unaddressed, rapidly leads to organ failure and death. Signs of compensation include increased respiratory rate, increased heart rate and peripheral vasoconstriction causing cold extremities. Health workers must be vigilant to signs of compensation so that intervention and treatment can be started promptly. Signs of decompensation include hypotension and bradycardia, and babies may develop apnoeic episodes.

Assessment of end-organ function is important and can also indicate decompensation. For instance, children may appear outwardly well, but they are listless, not interested in their surroundings and tolerate examination and

SUMMARY

Use the ABC approach to assess and treat the seriously ill child

Normal physiological values vary by age

If the physiological values are normal, this does not always mean the child is well

Children can compensate for serious illness effectively, which may mask how unwell they are

When treating an unwell child, it is important to reassess them after every intervention

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Table 1. Normal heart rate, respiratory rate and blood pressure values for children

	Neonate	Infant	Small child	Adolescent
	< 1 month	< 2 years	2 - 5 years	5 - 12 years
Heart rate (min⁻¹)	110-160	100-150	80-120	60-100
Resp rate (min⁻¹)	30-40	25-35	25-30	15-20
Systolic BP (lower limit of normal, mmHg)	5-70	70-80	65 + age x2	90-120

interventions without complaint; this can be an early sign of neurological decline or fatigue. Confusion in a child is a very worrying sign and indicates inadequate cerebral perfusion. Conscious level can be assessed quickly using the 'AVPU' score (Alert, responds to Voice, responds to Pain, Unconscious). Unlike adults, reduced urine output due to inadequate renal perfusion is often a late sign in children, and therefore not useful in initial emergency care. However, if a mother reports that the child has not passed urine, this is a serious sign.

PREPARATION

Health facilities caring for sick children must provide not only essential drugs and equipment but also ensure competence and ongoing training of all staff to foster excellent resuscitation team performance.

When preparing to receive a sick child to your facility, individual roles and responsibilities of the resuscitation team members should be clearly understood. The WETFLAG mnemonic (Weight, Energy, Tube, Fluids, Adrenaline, Glucose) is helpful to prepare to receive a sick child to the hospital facility⁸ (see Box 1). A worked example of the WETFLAG calculations for a 5-year-old child is given in Box 2, and a table of calculations for children <1 year in Table 2.

Box 1. 'WETFLAG' mnemonic

Weight*

0-12 months = (0.5 x age (months)) + 4

1-5 years = (2 x age) + 8

6-12 years = (3 x age) + 7

(* Estimation of weight is a guide only)

Energy = 4 x weight (J)

(Energy required for defibrillation)

Tube = age/4 + 4

(Approx. size of uncuffed tracheal tube)

Fluids = 20ml.kg⁻¹ of 0.9% saline

(Fluid bolus for shocked child)

Adrenaline = 10mcg.kg⁻¹ = 0.1ml.kg⁻¹ of 1:10,000

(Dose in cardiac arrest)

Glucose = 2ml.kg⁻¹ 10% glucose

(Treatment of hypoglycaemia)

Box 2. Example of WETFLAG calculations for a 5-year-old child

5-year-old child

Weight = (2 x 5) + 8 = 18kg

Energy = 4 x 18 = 72J

Tube = 5/4 + 4 = 5.5 - 6

Fluids = 20ml x 18 = 360ml of 0.9% saline

Adrenaline 0.1ml.kg⁻¹ = 1.8ml 1:10,000

Glucose = 2ml x 18 = 36ml 10% glucose

TRIAGE AND THE ABCDE + DEFG APPROACH

Triage is a system to prioritise who needs to be treated first. Initial triage involves categorizing children who present to the emergency department into three groups of urgency: emergency cases, priority cases and non-urgent cases (see Box 3).

Emergency cases

Signs of a potential emergency case are identified from conducting a rapid primary survey of any child presenting for treatment. The child is assessed using the ABC approach in order to identify those abnormalities that are most rapidly lethal:

Airway. Are there signs of airway obstruction?

Breathing. Is the child having difficulty breathing? (e.g. increased work of breathing, using accessory muscle, cyanosis, abnormal noise such as stridor, wheeze, or silent chest.)

Circulation. Does the child have signs of circulatory failure? e.g. cold peripheries, a rapid, weak pulse or capillary refill time > 2 seconds?

Disability or Dehydration. Is the child Awake, or do they have a decreased level of consciousness (assessed quickly using the AVPU score)

Table 2. WETFLAG calculations for a child less than 1 year. Reproduced with permission. Lorazepam is included as first line treatment for seizures.

University Hospital Southampton  NHS Foundation Trust							
Infant	Weight (KG)	Energy	ET Tube (mm)	Fluid	Lorazepam	Adrenaline (ml)	Glucose (ml)
Months	(0.5 x Age) + 4	4j/KG	Uncuffed (Cuffed)	20ml/kg NaCL 0.9%	0.1mg/kg	0.1ml/kg 1:10'000	2ml/kg 10%
0	4	16	3-3.5 (3)	80	0.4	0.4	8
1	4.5	18	3-3.5 (3)	90	0.45	0.45	9
2	5	20	3-3.5 (3)	100	0.5	0.5	10
3	5.5	22	3-3.5 (3)	110	0.55	0.55	11
4	6	24	3-3.5 (3)	120	0.6	0.6	12
5	6.5	26	3-3.5 (3)	130	0.65	0.65	13
6	7	28	3-3.5 (3)	140	0.7	0.7	14
7	7.5	30	3-3.5 (3)	150	0.75	0.75	15
8	8	32	3-3.5 (3)	160	0.8	0.8	16
9	8.5	34	3-3.5 (3)	170	0.85	0.85	17
10	9	36	3-3.5 (3)	180	0.9	0.9	18
11	9.5	38	3-3.5 (3)	190	0.95	0.95	19
12	10	40	3-3.5 (3)	200	1	1	20

NB - Fluid for trauma = 10ml/kg NaCl 0.9% x 4 then consider blood transfusion

NB - Always escalate Defib Energy to next Joules Up, Remembering 150j is adult dose (Biphasic)

NB - Drugs are maximum drug calculation - use clinical judgement

Box 3. Triage categories

5-year-old child

Emergency cases	→	Immediate emergency treatment
Priority cases	→	Assessment and rapid attention
Non-urgent cases	→	Can wait their turn in the queue

Exposure. Are there visible signs of trauma (e.g. fracture) or disease (e.g. rash)? Is the temperature normal? (very hot or very cold)

+ **Don't Ever Forget the Glucose.** Does the child have hypoglycaemia?

These emergency signs must be treated IMMEDIATELY they are discovered, before moving on to the next step.

Priority cases

Once emergency signs are excluded, look for the conditions that need to be treated as a priority. Deciding which children fit in the 'priority' category can be difficult; in the ETAT course this has been developed as the '3TPR-MOB' mnemonic.⁸ The mnemonic stands for 3xT (tiny, temperature, trauma), 3xP (pallor, poisoning, pain), 3xR (respiratory, restless, referral), and malnutrition, oedema, burns (see box 4 for an explanation of the 3TPR-MOB priority clinical signs).

If any of the priority signs are identified, the child must be seen quickly, blood taken for emergency investigations including glucose, malaria smear and haemoglobin (Hb), and senior help should be sought.

There are a few conditions such as severe malnutrition, anaemia or cardiac disease that must be identified as part of the initial triage process, as modifications in management will be needed. For example, children with malnutrition who develop diarrhoea are at a higher risk of death than those who are well nourished, and management of children with malnutrition must be modified.¹³ These special circumstances are discussed below.

The ABCDE + DEFG approach

The core cycle of assessment, treatment and reassessment using Airway, Breathing, Circulation, Disability, Exposure (ABCDE) is fundamental to the safe and effective care of sick children, and facilitates communication between healthcare workers.

For this approach to be successful, the child must be reassessed regularly, and accurate timed records kept, with results communicated to all team members.

Box 4. 3TPR-MOB – priority signs when assessing the ill child (ETAT)

3 Ts:

Tiny baby less than 2 months (because difficult to assess, deteriorate quickly)

Temperature: child very hot (high fever)

Trauma (including hidden head and abdominal injuries)

3Ps:

Pain (may indicate a severe condition)

Pallor (severe anaemia)

Poisoning (if history, may need specific urgent treatment)

3 Rs:

Respiratory Distress (if severe, this is an emergency)

Restless: continuously irritable or lethargic (may indicate a severe condition)

Referral: urgent referral to your facility

MOB:

Malnutrition: visible severe wasting (specific treatment protocols are used)

Oedema: both feet (may indicate severe malnutrition)

Burns (may cause urgent airway problem, severe respiratory distress, severe pain, be associated with large fluid losses or other injuries)

Airway

There are some important features to be aware of when assessing and managing the airway in a child (see also page 4)

- Nasal breathing. Infants less than 6 months old breathe predominantly through their nose; nasal obstruction can result in severe respiratory distress, which may be relieved by simple suction to clear the airway.
- Tongue. The tongue in an infant is relatively large and may obstruct the airway when the conscious level is impaired.
- Teeth. These may be loose in children between around 6-13 years of age.
- Adenotonsillar hypertrophy. This is common in 3-8 year olds and may cause upper airway obstruction.
- Soft palate and tonsils. These may be damaged when an oropharyngeal airway is inserted; airways must be inserted with care.
- Trachea. Soft and short, and prone to external compression, including from cricoid pressure. It is very easy to put a tracheal tube down too far.
- Large occiput with short neck in infants tends to force the head into flexion, potentially making airway obstruction worse.
- The airway is narrow in absolute terms. A small amount of airway swelling or obstruction by secretions may result in severe airway obstruction.
- Cricoid ring. This sub-glottic region is the narrowest part of the airway in a small child (compared with the vocal cords in an adult) and is susceptible to oedema. In general, uncuffed tracheal tubes are preferred for children < 8 years old; modern 'microcuff' tracheal tubes with cuff pressure monitoring are a new alternative.

The first question to consider when evaluating the airway is 'is the airway obstructed'?

Observe for:

- Talking or crying – the airway is open
- Noisy breathing. Is this due to stertor or stridor – i.e. partial obstruction above or below the larynx respectively
- 'See-saw' chest and abdominal movements – respiratory effort is present, but potentially with complete airway obstruction.

Common causes of airway obstruction are shown in Box 5.

If there is total airway obstruction, the airway must be opened immediately with simple manoeuvres: head-tilt, chin lift or jaw thrust. Airway adjuncts e.g. oropharyngeal (Guedel) or nasopharyngeal airway may be required. If total airway obstruction

persists despite these simple measures, any subsequent intervention will be determined by the specific history and condition of the child. For example, a child with suspected inhaled foreign body might require specific interventions to clear the airway.

Attempt to provide breaths whilst an assistant maintains airway-opening manoeuvres. If unable, consider direct laryngoscopy and intubation. If unable to intubate or ventilate, proceed urgently to a surgical airway down the 'can't ventilate, can't intubate' emergency airway algorithm (see page 116).

Box 5. *Common causes of airway obstruction in children*

- Reduced level of consciousness
- Inhaled foreign body – sudden onset, often witnessed
- Dental abscess
- Croup – gradual onset, viral illness usually due to parainfluenza virus, with characteristic 'barking' cough
- Epiglottitis – rapid onset, severe, bacterial illness usually due to *Haemophilus influenzae*. The child is 'toxic', with sore throat, drooling, muffled voice and a high temperature, and often adopts a 'tripod' position to maintain the airway
- Tracheitis – systemically unwell, bacterial illness usually due to *Staphylococcus aureus*. Bacterial tracheitis usually occurs as a complication of a viral infection.
- Retropharyngeal abscess – usually due to lymphatic spread of infection from sinuses, teeth or middle ear. The child presents with fever, sore throat and neck pain and swelling. Stridor is not often a major feature.

If the child has a traumatic injury, the cervical spine must be immobilised and the airway must be opened using a jaw thrust to keep the head in a neutral position.

Partial airway obstruction

If there is partial airway obstruction, and the child is unconscious, open the airway using the manoeuvres described above (chin lift, jaw thrust).

If there is partial obstruction and the child is conscious, allow the child to adopt a comfortable position – this will often be sitting up or leaning forwards. Leave the child with their parent or carer as this reduces distress; avoid inspecting

the oropharynx as this can rapidly worsen partial airway obstruction. Do not attempt intravenous cannulation before the airway can be improved; causing the child added distress may worsen the situation. Strategies to manage a child with stridor are shown in Box 6.

Breathing

Look, listen and feel for effort and efficacy of breathing. There are a number of signs to look for in the rapid assessment of breathing, but increased respiratory rate is one of the key indicators of severe illness. Increased respiratory rate may be due to a variety of causes, such as respiratory disease, sepsis or hypovolaemia. The respiratory rate should be assessed and compared to normal values for age, and the trend followed by repeated assessment. Increasing respiratory rate is a worrying sign and usually indicates a child that is tiring and at risk of imminent collapse; a sudden fall in respiratory rate is a sign of this collapse (it is a pre-terminal sign).

A child with respiratory disease will have to work hard to breathe and will tire easily. The airways are relatively narrow and are

Box 6. *Initial management of stridor in a child with partial airway obstruction.*

- Reassure the child; keep them close to their parents or carer
- Give high flow humidified oxygen via a mask
- Consider adrenaline nebuliser: 5ml of 1:1000 adrenaline with oxygen
- Consider antibiotics (epiglottitis) or steroids (croup)

easily obstructed by oedema or secretions. A small reduction in airway diameter leads to a large increase in resistance, and hence the work of breathing. In infants, respiratory mechanics are not very efficient; the ribs are soft and horizontal and the diaphragm is a major muscle of respiration. Abdominal distension is poorly tolerated. The soft ribs mean that subcostal and intercostal recession is relatively common in infants with respiratory infection, but is an ominous sign in an older child in whom the chest wall is relatively more rigid.

The signs to look for in assessment of breathing are shown in Table 3.

Increased work of breathing is a compensatory mechanism and may be effective in keeping oxygen delivery normal. The child will still require support and close watching. Carbon dioxide retention is unusual unless there is a reduced conscious level or lower airway obstruction.

Detecting hypoxaemia with pulse oximetry is an important measurement, especially for children with suspected pneumonia. Regular monitoring of oxygen saturation can be used to guide effective use of oxygen, and is associated with improved outcomes. Untreated hypoxaemia in children with pneumonia is associated with increased mortality.^{14,15} Note that inability to obtain a pulse oximetry reading may be due to reduced perfusion of the extremities due to shock.

It is important to review other systems to look for signs of respiratory distress, such as an increased heart rate (compensation), bradycardia (a pre-terminal sign), skin colour for cyanosis, and level of consciousness for evidence of cerebral hypoxia. Exhaustion, reduced conscious level and slow breathing or apnoea (stopping breathing) are signs of decompensation and are pre-terminal signs.

If the child has compensated respiratory distress, the management must include the following:

- Sit the child up
- Give oxygen – high flow via mask, humidified if possible. The oxygen mask can be held near to the child’s face if they are distressed by it

If the child has decompensated respiratory distress (reduced breathing or apnoeic episodes, cyanosis or desaturation, bradycardia, reduced level of consciousness), intervention to support breathing should be immediate, using 100% oxygen and bag and mask ventilation in the first instance.

Circulation

The next step is to assess circulation, assessing key elements at the same time (this is important to correctly diagnose shock):

- Feel the pulse rate (compare to normal values for age).
- Feel pulse strength (compare the strength of central and peripheral pulses).
- Capillary refill. Ideally the child should be normothermic and should be viewed in good light. Press on a central area such as the chest for 5 seconds; the skin will blanch but normal colour should return within 2 seconds when the pressure is released. Note that vasodilatation in ‘warm’ shock may mean that capillary refill appears to be normal, even if the child has severe sepsis. On its own, capillary refill time is not a reliable sign of cardiovascular compromise.
- Feel the extremities. Are the hands and feet cold compared with central parts of the body? Where does the zone of warmth extend to?
- Blood pressure. Choose the correct size of cuff (as large as fits comfortably on the arm); a normal BP does not always mean all is well. Remember that hypotension is a late sign that decompensation is occurring. As a guide, the lowest level of normal systolic pressure is 65 + (2 x age in years).
- Look for associated respiratory compensation (tachypnoea).
- Assess end-organ function: level of consciousness (AVPU – see Disability below). Confusion in a child is a worrying sign.

Signs to look out for when assessing the cardiovascular system are shown in Table 4.

The management of cardiovascular insufficiency in children must include:

Table 3. Assessment of breathing

Signs of increased effort i.e. increased work of breathing	Signs of efficacy of breathing i.e. is the respiratory effort effective?
<ul style="list-style-type: none"> • Respiratory rate (compare to normal values) • Body position - sitting forward or adopting ‘tripod’ position • Recession - intercostal, subcostal and or sternal • Tracheal tug • Grunting • Wheeze • Use of accessory muscles (e.g. sternocleidomastoid in the neck) • Nostril flaring • Head bobbing 	<ul style="list-style-type: none"> • Colour - look for central cyanosis • Oxygen saturation • Breath sounds (a silent chest is a pre-terminal sign) • Chest expansion • Conscious level. Reduction on conscious level is a late sign.

Table 4. Indicators of cardiovascular insufficiency or shock

Severe but compensated shock	Decompensated shock – pre-terminal
<ul style="list-style-type: none"> • Mottled, cold skin • Tachycardia • Weak peripheral pulses • Cold peripheries – to knees or elbows • Prolonged capillary refill (>2 seconds) • Increased respiratory rate 	<ul style="list-style-type: none"> • Hypotension • Bradycardia • Unconscious

- Oxygen – high flow via face mask
- Stop any bleeding
- IV access – intravenous or intraosseous
- Fluids - oral, nasogastric or intravenous.

Intravenous access can be difficult in children with shock. An intraosseous needle is an effective method of fluid and drug administration and should be considered early if intravenous access is difficult. (See article, page 240)

Traditional fluid management for a child in shock has been to give a fluid bolus of 10-20ml.kg⁻¹ 0.9% saline or Ringer’s lactate, followed by reassessment. However, intravenous fluid therapy for children in resource-poor settings has been addressed in an important new study, the Fluid Expansion as Supportive Treatment (FEAST) study, published in the New England Journal of Medicine in 2011. This is a randomised controlled study of over 3000 children in six hospitals in Uganda, Kenya and Tanzania, comparing fluid resuscitation starting with a bolus of fluid to just starting maintenance fluids without a bolus, in children with fever and shock. Shock was defined as signs of impaired perfusion plus impaired consciousness or respiratory distress, or both. Children with gastroenteritis, severe malnutrition, burns or surgical conditions were

excluded. The main finding was that children given a fluid bolus of 20-40 ml.kg⁻¹ 0.9% saline or 5% albumin did worse, with an increased risk of mortality compared to the control group who received maintenance fluids only.¹⁶

This was a well-conducted study and has provoked much debate.¹⁷⁻²¹ The children were severely unwell by any measure, but there were no intensive care facilities in the study hospitals. Many children had malaria and were anaemic, but the detrimental effects of fluid bolus were still seen in those without malaria and those without severe anaemia. The children appeared to die from cardiovascular collapse (rather than fluid overload), within 24-48 hours of treatment.²⁰ Excess mortality associated with fluid bolus was still seen in a smaller group of children who met the more strict WHO criteria for sepsis (i.e. capillary refill time > 3 secs, cold peripheries, a weak pulse, and a fast pulse).²¹ The implications of the FEAST study are that children with febrile illness and shock in Africa should receive maintenance fluids only (Ringer’s lactate 5% dextrose or 0.9% saline 5% dextrose), and aggressive resuscitation with boluses of 0.9% saline or albumin should be avoided. From a pragmatic point of view, this would appear to be a safer course of action in hospitals with low numbers of nursing staff and without burettes to accurately measure fluid volumes, and no backup intensive care facilities.

Figure 1. Intraosseous needle



Disability: neurological assessment

Make a quick assessment of neurological function. This is essential to assess end-organ function. If the child is alert, this indicates that there is adequate cardio-respiratory compensation; a child with decompensated cardiorespiratory failure will have a depressed conscious level. Depressed conscious level or confusion may also be due to a primary cerebral cause (trauma or cerebral infection).

The three quick assessments are:

- Pupils (size and reactivity to light) – always compare left and right
- Posture

- Conscious level assessed using the AVPU system:
- **A** – Alert
- **V** – responds to Voice
- **P** – only responds to Painful stimuli
- **U** – Unresponsive to painful stimuli.

AVPU is a quick reliable method of assessing conscious level without using an age-specific Glasgow or Blantyre coma scale.¹⁵ In general, a child responding to pain (P) or unresponsive (U) corresponds to a GCS of 8 or below and will likely need airway support.

Exposure

- Check for rashes, burns and bruises or other injuries
- Check temperature.

Glucose

Don't Ever Forget Glucose (DEFG) is the final part of the disability assessment, especially in children with a reduced conscious level. Aim for a blood glucose of >2.5mmol in a well nourished child, >3mmol in a malnourished child.

**Treat hypoglycaemia with 10% dextrose
2 ml.kg⁻¹ IV, or with oral glucose**

Review other systems for signs of neurological failure:

- *Airway.* Reduced conscious level will eventually lead to airway obstruction

- *Breathing.* Increased intracranial pressure may present as hyperventilation, Cheynes Stokes respiration or apnoea
- *Circulation.* Bradycardia + hypertension = Cushing's response, a pre-terminal sign of elevated intracranial pressure

Dehydration

Once shock has been treated (if present), make an assessment of fluid deficit in order to calculate the fluid requirements of the child over the next 24 hours:

**Total fluid requirement = degree of dehydration +
maintenance fluid + ongoing loss**

A guide to assessing dehydration in children is provided in Table 5, and a case example putting everything together is shown in Box 7.

SPECIAL CIRCUMSTANCES

Cardiac disease

It is important, particularly in newborns, to consider cardiac disease as a cause for cardiovascular insufficiency and shock. Signs to look for include:

- Cyanosis – not correcting with oxygen. Ideally all newborn infants should be screened for cyanotic heart disease using pulse oximetry
- Tachycardia
- Raised jugular venous pressure

Table 5. Clinical assessment of dehydration in children

Clinical sign	Mild	Moderate	Severe
Weight loss	Less than 5%	5-10%	Greater than 10%
Total fluid deficit	Less than 50 ml.kg ⁻¹	50-100 ml.kg ⁻¹	>100 ml.kg ⁻¹
General appearance	Alert	Irritable, thirsty	Lethargic, drinks poorly
Mucous membranes	Normal	Dry	Dry
Eyes	Normal	Normal	Sunken
Respiration	Normal	Fast	Fast
Pulse	Normal	Fast	Fast, weak
Blood pressure	Normal	Normal	Low
Pinched skin* (Pinch skin between thumb and forefinger on abdomen or thigh)	Springs back	Slow – skin fold present less than 2 seconds	Very slow – skin fold greater than 2 seconds

Box 7. Putting it all together - case example

A 5-year-old girl is brought to the emergency department with diarrhoea and a poor appetite. She is lethargic and refusing water. She has a respiratory rate of 40 and a weak femoral pulse with a rate of 140bpm. Her blood pressure is 100mmHg systolic. How would you assess and manage this child?

Preparation. Ensure the team and equipment are prepared to receive the child – remember **WETFLAG**:

5 year-old child...

$$\text{Weight} = (2 \times 5) + 8 = 18$$

$$\text{Energy} = 4 \times 18 = 72\text{J}$$

$$\text{Tube} = 5/4 + 4.5 = 5.5\text{-}6.0$$

$$\text{Fluids} = 20 \text{ ml.kg}^{-1} = 360\text{ml } 0.9\% \text{ saline}^*$$

$$\text{Adrenaline} = 0.1 \text{ ml.kg}^{-1} : 10,000 = 1.8\text{ml}$$

$$\text{Glucose} = 2 \times 18 = 18\text{ml } 10\% \text{ dextrose}$$

Triage – look for emergency signs

She has a clear airway but has rapid breathing, fast pulse and has dehydration with lethargy. This child must be treated immediately with:

- Breathing – high flow oxygen via face mask and sit upright
- Circulation and Dehydration – If possible give oral fluids, however with a lethargic child it may be necessary to consider an intravenous fluid bolus 20 ml.kg^{-1} 0.9% saline if there are signs of shock in association with a surgical diagnosis or acute dehydration from gastroenteritis.
- Check blood for glucose, malaria and Hb

Assess and treat ABCD + DEFG

Move on to a full ABC assessment. Her airway is clear and maintained unaided. For assessment of breathing look at effort and efficacy (see Table 2). She has a respiratory rate of 40 breaths per minute, indicating an increased effort of breathing. She is not using accessory muscles. She has good air entry on auscultation with SpO_2 99% in room air, demonstrating good efficacy despite increased effort. We have already started oxygen via facemask and sat her upright as part of our emergency management. This is sufficient treatment for breathing.

As A+B are now stable, move on to circulation. Her pulse rate is fast and thready. She is in the 'compensated' phase of shock - her blood pressure is normal for her age and she remains conscious. She has acute dehydration and is refusing oral fluids. There are no signs of cardiac disease, such as cyanosis or liver enlargement or malnutrition. She should receive an IV fluid bolus 20 ml.kg^{-1} . After IV fluids her heart rate reduces to 110bpm.

To calculate her ongoing fluid requirement, first calculate her level of dehydration (Table 4). She is lethargic with a raised respiratory rate and tachycardia. She has at least a 10% deficit with clinical signs of hypovolaemia. She is too unwell for oral rehydration. Her fluid requirements for the next 24 hours are: (Total fluid requirement = degree of dehydration + maintenance fluid + ongoing loss)

10% deficit (100 ml.kg^{-1}) in an 18kg child = 1800ml

- Replace 50% over 8 hours = 900ml
- Replace the remaining volume over the next 16 hours = 900ml

Maintenance = 100 ml.kg^{-1} for first 10kg (100×10) + 50 ml.kg^{-1} for next 10kg (50×8) = 1400ml

- i.e. IV fluids for first 8 hours = 900ml replacement + 466ml maintenance.
- For next 8-24 hours = 900ml replacement + 933ml maintenance.
- Give as Ringer's lactate 5% dextrose

We can move on to assess Disability. Using the AVPU assessment tool she is responding to voice and is only lethargic. This is still an important sign and must be reassessed during and after treatment. A blood glucose is taken and is within normal limits.

After so many interventions it is important to reassess her and treat any abnormal signs before she is transferred to a paediatric ward for ongoing fluid resuscitation and investigation.

- Gallop rhythm/murmur
- Enlarged liver
- Absent femoral pulses.

Fluids must be given cautiously. This topic is covered in more detail on page 81.

Malnutrition

Malnutrition is a contributing factor in approximately one third of child deaths, making children more susceptible to severe disease.¹⁵ It is vital that malnutrition is identified in seriously ill children as specific management strategies must be adopted.

The child with serious malnutrition undergoes metabolic and physiological changes to conserve energy and preserve essential processes.²² If these changes are not acknowledged when initiating treatment, the child will be at increased risk of death from heart failure, electrolyte imbalance, hypoglycaemia, hypothermia and they may have untreated infection. Mortality rates of up to 60% are seen in the most severe group.²³

Signs of malnutrition include:

- Severe wasting
- Oedema in feet
- Underweight for age.

The WHO has produced guidelines for the management of severe malnutrition, which outlines key steps for initial management²⁴:

- The child should be fed every 2–3 hours, day and night, to prevent hypoglycaemia and hypothermia
- Keep the child warm
- Rehydrate with low sodium fluids; monitor closely for signs of fluid overload; avoid intravenous fluids, except in shock
- Give 100kcal.kg⁻¹.day⁻¹ and 1g protein.kg⁻¹.day⁻¹
- Give potassium and magnesium to correct electrolyte imbalance; restrict sodium
- Give micronutrient supplements; do not give iron
- Give broad spectrum antibiotics even when clinical signs are absent as infections can be silent.

Critically, children with severe malnutrition must not be aggressively resuscitated with IV fluids as this may lead to heart failure. Intravenous fluids should not be given unless the child is lethargic or unconscious and shocked. When restarting feeding malnutrition protocols should be used. A suggested regimen for fluid resuscitation for a child with malnutrition and acute dehydration is shown in box 8.

Box 8. Fluid resuscitation for a child with malnutrition and shock due to acute dehydration from gastroenteritis

Slow IV fluid bolus = 15ml.kg⁻¹ Ringers lactate with 5% dextrose over 1 hour

Reassess

Oral rehydration with low sodium (ReSoMAL) oral rehydration solution

Measles

Despite an effective vaccine against the virus, more than 20 million people are affected by measles every year, predominantly in parts of Africa and Asia. The majority of deaths occur in low-income countries and in children who are malnourished, particularly with vitamin A deficiency.²⁵

Children with measles present with symptoms which usually appear 10–12 days after infection, including a fever, runny nose and white spots on the inside of the mouth. Several days later a rash appears, starting on the face and neck, gradually spreading downwards.

The most serious complications of measles include blindness, encephalitis (an infection that causes brain swelling), severe diarrhoea with dehydration, and severe respiratory infections such as pneumonia.

Measles is caused by a virus for which there is no specific treatment. Children should be assessed using the ETAT triage tool followed by a thorough ABC assessment, with particular attention to assessment of nutritional status and dehydration, and treated symptomatically with supportive therapy.

Malaria

Malaria is one of the five main causes of death in children under 5 years, with symptoms appearing 7–15 days after the infective mosquito bite. It typically presents with non-specific symptoms such as fever, headache and vomiting.²⁶ It is frequently over-diagnosed and over-treated, yet it is also often treated sub-optimally with incorrect doses of anti-malarial medication prescribed.^{27–32} Over-diagnosis of malaria may result in failure to treat other potential causes of febrile illness.³³ Bedside testing is now available in many countries for malaria parasites.

Children with malaria commonly present with:

- High temperature
- Shock
- Severe anaemia
- Hypoglycaemia

- Jaundice.

In severe cases of cerebral malaria they may also present with:

- Convulsions
- Coma.

The assessment and management of malaria should follow ETAT guidelines, with identification and treatment of emergency signs followed by a thorough ABC assessment. Important additional points to remember for suspected malaria are:

- Treat hypoglycaemia
- Assess conscious level and consider lumbar puncture to rule out meningitis
- Do an early blood film to establish diagnosis
- Treat using local anti-malarial guidelines ensuring accurate dosing
- Consider broad spectrum antibiotics if there is some doubt as to the diagnosis
- Give cautious fluids if there is impaired perfusion or shock, especially if there is anaemia or cerebral impairment.

CONCLUSION

In this article we have looked at the initial assessment and management of the seriously ill child. Key points to consider are rapid initial assessment and triage using the ETAT criteria followed by treatment of emergency signs. This must then be followed by a thorough review using the ABCDE + DEFG approach, commencement of appropriate treatment with frequent reassessment of ABCDE +DEFG. This system will ensure effective and accurate initial management for all seriously ill children.

REFERENCES

1. Murray C, Lopez A. The global burden of disease: comprehensive assessment of mortality and disability from diseases, injuries and risk factors in 1990 and projected to 2020. Geneva: WHO, 1996.
2. UNICEF. The state of the world's children 1998. New York: Oxford University Press, 1998.
3. World Health Organisation. Global Health Observatory http://www.who.int/maternal_child_adolescent/epidemiology/child/en/index.html (accessed 3rd October 2014).
4. Advanced Life Support Group. Advanced paediatric life support—the practical approach, 2nd ed. London: BMJ Publishing Group, 1997.
5. American Academy of Pediatrics and American Heart Association. Textbook of pediatric advanced life support. AHA 1994 revision.
6. World Health Organization, Division of Child Health and Development. Hospital study of the referral care project. Final Report, February 1998.
7. Nolan T, Angos P, Cunha A et al. Quality of hospital care for seriously ill children in less-developed countries. *Lancet* 2001; **357**: 106-10.
8. World Health Organization. Emergency Triage and Assessment (ETAT) Course. http://www.who.int/maternal_child_adolescent/documents/9241546875/en/ (accessed 4th October 2014).
9. Irimu G et al. Performance of Health Workers in the Management of Seriously Sick Children at a Kenyan Tertiary Hospital: Before and after a Training Intervention. *PLoS One* 2012; 7: e39964.
10. Molyneux E. Paediatric emergency care in developing countries. *Lancet* 2001; **357**: 86-7.
11. English M, Wamae A, Nyamai R, Bevins B, Irimu G. Implementing locally appropriate guidelines and training to improve care of serious illness in Kenyan hospitals: a story of scaling-up (and down and left and right). *Arch Dis Child* 2011; **96**: 285-90.
12. Fleming S et al. Normal ranges of heart rate and respiratory rate in children from birth to 18 years of age: a systematic review of observational studies. *Lancet* 2011; **377**: 1011-18.
13. World Health Organisation. Management of severe malnutrition: a manual for physicians and other senior health workers. Geneva: WHO, 1999.
14. Molyneux ME, Taylor TE, Wirima JJ, Borgstein A. Clinical features and prognostic indicators in paediatric cerebral malaria: a study of 131 comatose Malawian children. *Q J Med* 1989; **71**: 441-59.
15. World Health Organization. Children: reducing mortality. <http://www.who.int/mediacentre/factsheets/fs178/en/index.html> (accessed 4th Oct 2014).
16. Maitland K, Kiguli S, Opopka RO et al. Mortality after fluid bolus in African children with severe infection. *NEJM* 2011; **364**: 2483-95.
17. Southall D, Samuels M. Treating the wrong children with fluids will cause harm: response to 'mortality after fluid bolus in African children with severe infection'. *Arch Dis Child* 2011; **96**: 905-6.
18. Duke T. What the African fluid-bolus trial means. *Lancet* 2011; **387**: 1685-87.
19. Myburgh J1, Finfer S. Causes of death after fluid bolus resuscitation: new insights from FEAST. *BMC Med*. 2013 Mar 14; 11: 67
20. Maitland K, George EC, Evans JA et al. Exploring mechanisms of excess mortality with early fluid resuscitation: insights from the FEAST trial. *BMC Med*. 2013; 11: v68.
21. Kiguli S, Akech SO, Mtove G et al. WHO guidelines on fluid resuscitation in children: missing the FEAST data. *BMJ* 2014; 348: f7003 doi: 10.1136/bmj.f7003 (Published 14 January 2014)

22. Jackson AA. Severe malnutrition. In: Warrell DA, Cox TM, Firth JD, Benz EJ, eds. Oxford textbook of medicine, 4th edn, vol 1. Oxford: Oxford University Press, 2003. 1054–1061.
23. Schofield C, Ashworth A. Why have mortality rates for severe malnutrition remained so high? *Bull World Health Organ* 1996; **74**: 223–229.229
24. World Health Organization. Management of the child with a serious infection or malnutrition. Guidelines for care at the first–referral level in developing countries. Geneva: WHO, 2000 http://whqlibdoc.who.int/hq/2000/WHO_FCH_CAH_00.1.pdf (accessed 4th October 2014).
25. World Health Organization. Measles. <http://www.who.int/immunization/topics/measles/en/index.html> (accessed 3rd October 2014).
26. World Health Organization. Malaria <http://www.who.int/immunization/topics/malaria/en/> (accessed 3rd October 2014).
27. Amexo M, Tolhurst R, Barnish G, Bates I. Malaria misdiagnosis: effects on the poor and vulnerable. *Lancet*; 2004; **b364**: 1896-98.
28. Rowe AK, Hamel MJ, Flanders WD, Doutizanga R, Ndoyo MS. Predictors of correct treatment of children with fever seen at outpatient health facilities in the Central African Republic. *Am J Epidemiol* 2000; **151**: 1029-35.
29. Rowe AK, Onikpo F, Lama M, Deming MS. Risk and protective factors for two types of error in the treatment of children with fever at outpatient health facilities in Benin. *Int J Epidemiol* 2003; **32**: 296-303.
30. Zurovac D, Rowe AK, Ochola SA et al. Predictors of the quality of health worker treatment practices for uncomplicated malaria at government health facilities in Kenya. *Int J Epidemiol* 2004; **33**: 1080-1091.
31. Osterholt DM, Rowe AK, Hamel MJ et al. Predictors of treatment error for children with uncomplicated malaria seen as outpatients in Blantyre district, Malawi. *Trop Med Int Health* 2006; **11**: 1147-56.
32. Nshakira N, Kristensen M, Ssali F et al. Appropriate treatment of malaria? Use of antimalarial drugs for children's fevers in district medical units, drug shops and homes in eastern Uganda. *Trop Med Int Health* 2002; **7**: 309-316.
33. Reyburn H, Mbatia R, Drakeley C et al. Overdiagnosis of malaria in patients with severe febrile illness in Tanzania: a prospective study. *BMJ*. 2004; **329**: 1212.

Meningococcal disease in children

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Case history 1

A three-year-old girl presents to her doctor with fever, lethargy and a rash. The rash is initially petechial but spreads rapidly. The doctor makes a presumptive diagnosis of meningococcal disease and gives her intramuscular penicillin and refers her to hospital by ambulance. On arrival she is confused, shocked and has widespread purpura. She receives appropriate resuscitation and emergency treatment in the emergency department and is transferred to the Intensive Care Unit. Meningococci are seen on microscopy of a skin scraping of a purpuric area. She develops multiple organ failure and requires inotropes and ventilation. Three fingers on her left hand become necrotic and require amputation. After 5 days she has recovered sufficiently to leave ICU.

Case history 2

A fifteen-year-old boy presents to hospital with fever, vomiting and lethargy. He has no neck-stiffness, photophobia or rash and is admitted with a diagnosis of a viral infection. Over the next few hours he becomes irritable and drowsy. After a blood culture is taken, he is started on ceftriaxone and intravenous fluids. His level of consciousness continues to decline and he has a seizure on the ward. He is admitted to Intensive Care where he is ventilated but he later dies from raised intracranial pressure.

These two cases represent the opposite ends of the spectrum of meningococcal disease. The first case is an example of meningococcal septicaemia whilst case 2 is an example of meningococcal meningitis. A mixed picture is also very common. It is vital that all doctors that may treat sick children have a good understanding of how to diagnose and treat this condition, as it occurs worldwide and is currently the leading infective cause of death in children in the developed world.

MICROBIOLOGY

Neisseria meningitidis (meningococcus) is a capsulated gram-negative diplococcus. There are more than ten serogroups based on the polysaccharide that makes up their capsule. The commonest serogroups are A, B, C, Y and W-135. They can be further serotyped and subtyped based on proteins in the outer membrane of the bacterium. More complex techniques of enzyme electrophoresis and DNA typing allow the accurate identification of individual strains of individual meningococci to be determined. This is important public health information.

EPIDEMIOLOGY

The disease occurs worldwide but the incidence varies greatly. In the UK the incidence is about 5

cases/100 000 population/year. In sub-Saharan Africa (the "meningitis belt") epidemics occur every 5-10 years with rates of 500 cases/100 000 population/year. In the UK serogroups B and C, and worldwide serogroups A, B and C, are responsible for the majority of cases. Serogroup W-135 has been particularly associated with pilgrims attending the Haj religious festival in Saudi Arabia. The disease is characterised by local clusters or outbreaks and there is a winter predominance in the UK. Nasopharyngeal carriage of the organism occurs in about 10% of the population. Most of these strains are non-pathogenic. The factors associated with pathogenicity are not well understood at present. Risk factors include:

- Age (<1 year of age)
- Overcrowding
- Poverty
- Smoking
- Complement deficiency.

Although the relative risk of developing meningococcal disease following exposure to a case is high (500-1000 times the background rate in the population), only about 1 in 200 contacts will develop the disease.

Summary

Both meningitis and meningococcal septicaemia can present with non-specific initial signs and symptoms but progress rapidly. Prompt diagnosis and treatment are vital. Mortality of those reaching hospital remains 5-10% with a further 10% long term morbidity.

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The epidemiology of this disease may change due to **vaccines** being developed. Purified polysaccharide vaccines have been developed against serogroups A, C, Y and W-135, but they are poorly immunogenic in young children and the immunity is short lived. This is because the immunological response is T-cell independent. These vaccines may be useful for controlling outbreaks and epidemics, but are not suitable for use as part of a primary vaccination program. A conjugated group C vaccine has been developed where the polysaccharide antigen is conjugated to a carrier protein. The immunological response to this is T-cell dependent, which overcomes the problems associated with the purified vaccines and makes it suitable for primary immunisation. In the UK all children receive conjugated meningococcal C vaccine at 3, 4 and 12 months of age. A conjugated group A vaccine has recently been developed and was introduced into African countries in December 2010, with the aim of introducing it into all 25 countries in the African belt by 2016. It has already led to a decrease in the number of confirmed cases in these countries. Group B polysaccharide appears not to be immunogenic.

PATHOPHYSIOLOGY

Development of the disease involves:

- Colonisation of the nasopharynx
- Invasion
- Multiplication.

Both innate and acquired immune mechanisms are responsible for host protection. The resultant disease process may be focal infection (normally meningitis), septicaemia or both. About 60% of cases in Europe have evidence of meningitis and septicaemia, while about 20% have meningitis only and 20% septicaemia only. Endotoxin and other bacterial factors cause a host response that results in much of the damage. This pattern of events is shown in Figure 1. In meningococcal septicaemic shock,

- Endothelial changes cause capillary leak (leakage of fluid into the interstitial space and hypovolaemia) and pathological vasospasm and vasodilatation.
- Intravascular thrombosis causes organ ischaemia and consumptive coagulopathy.
- Generalised endothelial injury activates procoagulant pathways.
- Anticoagulant pathways (protein-C and fibrinolytic) are down-regulated.
- Multiple organ failure is caused by cytokine production and by ischaemia due to intravascular thrombosis and shock.
- Cardiac dysfunction is often an important feature of septic shock due to meningococci.

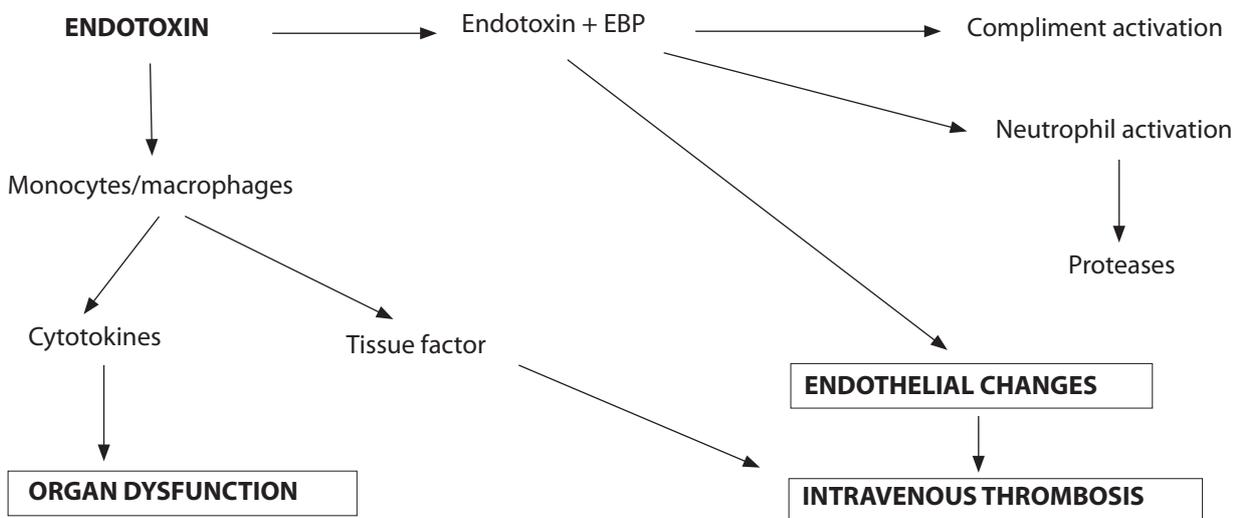
CLINICAL FEATURES

Patients who present early may have very non-specific symptoms and signs. The disease may progress very rapidly, so a high index of suspicion needs to be maintained if the diagnosis is to be made early enough for treatment to be effective. The classical feature of the disease is a petechial or purpuric **rash** (purple rash, which does not fade on pressure), but up to 20% of cases may have no rash or an atypical maculopapular rash. Other infections rarely can produce a similar rash and septicaemia.

Symptoms of **meningitis** include:

- Headache
- Fever
- Vomiting
- Photophobia
- Lethargy or confusion

Figure 1. Pathophysiology of meningococcal disease



EBP = Endotoxin binding protein

- Some patients present with seizures.

Neck stiffness, neurological signs and signs of raised intracranial pressure should be sought on examination. In infants, particularly, the features can be very non-specific; they frequently present with only:

- Irritability
- Refusal to eat
- Drowsiness
- Fever.

Death is usually caused by refractory raised intracranial pressure.

Septicaemia is characterised by:

- Fever
- Rash
- Vomiting
- Headache
- Myalgia (muscle pains)
- Abdominal pain
- Tachycardia
- Cool peripheries
- Hypotension.

Typically the rash spreads rapidly and can lead to widespread necrosis and gangrene of skin and underlying tissues. The rash is a visible sign of the endothelial changes and coagulopathy, which is occurring throughout the body. Death due to septic shock will ensue rapidly if these patients are not resuscitated promptly.

DIAGNOSIS

Because of the need for immediate treatment once the disease is suspected, laboratory tests are not of use in making the initial diagnosis. They may also offer false reassurance since in fulminant infections the white cell count, C-reactive protein and lumbar puncture may all be normal early in the disease. The **initial diagnosis is based on clinical history and examination**. Following the institution of treatment, the diagnosis can be confirmed later by microbiological culture (blood, CSF or skin), antigen detection (PCR, latex agglutination test) or serology. Blood cultures and CSF cultures are more likely to be positive if taken before antibiotics are given.

There have been a number of reports suggesting that major morbidity (particularly death following cerebral herniation) was caused by performing lumbar punctures (LP) in patients with meningitis. Cephalosporins are effective in treating all the common causes of bacterial meningitis. There has thus been a trend not to perform LP in these patients. Some experts believe that too few lumbar punctures are done and this remains a controversial area.¹ Contra-indications to lumbar puncture are:

- GCS <13
- Focal neurological signs
- Raised intracranial pressure
- Recent or prolonged seizures
- Cardiorespiratory compromise
- Coagulopathy
- Infection at the site.

If a positive microbiological diagnosis can be made from a skin scraping, LP is unnecessary. However LP may be useful for the following reasons:

- Gram stain is frequently diagnostic and thus allows a definite diagnosis to be made early.
- It will detect resistance - in some areas pneumococci are resistant to penicillin and cephalosporins.
- Will identify unusual pathogens and allow a positive diagnosis of viral meningitis to be made (enteroviral meningitis can be diagnosed on PCR allowing hospital discharge on no antibiotics).
- Allows public health monitoring of the aetiology of meningitis.
- Allows appropriate prophylaxis to be given to contacts.
- Makes it possible to investigate vaccine failures.

Unless contra-indication exists, patients with suspected meningitis should have a lumbar puncture, but it should be done promptly and should not delay giving the antibiotics by more than thirty minutes.

CT scanning is of no benefit in making the diagnosis of meningitis or in determining whether the intracranial pressure is raised in patients with known meningitis. It should only be used to exclude other causes for focal neurological signs or to investigate complications of meningitis.

TREATMENT

Initial assessment and resuscitation

Early recognition and prompt treatment is vital. If the diagnosis is suspected in the primary care setting the patient should receive intramuscular penicillin or a cephalosporin, if available, and be referred immediately to hospital. In hospital, **assessment and resuscitation of vital functions should occur together**, with problems treated as they are found. Priorities are:

1. Maintaining a patent **airway**. Patients with a decreased level of consciousness due to meningitis or shock may need assistance in maintaining their airway.
2. Supporting **ventilation** as necessary.
 - All patients should receive a high concentration of inspired oxygen. Ventilatory drive may be impaired due to raised intracranial pressure.

- Hypoxia is common due to the capillary leak associated with shock (acute lung injury).
- Intubation and ventilation may be required soon after the patient reaches hospital.

3. Circulation

- Shock is recognised by the presence of an increased heart rate and respiratory rate for age, a prolonged capillary refill time and cool skin and peripheries
- Reduced end organ perfusion will cause a metabolic acidosis (Kussmaul breathing), oliguria (not a sign that can be elicited immediately) and a decreased level of consciousness
- Hypotension is often a very late clinical sign

Treatment of shock requires:

- Stabilisation of the airway and breathing.
- Intravenous or intraosseous access.
- Replacement of circulating blood volume. Give 20ml.kg^{-1} boluses of resuscitation fluid (crystalloid or colloid) and assess the response. As soon as intravenous access is obtained, take blood for culture, biochemical (including glucose) and haematological tests, and give antibiotics (see later).
- Large volumes of fluid may be required, frequently 60ml.kg^{-1} in the first hour. The increased vascular permeability that is associated with septic shock means that fluid will continue to extravasate and these patients may become very oedematous. If more than 40ml.kg^{-1} of resuscitation fluid is required initially, consider intubation and ventilation since pulmonary oedema is likely to develop. Use a tidal volume of $6\text{--}7\text{ml.kg}^{-1}$ and add positive end-expiratory pressure (PEEP).

4. Determine whether major neurological compromise exists:

- Rapid assessment of level of consciousness (AVPU - alert, responds to voice, responds only to pain or unresponsive).
- Examination of the pupils.
- Observation for seizures or abnormal posturing.

Patients with meningitis rather than septicaemia may develop raised intracranial pressure. Look for:

- Fluctuating or decreasing level of consciousness
- Unequal, dilated or poorly reacting pupils
- Focal neurological signs
- Abnormal posturing
- Seizures
- Hypertension accompanied by tachycardia or bradycardia
- Papilloedema is sometimes seen.

It may be difficult to distinguish the central nervous system effects of shock (caused by decreased cerebral perfusion) from those of raised intracranial pressure, especially since raised intracranial pressure can sometimes be associated with abnormal vasoconstriction. Patients with raised intracranial pressure require treatment to optimise cerebral perfusion. Protect their airway by intubation and control their breathing by mechanical ventilation to a normal PaCO_2 . Treat shock aggressively if present.

Patients with isolated meningitis (i.e. no shock) should receive dexamethasone (0.4mg.kg^{-1} BD for 2 days) either with or before the first dose of antibiotic (see later).² Mannitol and frusemide may be used if the intracranial pressure is raised. The patient should be examined for the typical rash but this may not always be present.

Antibiotic therapy

A **third generation cephalosporin** is the drug of choice for suspected meningococcal disease and should be given intravenously for 7 days. Cefotaxime 50mg.kg^{-1} 4 times a day or ceftriaxone 80mg.kg^{-1} as a single daily dose are appropriate. Advantages of these agents over penicillin are:

- Broader spectrum (covering the other common causes of bacterial meningitis)
- Activity against meningococci that are less sensitive to penicillin (due to a different penicillin binding protein) or resistant to penicillin (rarely meningococci can produce β -lactamases)
- Better CSF penetration
- Less CNS toxicity (especially important if renal failure is present)
- They eliminate carriage of the organism, which penicillin does not do.³

Ongoing treatment and Intensive Care

The initial priority of management is the identification and treatment of immediately life threatening problems. These problems (e.g. airway obstruction or shock) should be treated as they are detected, even if the cause for them is not immediately obvious. After the resuscitation has commenced, a focussed **medical history, fuller examination** and the results of special investigations will either confirm the initial diagnosis of meningococcal disease, or allow a differential diagnosis to be made which will determine what further treatment is required.

Other complications that may need treatment include:

- Hypoglycaemia. This is particularly common, causes major morbidity if unrecognised, and is easy to treat. Determine the blood glucose when intravenous access is first obtained
- Hypokalaemia
- Hypomagnesaemia
- Hypocalcaemia
- Anaemia
- Coagulopathy.

Many of these patients will require ongoing **intensive care**. Patients who remain hypotensive following intravenous fluid resuscitation need vasoactive drug administration to counter ventricular dysfunction and vasodilation. A central venous line should be inserted as a route for **inotropic/vasopressor** agents, although their use as a guide for fluid therapy is limited. In very young children a femoral line may be inserted as it is associated with less morbidity than jugular or subclavian lines. Where available an arterial line will be required for cardiovascular monitoring and to facilitate blood sampling. Ventilated patients should have a nasogastric tube and urinary catheter inserted.

The use of inotropes/vasopressors should be guided by clinical assessment and markers of 'global metabolic status':

- Clinical signs
- Arterial blood pH
- Blood lactate
- Base deficit
- Mixed venous oxygen saturation.⁴

Choice of vasoactive drug should be guided by the clinical picture (warm shock and low BP, cold shock with low BP, or cold shock with normal BP) and titrated to achieve an acceptable cardiac output and systemic vascular resistance. The haemodynamic picture can change frequently during the first 48 hours and high doses of drugs may need to be given due to receptor down-regulation. Dopamine, epinephrine, norepinephrine, vasopressin and various vasodilators may all have a place in managing the haemodynamic changes associated with this condition. Children with septic shock, particularly those with meningococcal disease, often die with a high systemic vascular resistance and low cardiac output (compared to adults that tend to have a low systemic vascular resistance that is refractory to therapy). This makes dopamine an appropriate first line agent. High dose epinephrine or norepinephrine are usually required in severe cases.

The skin and limb involvement in meningococcal septicaemia distinguishes it from most other causes of sepsis and can be responsible for major morbidity. Widespread thrombosis and haemorrhagic necrosis of the skin and underlying tissues is called "purpura fulminans". When the thrombosis involves large vessels, infarction and gangrene of the limbs occurs. The combination of ischaemia, necrosis and oedema can cause compartment syndrome. The management of these problems is difficult. It has been suggested that fasciotomies are only indicated in the first 24 hours after onset of purpura fulminans and only for compartment syndrome of the lower limb and where there is no major bleeding diathesis. A combination of clinical assessment, doppler flow studies and compartment pressures should be used as a guide to the decision to perform a fasciotomy. Leave gangrenous limbs to demarcate if possible; amputation should be an elective procedure.⁵

There is now evidence from randomised controlled trials of adults and children with septic shock that low dose hydrocortisone

treatment ($1\text{mg}\cdot\text{kg}^{-1}$ 6 hourly IV) decreases 28 day mortality.⁶ In paediatric septic shock adrenal insufficiency has been shown to be associated with an increased vasopressor requirement and duration of shock. Also, in paediatric meningococcal septicaemia a low serum cortisol and a high ACTH has been shown to be associated with severe disease or death. As a result, many paediatric intensivists give hydrocortisone in a replacement dose ($1\text{mg}\cdot\text{kg}^{-1}$ 6 hourly) to patients with meningococcal septicaemia either on the basis of an ACTH stimulation test or to all those that have shock requiring high dose inotropic support. If the patient is already receiving dexamethasone, further steroid supplementation is not required.

Coagulopathy

Deranged clotting is commonly seen as part of the septic process and blood products are often required to correct this.

OUTCOME

The mortality of all patients admitted to hospital is about 5-10%, but the mortality of those admitted to intensive care varies from 5-35%. The mortality is greater in those patients who have septicaemia. Approximately 10% of patients will have long term morbidity due to neurological complications (especially deafness) or amputations. Long-term problems related to renal or myocardial function are less common.

SECONDARY PREVENTION

Patients remain infectious for 24 hours after receiving a cephalosporin and should be isolated during this period. Household contacts and carers exposed to oropharyngeal secretions should receive chemoprophylaxis:

- Ciprofloxacin as a single oral dose (>12 years 500mg, 5-12 years 250mg, <5 years $30\text{mg}\cdot\text{kg}^{-1}$ up to 125mg); or
- Rifampicin twice daily for 2 days (> 12 years 600mg, 1-12 years $10\text{mg}\cdot\text{kg}^{-1}$, < 12 months $5\text{mg}\cdot\text{kg}^{-1}$); or
- A single IM injection of ceftriaxone (<12 years 125mg and >12 years 250mg)
- If the infection is due to serogroup C, contacts should also receive the conjugated group C vaccine
- If infection is due to serogroup A, W-135 or Y, contacts should also receive the quadrivalent conjugate vaccine.

Further information on meningitis and meningococcal disease is available at www.meningitis.org

REFERENCES

- Law R. Meningococcal disease in children. *Update in Anaesthesia* (2007)**22**:26-30. Available from: http://www.wfsahq.org/images/wfsa-documents/updates_in_english/Update_22_2007.pdf
1. Kneen R et al. The role of lumbar puncture in suspected CNS infection - a disappearing skill? *Arch Dis Child* 2002; **87**: 181-3
 2. McIntyre PB et al. Dexamethasone as adjunctive therapy in bacterial meningitis: A meta-analysis of randomised clinical trials since 1998. *JAMA* 1997; **278**: 925-31.

3. Wall RA. Meningococcal disease: treatment and prevention. *Annals of Medicine* 2002; **34**: 624 - 34.
4. Rivers E et al. Early goal directed therapy in the treatment of severe sepsis and septic shock. *NEJM* 2001; **345**: 1368 - 77.
5. Davies MS et al. The orthopaedic management of peripheral ischaemia in meningococcal septicaemia in children. *Journal of Bone and Joint Surgery* 2000; 82B No.3: 383 - 86
6. Annane D et al. Corticosteroids for treating severe sepsis and septic shock. *Cochrane Database of Systemic Reviews* 2004, Issue 1. Art. No: CD002243. DOI: 10.1002/14651858.CD002243.pub2.

Intraosseous infusion

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INTRODUCTION

The technique of intraosseous infusion was first described in humans in 1934 and it became increasingly popular in the 1940s. In recent years it has regained popularity in both adult and paediatric resuscitation. Unfortunately many doctors do not know this technique or do not employ it.

However, intraosseous infusion is one of the quickest ways to establish access for the rapid infusion of fluids, drugs and blood products in emergency situations as well as for resuscitation. In many countries, where children are the victims of war trauma, road traffic accidents or severe dehydration and need good circulatory access, this technique can be life-saving. In these situations peripheral venous access can be difficult to obtain and alternatives such as central venous access can be difficult and/or dangerous.

INTRODUCTION TO THE TECHNIQUE

The marrow cavity is in continuity with the venous circulation and can therefore be used to infuse fluids and drugs, and to take blood samples for crossmatch, for example. The procedure must be performed under sterile conditions to avoid causing osteomyelitis. It is also recommended to limit the duration of the use of intraosseous infusion to a few hours until intravenous access is achieved. It is thus a temporary emergency measure. In experienced hands intraosseous access can be established within 1 minute.

It has been shown that the onset of action and drug levels during cardiopulmonary resuscitation using the intraosseous route are similar to those given intravenously.

INDICATIONS

Placement of an intraosseous needle is indicated when vascular access is needed in life-threatening situations in babies, infants and children under the age of six years. It is indicated when attempts at venous access fail (three attempts or 90 seconds) or in cases where it is likely to fail and speed is of the essence. Although principally advocated for use in young children, it has been successfully used in older children where the iliac crest may also be used.

CONTRA-INDICATIONS

- Femoral fracture on the ipsilateral side
- Do not use fractured bones
- Do not use bones with osteomyelitis.

EQUIPMENT

1. Skin disinfectant
2. Local anaesthetic
3. 5ml syringe
4. 50ml syringe
5. Intraosseous infusion needle or Jamshidi bone marrow needle. There are different needle sizes; 14, 16 and 18G. The 14 and 16G are usually used for children older than 18 months. However any size can be used for all ages.

It is possible but not ideal to use a 16 – 20G butterfly needle, spinal needle or even hypodermic needle. The chance that the needle gets blocked with bone marrow however, is much increased when not using a needle with a trochar.

SITE

The best site to use is the flat anteromedial aspect of the tibia. The anterior aspect of the femur and the superior iliac crest can also be used. The tibia is preferred since the anteromedial aspect of the bone lies just under the skin and can easily be identified. Avoid bones with osteomyelitis or fractures and do not use the tibia if the femur is fractured on the same side.

TECHNIQUE

1. Palpate the tibial tuberosity. The site for cannulation lies 1 - 3cm below this tuberosity on the anteromedial surface of the tibia.
2. Use sterile gloves and an aseptic technique and a sterile needle.
3. Clean the skin. Placing a bone marrow needle without using a sterile technique obviously increases the chance of osteomyelitis and cellulitis.

Summary

Intraosseous infusion is a temporary emergency measure

Indicated in life-threatening situations when intravenous access fails (3 attempts or >90 seconds)

Use the anteromedial aspect of the tibia

Insert pointing caudal to avoid the epiphyseal growth plate

Use an aseptic technique
Crystalloids, colloids, blood products and drugs can be infused

Remove as soon as the child has been resuscitated and intravenous access has been established.

The authors are members of the international medical aid organisation Médecins Sans Frontières (MSF).

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4. Inject a small amount of local anaesthetic in the skin and continue to infiltrate down to the periosteum. When the child is unconscious it is not necessary to use local infiltration.
5. Flex the knee and put a sandbag as support behind the knee.
6. Hold the limb firmly above the site of insertion, usually at the level of the knee. Avoid putting your hand behind the site of insertion to avoid accidentally injuring your own hand.
7. Insert the intraosseous needle at 90 degrees to the skin (perpendicular) and slightly caudal (towards the foot) to avoid the epiphyseal growth plate.

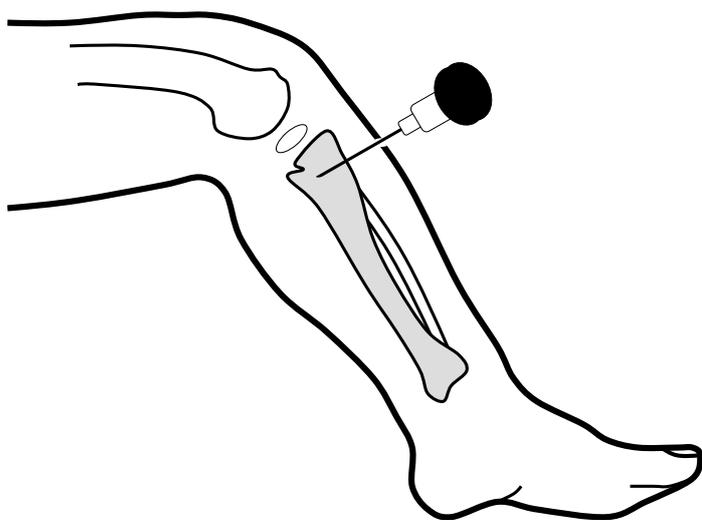


Figure 1. Insertion of intraosseous needle in the right tibia

8. Advance the needle using a drilling motion until a 'give' is felt – this occurs when the needle penetrates the cortex of the bone. Stop inserting further.
9. Remove the trochar. Confirm correct position by aspirating blood using the 5ml syringe. If no blood can be aspirated the needle may be blocked with marrow. To unblock the needle, slowly syringe in 10ml of saline. Check that the limb does not swell up and that there is no increase in resistance.
10. If the tests are unsuccessful remove the needle and try the other leg.
11. Secure the needle in place with sterile gauze and strapping.

Correct placement is further confirmed by the following:

- A sudden loss of resistance on entering the marrow cavity (less obvious in infants who have soft bones).
- The needle remains upright without support (because infants have softer bones, the needle will not stand as firmly upright as in older children).
- Fluid flows freely through the needle without swelling of the subcutaneous tissue.

COMPLICATIONS

Important complications are tibial fracture especially in neonates, compartment syndrome, osteomyelitis and skin necrosis. When an aseptic technique is used, the incidence of osteomyelitis is less than 1%. Microscopic pulmonary fat and marrow emboli do not seem to be a clinical problem.

Provided the correct technique is employed there does not seem to be any long-term effects on bone growth.

INFUSION

Fluid can be infused under gentle pressure, manually by using a 50ml syringe or by inflating a blood-pressure cuff around the infusion bag. Crystalloids, blood products and drugs can be infused using this technique.

The intraosseous route should be replaced as soon as a normal vein can be cannulated and certainly within a few hours. The longer the period of use the greater the risk of complications.

CONCLUSION

In emergencies rapid intravenous access in children may be difficult to achieve. Intraosseous access is an easy, safe and life-saving alternative.

Editors' note

Since this article was first published, battery-powered insertion devices for intraosseous needles became available. Insertion technique is as described above for the hand-held intraosseous needles. These new devices are more expensive; some find them easier to insert and hence more reliable.

REFERENCES

- Vreede E, Bulatovic A, Rosseel P, Lassalle X. Intraosseous infusion. *Update in Anaesthesia* (2000) **12**:38-40. Available from: http://www.wfsahq.org/components/com_virtual_library/media/88e559324c3f259ce183b8f9fbf6047c-Intraosseous-Infusion-Update-12-2000-.pdf
1. Chameides L, Hazinski MF, Eds. Textbook of Pediatric Advanced Life Support. 1994; 5-5 to 5-7
 2. Brickman KR, Krupp K, Rega P, Alexander J, Guinness M. Typing and screening of blood from intraosseous access. *Annals of Emergency Medicine* 1992; **21**: 414-7.
 3. Sawyer RW, Bodai BI, Blaisdell FW, McCourt MM. The current status of intraosseous infusion. *Journal of American College of Surgeons* 1994; **179**: 353-60.
 4. Claudet I, Fries F, Bloom MC, Lelong-Tissier MC. Etude retrospective de 32 cas de perfusion intraosseuse. *Archives of Paediatrics* 1999; **6**: 566-9.
 5. Nafiu OO, Olumese PE, Gbadegesin RA, Osinusi K. Intraosseous infusion in an emergency situation: a case report. *Annals of Tropical Paediatrics* 1997; **17**: 175-7.

The child with malaria

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Summary

Malaria is a multi-system disease that can coexist with other infections and conditions, including those that may require surgery. Intensive care may be necessary.

Malaria can vary from an insidious febrile illness to an acute life-threatening disease. Rapid deterioration is much more likely in children. If malaria is suspected, it should be investigated rapidly and treated appropriately. WHO and national diagnostic and treatment guidelines should be followed. Children should be observed closely for the development of complications. Anaesthesia and surgery should be avoided in the child with malaria if at all possible, but, if it is necessary, the multi-system nature of the disease should be considered.

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MALARIA: CAUSE, TRANSMISSION & EPIDEMIOLOGY

Malaria is a life-threatening disease caused by four protozoan parasite species of the genus *Plasmodium* infecting humans: *P. falciparum*, *P. vivax*, *P. ovale*, and *P. malariae*. Co-infection with more than one species is possible. A 5th species *P. knowlesi*, which primarily affects primates, has also recently been found to infect humans. *P. falciparum* is the most deadly species, and *P. falciparum* and *P. vivax* the most common. *P. vivax* and *P. ovale* both lead to a dormant liver form (hypnozoites) that may cause relapses months or years later.

The parasites are transmitted through the bite of a vector, the infected female *Anopheles* mosquito. There are around 20 *Anopheles* species across the world. The intensity of malaria transmission depends on factors relating to the parasite, vector, host and environment. Mosquitoes exhibit different breeding and biting preferences, with the important vectors tending to bite at night. High humidity and warmer temperatures (between 20°C and 30°C) favour transmission of malaria due to increase in mosquito numbers. Consequently, the disease is often seasonal, relating to rainfall patterns. Some areas, with constant temperatures and humidity, have steady parasite rates. Climate changes may lead to alterations in the pattern of disease.

40% of the global population reside in malaria endemic areas. Most cases of malaria are found in sub-Saharan Africa, Asia and Latin America; a few cases occur in the Middle East and in some areas of Europe. Increased international air travel has also introduced malaria into malaria free zones, resulting in disease misdiagnosis. *The World Malaria Report 2011* estimated 216 (uncertainty range of 149-274) million cases of malaria with 655,000 deaths (uncertainty range 537-907,000). 80% of the deaths occur in African children, most of them being in the under five age range. Since 2000, deaths from malaria have been reduced by >25 % globally, and by 33% in Africa.

Partial immunity to the disease can develop over years of exposure; consequently the majority of deaths

occur in children. The risk is greater across all ages when natural immunity is reduced:

- Limited previous exposure
- Pregnancy
- Severe concomitant illness
- Surgery.

Malaria in pregnancy affects both the mother and the foetus, which can lead to loss of the pregnancy or low birth weight.

Infants can be protected by maternal antibodies and by foetal haemoglobin, up to around 6 months of age. A behavioural tendency to cover infants may also be protective. Some inherited abnormalities of red cells can be protective against malaria, for instance, the sickle cell trait and Melanesian ovalocytosis, a genetic polymorphism associated with mild haemolytic anaemia, common in South East Asia.

Malaria is both preventable and curable. Where prevention and control measures have been applied aggressively, the malaria burden has been effectively reduced.

PREVENTION

Methods used to avoid disease transmission include prevention of mosquito bites using the following:

- Insecticide treated nets (ITNs)
- Use of mosquito repellents
- Indoor residual spraying with insecticides
- Maximum coverage clothing
- Reduction of mosquito breeding grounds by drainage of stagnant water and clearing of bushes.

Chemoprophylaxis is required for

- High-risk populations, such as travellers to malaria endemic regions
- Intermittent preventive treatment in pregnancy
- Infants in high transmission areas (infants receive 3 doses of sulfadoxine –pyrimethamine alongside routine vaccines).

Seasonal malaria chemoprevention was recommended in 2012 by the World Health Organisation (WHO) in areas of the Sahel sub-region of Africa.

Vaccinations against malaria are currently being evaluated in clinical trials, but there is, as yet, no licensed vaccine. One such study, for a vaccine against *P. falciparum*, is expected to finalise results towards the end of 2014, which will subsequently lead to a review by the WHO.

Anti-malarial drug treatment and drug resistance

Chloroquine: forms complexes with haem molecules in haemoglobin, interfering with haem polymerisation. This is effective at preventing formation of intraerythrocytic trophozoites. Resistance has occurred relating to genetic mutations in the transporter molecule, the '*P. falciparum* chloroquine resistance transporter' (PfCRT).

Doxycycline: binds to ribosomes to inhibit parasite protein synthesis.

Sulfadoxine-Pyrimethamine combination: interferes with the folate pathway and therefore parasitic nucleic acid synthesis through inhibition of dihydrofolate reductase (DHFR). Sulfadoxine inhibits parasite dihydropteroate synthase (DHPS). Pyrimethamine inhibits parasite (DHFR). Resistance due to point mutations in the DHPS has been found.

Atovaquone-Proguanil combination: affects the function of the parasite mitochondria via inhibition of the electron transport chain. Resistance to Atovaquone and combination therapy has been described due to substitution mutations. DHFR point mutation has been found in Proguanil resistance.

Artemisinin derivatives: the mechanism of action is not clear, but may relate to peroxide bond and production of oxide radicals, which destroy the parasite, or to inhibition of cellular redox cycling. Monotherapy is discouraged for fear of resistance. Consequently, they are usually used with other classes of anti-malarials, referred to as 'Artemisinin based Combination Therapy' (ACT).

Mefloquine, quinine, quinidine: bind with haem molecules, leading to the creation of parasite-toxic complexes. Mutations in the P-glycoprotein homolog-1 gene *pfmdr-1* and PfCRT have been identified.

Resistance to antimalarials remains a concern. Combination therapy is preferred in order to prevent the development of resistant parasites. Routine monitoring of antimalarial resistance is essential. Resistance of mosquitoes to insecticides has also emerged in some countries, although in most areas they remain an effective prevention tool.

Widespread resistance to chloroquine and the sulfadoxine-pyrimethamine (SP) combination was identified in the 1970-1980s. Resistance to artemisinin derivatives was reported around the Cambodia/Thailand border in 2009. There has also been documented resistance to quinine in Africa and concerns over reduced efficacy of quinine in Southeast Asia.

HISTORY, CLINICAL FEATURES AND PARASITOLOGICAL DIAGNOSIS

History and clinical presentation

Children with malaria may present acutely unwell, or with a more

indolent and asymptomatic picture. Malaria can be classified as simple/uncomplicated or severe/complicated.

Uncomplicated malaria is defined by the WHO as 'symptomatic infection with malaria parasitaemia without signs of severity and/or evidence of vital organ dysfunction'. Typical presentation of uncomplicated malaria includes a febrile illness with headache, night sweats, weakness, myalgia, arthralgia, diarrhoea, abdominal cramps and vomiting. Gastrointestinal symptoms are more common in children.

The acute febrile illness is sometimes associated with classic cyclical 'paroxysms', each lasting several hours. Paroxysms consist of chills and rigors, followed by fever spikes, then profuse sweating and, finally, extreme exhaustion. They occur in regular cycles every 48 or 72 hours, depending on the *P. lasmodium* species, and correspond with schizont rupture. However, these patterns may vary, and fever may be absent. Lack of such features, particularly with *P. falciparum*, should not delay diagnosis or treatment.

Complicated malaria occurs with falciparum malaria and is due to significant multisystem involvement. Presentation may include extreme weakness, confusion or drowsiness. WHO diagnostic features and manifestations of complicated malaria include:

- Cerebral malaria
- Generalised convulsions
- Hyperparasitaemia
- Hyperpyrexia
- Prostration
- Severe anaemia
- Hypoglycaemia
- Acute renal failure
- Acute pulmonary oedema
- Fluid and electrolyte abnormalities
- Metabolic acidosis with respiratory distress
- Shock
- Haemoglobinuria
- Abnormal bleeding
- Jaundice.

Falciparum malaria can be much more acute and severe compared to malaria caused by the other species and carries the greatest mortality. *P. vivax* may also be fatal.

The severity of *falciparum* malaria relates to the ability of the parasite to sequester in the microvasculature. Severe illness may be due to delayed or inadequate treatment and can occur very rapidly in children and in visitors from non-endemic areas. Rapid recognition and treatment is crucial and influences outcome. There should be a high index of suspicion of malaria in both endemic and non-endemic areas. It is important to elicit a travel history to at risk areas, as well as a history of exposure to infected blood through transfusion.

Table 1. Differences between severe malaria in adults and in children. These features will vary depending on the region and levels of immunity in the population

Signs or symptoms	Adults	Children
Duration of illness before severe features	Long (5-7 days)	Short (1-2 days)
Anaemia	Common	Very common
Convulsions	Common	Very Common
Pre-treatment hypoglycemia	Less common	Common
Metabolic acidosis	Less common	Common
History of cough	Uncommon	Common
Cerebral malaria	Common	Very common
Jaundice	Very common	Uncommon
Renal failure	Common	Uncommon
Pulmonary oedema, Acute Respiratory Distress Syndrome (ARDS)	More common	Rare
CSF pressures	Usually normal	Usually raised
Resolution of coma	Longer (2-4 days)	Shorter (1-2 days)
Bleeding/clotting disturbances	Up to 10%	Rare
Abnormality of brainstem reflexes (e.g. oculovestibular, oculocervical)	Rare	More common

Signs and symptoms can be non-specific and so other diagnoses must also be considered. Differential diagnoses include:

- Meningitis
- Influenza
- Typhoid and paratyphoid enteric fever
- Dengue fever
- Hepatitis
- Acute schistosomiasis
- Leptospirosis
- African tick fever
- East African trypanosomiasis
- Yellow fever
- Viral encephalitis.

For primary attacks, the incubation period may last 8-25 days, but can be longer. This relates to the patient's immune status, the *Plasmodium* strain, the sporozoite load, and chemoprophylaxis use. Relapses can cause delayed presentation months to years later and are due to dormant hypnozoites.

The features found in children may differ to those found in adults (see Table 1).

Parasitology

Diagnosis is based on clinical suspicion and on parasite detection in blood (parasitological diagnosis). Current WHO advice recommends rapid parasite diagnostic testing before treatment in suspected cases. Where such testing is not possible, or delayed, treatment can still be considered. Parasitological testing includes either light microscopy or rapid diagnostic tests (RDT).

Microscopy identifies the species, parasite density and parasite stage. Giemsa stained thick and thin films are the accepted standard for diagnosis, but require experienced personnel.

Malarial parasitaemia may be reported as:

- the percentage of parasite infected red blood cells, or
- the number of parasites per microlitre of blood.

The higher the parasite density, the greater the risk of developing severe malaria. The stage of the parasite in peripheral blood also influences prognosis.

RDTs detect parasite specific antigen. They may not identify low level infections and accuracy will depend on the manufacturer. They are useful if microscopy skills are not available or well developed.

Antimalarial treatment should be reserved for test positive cases. Occasionally the film can be negative when intense tissue sequestration has occurred. False negative cases are also more likely in recent artemisinin-derivative use. Rarely, treatment may therefore be considered in test negative cases where severity of suspected disease is significant. Differential diagnoses must be remembered.

Blood smears should be repeated 24-48 hours after initiation of treatment to monitor efficacy of the drugs used. A change in medication may be required if parasites have not been cleared.

Where clinically indicated, other laboratory tests include full blood count, clotting studies, renal function test, liver function test, blood glucose measurement, chest X-ray and lumbar puncture. Children with complicated malaria may be profoundly anaemic and hypoglycaemic. Hyponatraemia is common.

TREATMENT

Delayed diagnosis and treatment leads to increased morbidity and mortality. This may be due to

- Low index of suspicion

- Unclear history
- Failure to request blood films
- False negative results.

Treatment includes targeted antimalarial therapy as well as supportive therapy.

Drug therapy

WHO recommends use of ACTs for uncomplicated *P. falciparum*. Initial treatment of *P. falciparum* malaria should be started according to national and/or local guidelines. National malaria treatment policies should be in place, and should be assessed by in vivo monitoring for therapeutic efficacy to ensure the correct antimalarial regimen is in use. Treatment depends on local resistance patterns and species of plasmodium. Uncomplicated malaria can be treated with oral medication and on an outpatient basis.

ACTs are commonly used in Africa and South East Asia for uncomplicated malaria due to multi-drug resistant *P. falciparum*. Combination with another drug is necessary to reduce the risk of recrudescence and resistance. Combinations will depend on patterns of resistance to the partner drug.

In severe malaria, treatment should commence with one of the following parenteral drugs: artesunate IV or IM/ quinine IV or IM / artemether IM. If definitive treatment is delayed, pre-referral treatment may include rectal artesunate. Parenteral antimalarials should be given for 24 hours. Once this is complete, and oral therapy can be tolerated, antimalarial cover should be continued with either 1) an ACT, or 2) artesunate with clindamycin or 3) quinine with clindamycin. Refer to the latest online WHO guidelines on treatment of malaria.

Adult studies comparing quinine to artemisinin derivatives (artesunate, artemether and artemortil) show superiority of the artemesinins. This relates to the artemesinins' wider range of action in the lifecycle of the parasite and faster clearance of the parasite, alongside a reduced incidence of hypoglycaemia. Neurological outcomes are not felt to differ between artesunate and quinine use.

Supportive therapy

Monitoring and supportive therapy are key to the management of the child with malaria. This includes regular measurement of vital signs, level of consciousness, urine output, blood sugars and oxygen saturation, alongside intensive nursing care.

Fluid resuscitation is often needed. There is a risk of pulmonary overload, although it is rare in children. The requirement for respiratory support ranges from oxygen supplementation to full ventilatory support. Renal failure may require haemodialysis. The nature of multi organ involvement will determine the support required.

Pharmacological adjuncts may include antipyretics, antibiotics, anticonvulsants and antiemetics.

ANAESTHETIC CONSIDERATIONS – THE CHILD WITH MALARIA PRESENTING FOR SURGERY

If a child with malaria presents for surgery, you must assess the urgency of the surgery. Surgery in the presence of acute malaria is

associated with increased morbidity and mortality, both intra- and post-operatively. Where possible, delay surgery to allow for time to respond to anti-malarial treatment. The following precautions should be taken if surgery cannot be delayed:

- Preoperative assessment should include routine history and examination, specific to the child, and with an emphasis on the multi-system effects associated with malaria. The anaesthetic plan will depend on which systems are affected. Determine features and severity of the malaria should be determined, assisted by a full examination. A complete set of observations is required, including temperature and blood sugar measurement.
- Preoperative assessment should include the level of consciousness, with documentation of the GCS/Blantyre Coma Scale and identification of any features of cerebral malaria (see below). There is an increased risk of deterioration post operatively if the child has signs of CNS involvement preoperatively. Place a nasogastric tube if there is a reduced level of consciousness.
- Avoid premedication with sedative drugs in complicated malaria, to prevent confusion between the drug's sedative effects and clinical deterioration, as well as to avoid the risk of airway compromise.
- Aim for low normal PaCO₂ and good oxygenation. Reduced respiratory effort may lead to raised PaCO₂ and falling PaO₂, risking cerebral vasodilatation and a raised ICP. The airway should be secured and controlled ventilation used to prevent a rise in intracranial pressure (ICP). Avoid drugs that lead to an increase in ICP such as halothane and ketamine and prevent hypertension at intubation and extubation. Vigilance is required to identify convulsions under anaesthesia: use signs such as hypertension, tachycardia and pupil changes.
- Consider atracurium or cis-atracurium due to its reliance on Hoffman degradation, although renal failure is less common in children compared to adults. Avoid halothane if hepatic dysfunction is present. If hyperkalaemia is present, avoid suxamethonium. Avoid vecuronium and pancuronium due to delayed clearance. Quinine will enhance the effect of neuromuscular blockade.
- Transfusion requirements and the use of invasive monitoring will depend on factors such as proposed surgery and expected blood loss. Beware of the possibility of a low platelet count and the presence of coagulopathy when considering regional techniques such as a caudal block or spinal anaesthesia.
- Intraoperative vigilance with blood glucose monitoring and treatment is vital.
- Post-operative assessment of consciousness is important - patients should not be returned to the ward if they are not awake and alert. If there is deterioration in the GCS/BCS, hypoglycaemia, seizures or the child is post-ictal, consider worsening cerebral malaria in addition to anaesthetic causes. The child should be monitored carefully postoperatively, ideally in a high dependency area. Anti-malarial treatment must be continued postoperatively.

MULTI-ORGAN INVOLVEMENT, ICU MANAGEMENT

Malaria is a multi-system disease that can coexist with other infections

Table 2. *The Blantyre Coma Scale*

Blantyre Coma Scale	
	Score
Best MOTOR response	
Localises to painful stimulus	2
Withdraws limb from painful stimulus	1
No response or inappropriate response	0
Best VERBAL response	
Cries appropriately with painful stimulus, or, if verbal, speaks	2
Moan or abnormal cry with painful stimulus	1
No vocal response to painful stimulus	0
EYE Movements	
Watches or follows (e.g. mother's face)	1
Fails to watch or follow	0

and conditions, including those that may require surgery. Intensive care may be necessary.

Remember, a child may deteriorate rapidly, particularly if hyperparasitaemia is present. Severe anaemia (Hb <5g.dl⁻¹), respiratory distress due to metabolic acidosis and cerebral malaria are seen more frequently in children.

Laboratory and diagnostic tests should include: full blood count, liver function tests, urea, creatinine and electrolytes, clotting profile, blood type and crossmatch where appropriate (depending on FBC and coagulation findings). Pulse oximetry is helpful. Arterial blood gas, ECG and chest Xray may also be indicated.

Criteria for intensive care admission include:

- Presence of immediate life threatening complications such as coagulopathy or end organ failure
- Presence of signs or symptoms of cerebral malaria
- Non-immune patients with *P. Falciparum* parasitemia >2% or semi-immune patients with *P. Falciparum* parasitemia >5%
- Presence of any other severe complications of malaria.

Cerebral malaria

Cerebral malaria has been defined by the WHO as:

- 'Severe *P. falciparum* malaria with cerebral manifestations, usually including coma (Glasgow Coma Scale <11, Blantyre Coma Scale <3)' (see below)
- 'Malaria with coma persisting for more than 30 minutes after a seizure'.

Other causes for reduced cerebral function should be sought and excluded.

Cerebral malaria is more common in children and non-immune adults. Mortality can be as high as 40% in children, who are also at a greater risk of developing neurological sequelae (10%). Such sequelae include hemiparesis, cerebellar ataxia, cortical blindness, hypotonia, mental retardation and cerebral palsy.

Pathogenesis is thought to include sequestration of parasitised erythrocytes in cerebral microvasculature with associated inflammatory responses. Inducible nitric oxide production is also thought to play a role through inhibition of neurotransmission. There is reduced oxygen and glucose delivery with raised temperature, hypoglycaemia and metabolic acidosis exacerbating the effects.

In children, febrile convulsions may occur, with a post-ictal state lasting several hours. Hyperpyrexia and hypoglycaemia should be excluded as causes of both coma and convulsions.

The optic fundi should be examined. Meningitis should be considered in the differential diagnosis. In cerebral malaria, Kernig's sign is negative (if positive, the child is unable to straighten the leg whilst lying down with the hip flexed – a sign of meningism e.g. due to meningitis). Neck stiffness, photophobia and focal neurology are rare in cerebral malaria. However, cerebral malaria may present with coma, convulsions or posturing. Retinal haemorrhages may be present in 15%. CSF pressure at lumbar puncture may be elevated in children, but is often normal in adults. Cerebrospinal fluid (CSF) is clear with < 10 white blood cells per microlitre but protein is often slightly raised. Computed tomography scans are usually normal.

In older children and adults the Glasgow Coma Scale (GCS) is used to measure the level of consciousness. In younger children either a modified GCS or the Blantyre Coma Scale (BCS) can be used (see Table 2).

Convulsions should be treated with intravenous or rectal diazepam, or intramuscular or rectal paraldehyde.

Patients require meticulous nursing care. The level of consciousness must be regularly monitored alongside temperature, blood pressure, heart rate, and respiratory rate. A urinary catheter is required, with strict input/output recordings. There is currently no role for drugs to reduce cerebral oedema. Respiratory and ventilatory support may be required.

Respiratory distress

There may be several processes present leading to respiratory distress including:

- Pulmonary oedema
- Respiratory compensation for metabolic disturbance, particularly acidosis
- Superadded chest infections
- Severe anaemia.

Non-cardiogenic pulmonary oedema involves sequestration of erythrocytes in the lungs and associated inflammatory responses in the pulmonary vasculature, alongside increased capillary permeability. Mild ARDS may develop and can progress to be severe. Patients can appear to otherwise be improving clinically at the time it develops. Excessive fluid administration, renal failure and hypoalbuminaemia can contribute. Pulmonary oedema can occur in both *falciparum* and *vivax* malaria.

Abnormal breathing patterns can be due to effects on the respiratory centre. Patients may have a superadded chest infection due to immune suppression.

Metabolic acidosis is more common in children than in adults. Confirm with arterial blood gas results where possible. Consider and treat bacterial infection and the impact of a reduced level of consciousness.

Although there is a risk of pulmonary oedema and ARDS in children, this is much less common than in adults. Care is needed with fluid therapy. In anaemic children, dyspnoea is more commonly related to acidosis and hypovolaemia, which therefore needs urgent correction. Ventilatory support may become necessary. Increased FiO_2 and positive end expiratory pressure may be required. In the event of fluid overload, intravenous furosemide can be used.

Circulatory collapse

Cardiovascular collapse in malaria may be due to:

- Secondary bacterial infection
- Metabolic acidosis
- Dehydration
- Bleeding, including a ruptured spleen
- Pulmonary oedema.

Often the bacterial infection is Gram-negative sepsis. Seek possible infection sites, including respiratory tract, urinary tract, meningitis and intravenous lines. Correct hypovolaemia and commence broad-spectrum antibiotics, ideally after blood cultures are sent. Myocardial function is often well preserved, however there is potential for impaired myocardial function and a preoperative echocardiogram may be advised.

Preoperative assessment of hydration is important, with identification and treatment of hypovolaemia, as well as sepsis and shock. Consider fluid therapy, possible blood transfusion and potential inotropes. Children with severe anaemia may present with tachycardia and dyspnoea.

Haematological disturbances

All patients with malaria are prone to anaemia. Children and non-immune patients with high parasite loads are at the greatest risk. The anaemia can be severe, with a haemoglobin $<5\text{g.dL}^{-1}$. Causes are multifactorial and relate to:

- Haemolysis
- Removal of both parasitized and non-parasitized erythrocytes in the spleen
- Impaired bone marrow function
- Reduced erythropoietin production and response to erythropoietin
- Nutrition
- Infective causes (e.g. hookworm).

Thrombocytopenia is very common in falciparum malaria, often occurring in the absence of other clotting dysfunction. It relates to an increase in splenic clearance of platelets. Disseminated intravascular coagulation (DIC) can occur and may present as bleeding gums, epistaxis, petechiae, haematemeses, and malaena. DIC and significant bleeding occurs in $<10\%$, with the greatest risk among non-immune patients.

Determine the degree of anaemia alongside the clinical picture and consider transfusion if the haematocrit is $<25\%$, or when hypovolaemic shock is present. Transfuse whole blood (20ml.kg^{-1}) or packed cells (10ml.kg^{-1}) as per hospital guidelines. Transfusion should be carried out cautiously due to the risk of pulmonary oedema. Children with a hyperdynamic circulation may need intravenous furosemide for transfusions. Platelet and fresh frozen plasma transfusions may be required in the presence of coagulopathy.

Hypoglycaemia

Hypoglycaemia is common in severe malaria. Pregnant women and children are at a greater risk, especially neonates and infants. It should be suspected in all those with a reduced conscious level, and may present with coma or convulsions. Regular blood sugar monitoring is essential and hypoglycaemia must be appropriately treated and observed. Hypoglycaemia can be due to:

- Increased demand (anaerobic glycolysis, febrile illness and demand from parasites)
- Failed hepatic glycogenolysis and gluconeogenesis
- Quinine-stimulated pancreatic beta-cell insulin secretion.

Hypoglycemia contributes to central nervous system dysfunction and associated neurological deficits in survivors of cerebral malaria.

Fluid and electrolyte disturbance, metabolic acidosis

There is often evidence of hypovolaemia and dehydration. Metabolic acidosis can occur with severe illness, hypoglycaemia, hyperparasitaemia, or renal failure. Lactic acidosis is mainly due to reduced oxygen delivery to tissues caused by hypovolaemia, sequestration, and anaemia. Contributing factors include parasite anaerobic glycolysis, impaired hepatic and renal function with reduced lactate clearance and cytokine release. In children with severe malaria, lactate level $>5\text{mmol.L}^{-1}$ is a major predictor of death. Hyponatraemia is a common finding. Children with acute renal tubular dysfunction may have raised potassium levels.

Hyperpyrexia

High fevers are more common in children and may contribute to convulsions (febrile convulsions) and coma, with increased mortality. Persistently high temperatures ($\geq 42^\circ\text{C}$.) may cause permanent neurological sequelae.

Monitor temperature carefully. Children are also prone to hypothermia and so hyperthermia must be aggressively treated, whilst avoiding hypothermia. Treatment of a raised temperature includes the use of antipyretics and cooling methods such as tepid sponges and fans, aiming to keep the temperature $<39^\circ\text{C}$.

Gastrointestinal (GI) system

GI symptoms of malaria are frequently found in children, presenting with nausea and vomiting, abdominal pain and diarrhoea. There may be gastric/duodenal ulceration, malabsorption and an increase in gastrointestinal infections such as *salmonella*. Jaundice may be present due to haemolysis, hepatocellular dysfunction, cholestasis, or a combination of each. Serum bilirubin and liver enzymes may be elevated, although less than with viral hepatitis. Splenomegaly is also common and most likely related to the clearance of erythrocytes.

Spontaneous splenic rupture can occur with *P. vivax* infection.

Renal dysfunction

Acute renal failure usually occurs in adults. It can be due to prerenal or renal causes and is often secondary to acute tubular necrosis (ATN). Microvascular obstruction and cellular damage due to filtration of free haemoglobin, myoglobin and cellular material can lead to ATN. Usually there is a protracted period of oliguria, followed by anuria; occasionally a polyuria may be found. IV fluids and antimalarial treatment will aid pre-renal factors.

Monitor blood urea, creatinine and electrolytes and hydration status. A urinary catheter is required, with close monitoring of input and output balance. In severe cases, dialysis may be required.

Blackwater fever, although uncommon, can occur in severe malaria and is due to massive haemoglobinuria. It is more likely found in adults with renal failure. The urine is tea/coca-cola coloured. Those with glucose-6-phosphate dehydrogenase (G6PD) deficiency are at a greater risk of developing the condition, especially if receiving oxidant drugs such as primaquine and sulphonamides.

Hyperparasitaemia

High parasite densities (>5%) and peripheral schizontaemia tend to be associated with severe disease, particularly in children and those who are non-immune. In endemic areas, and in the partially immune, higher mean densities (20-30%) may be found without clinical symptoms.

Exchange transfusions can be considered in high parasitaemia, but must be weighed against the risk of the transfusion itself.

Miscellaneous

Malaria can be transmitted via a needle stick injury. Observe universal precautions.

SUMMARY

Malaria can vary from an insidious febrile illness to an acute life-threatening disease. Rapid deterioration is much more likely in children. If malaria is suspected, it should be investigated rapidly and treated appropriately. WHO and national diagnostic and treatment guidelines should be followed. Children should be observed closely for the development of complications. ICU admission may be required. Anaesthesia and surgery should be avoided in the child with malaria if at all possible, but, if it is necessary, the multi-system nature of the disease should be considered.

REFERENCES AND FURTHER READING

1. Espina-Bertosio S. Malaria for the anaesthetist. Update in Anaesthesia (2010)26:24-27. Available from: http://www.wfsahq.org/components/com_virtual_library/media/3e15badd08cee5e7f909ba92d916a0b1-Malaria-for-the-anaesthetist-Update-26-2010-.pdf
2. Fairhurst RM, Wellem TE. Chapter 275: Plasmodium Species (Malaria). In: Mandell, G.L., Bennett, JE and Dolin, R. eds. Principles and Practice of Infectious Diseases (7th Edition) New York: Churchill Livingstone Elsevier, 2009 pp. 3437-3462.
3. Gupta D, Chugh K, Sachdev A, Soni A. ICU management of severe malaria. *Indian J Pediatr* 2001; **68**: 1057-1061.
4. Pavsol, G. Chapter 2A: Malaria. In: Eddlestone M, Davidson R, Wiklinson R, Perini S. eds. Oxford Handbook of Tropical Medicine (2nd Edition) New York: Oxford University Press, 2005: pp 9-37.
5. World Health Organization. Guidelines for the treatment of malaria. (2nd edition). Geneva: World Health Organization; 2010. Available at: http://whqlibdoc.who.int/publications/2010/9789241547925_eng.pdf
6. World Health Organization. Management of severe malaria: a practical handbook. (2nd edition). Geneva: World Health Organization; 2000.
7. http://www.who.int/malaria/world_malaria_report_2011/WMR2011_factsheet.pdf (Accessed 16 June 2012).
8. <http://www.who.int/mediacentre/factsheets/fs094/en>

Acute lower respiratory disease in children

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INTRODUCTION

Pneumonia is the leading cause of death in children worldwide. Vulnerability is highest in the first 28 days of life, remaining a huge burden in the subsequent five years. In 2010 it was estimated that throughout the world 21,000 children under the age of five died every day, with a total of 7.6 million deaths each year.¹ Of these 1.4 million deaths (18%) were attributable to pneumonia, which is more than those caused by AIDS, malaria and tuberculosis combined.^{1,2}

The vast majority of these childhood deaths occur in the developing world, particularly in Africa and South East Asia. The risk of a child dying under the age of five in a low-income country is 18 times greater than in a high-income country.¹ The highest under-5 mortality rates are seen in the poor rural communities with low levels of education.^{1,3-5} Poverty contributes to increasing susceptibility through risk factors such as malnutrition, inadequate sanitation, and reduced access to health care services.^{1,3,6} Inhalation of particulate matter from indoor air pollution caused by the use of biomass fuels for heating or cooking is responsible for almost half of deaths due to acute lower respiratory tract infections.^{2,7}

DEFINITIONS

Acute lower respiratory disease in children consists of asthma (acute severe or life-threatening asthma) and Acute Lower Respiratory Infections (ALRI), which includes pneumonia and bronchiolitis. All are acute, serious and potentially life threatening.

Pneumonia is an acute lower respiratory tract infection that presents with symptoms of cough, fever, and difficulty breathing.

Asthma is a condition of hyper-reactive, inflamed and narrowed airways resulting in difficulty breathing, wheeze, cough and chest tightness. Although a chronic condition, patients often present with acute exacerbations related to infective or non-infective triggers (physical exertion, allergens, irritants or cold weather).

Bronchiolitis is an acute, communicable condition mainly affecting infants between 3-6 months of age.

Starting as an upper respiratory tract infection, it is caused by viral infections, which then trigger lower respiratory symptoms of bronchospasm, wheeze, cough and respiratory distress.⁸

PNEUMONIA

Pneumonia is most commonly caused by the bacteria *Streptococcus pneumoniae* (Spn) or *Haemophilus influenzae* (HiB). Other significant bacteria include *Staphylococcus aureus* and *Klebsiella pneumoniae*. Viral causes of pneumonia include Respiratory Syncytial Virus (RSV), parainfluenza virus and adenovirus. Fungal infections such as pneumocystis jiroveci are important to consider in the child with AIDS.^{2,5} Increasing immunisation coverage against Spn and HiB is expected to change the aetiology of pneumonia. The Pneumonia Etiology Research for Child Health (PERCH) study is a large multinational study investigating the aetiology of pneumonia in children.⁹

Transmission of pathogens between individuals usually occurs via droplets, which are aerosolised through coughing and sneezing. Neonates are also at risk of blood borne infection at or shortly after birth.^{2,5}

Risk factors for childhood ALRI are shown in Box 1. It is estimated that more than 20 million children suffer severe malnutrition, which compromises their defence against infection and increases their risk of dying from pneumonia. Immunity can also be weakened by concurrent illnesses such as HIV or measles. Non-exclusively breast fed infants are 15 times more likely to die from pneumonia, and suffer more frequent and severe infections than exclusively breastfed children.^{2,4}

Approximately one million children could be saved through effective prevention and treatment of pneumonia every year.² The Global Action Plan for the Prevention and Control of Pneumonia presents a framework to reduce pneumonia morbidity and mortality through three facets:⁷

1. Protection - strategies include the provision of a healthy living environment to enhance natural defences.

SUMMARY

Pneumonia continues to be the leading cause of death in children under 5, worldwide

Poverty contributes notably - the risk of a child dying under the age of five is 18 times greater in a low income country

Assessment and initial management follows the ABC approach, as for any acutely ill child

Standardised and prompt treatment improves outcomes

Supplementary oxygen saves extra lives.

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2. Prevention - includes using immunisation against *Haemophilus influenzae* B, *Streptococcus pneumoniae*, measles and pertussis. Children with HIV or diarrhoea are at even greater risk of pneumonia, and infected or exposed children should receive daily cotrimoxazole prophylaxis.^{2,7}

3. Treatment - for children with pneumonia, the mainstay of reducing mortality is appropriate treatment, either in the community, health centre or hospital. Evidence shows mortality can be reduced through:

- Standardised guidelines for diagnosis and treatment
- Early recognition and treatment with antibiotics.

The majority of cases of pneumonia in developing countries are bacterial in origin, but it is not possible to differentiate pathogens by clinical assessment.⁵ Less than 30% of children with pneumonia receive the appropriate antibiotics.²

ASTHMA

Over the past four decades the global prevalence, morbidity and mortality of asthma has risen sharply, particularly in children. While most of the 180,000 annual deaths from asthma are in patients over the age of 45 years, there has been a marked increase in hospitalisation in very young children. Prevalence of asthma is already high in

Box 1. Risk factors for Childhood Acute Lower Respiratory Infection (ALRI)^{3,10-12}

Definite Risk Factors for ALRI

- Malnutrition
- Low birth weight
- Non-exclusive breast feeding for first 6 months
- Lack of measles immunisations (within first 12 months)
- Indoor air pollution
- Crowded living

Likely Risk Factors for ALRI

- Parental smoking
- Zinc deficiency
- Maternal experience
- Concomitant diseases (diarrhoea, cardiac disease, asthma)

Possible Risk Factors for ALRI

- Mother's education
- Day-care attendance
- Rainfall (humidity)
- High altitude (cold air)
- Vitamin A deficiency
- Birth order
- Outdoor air pollution

high-income countries, reflecting increasing atopic sensitization. Prevalence is increasing in developing countries, possibly as a result of urbanization.¹³

Prevention of acute life threatening exacerbations depends on effective management of chronic disease. As with many diseases, this is hampered by poverty, poor education and limited access to health care.

BRONCHIOLITIS

Bronchiolitis predominantly affects infants under six months old. It is a leading cause of ALRI and a major contributor to acute respiratory distress, hospitalisation and PICU admission^{14,15} The most common pathogen is RSV and it is estimated that almost 199,900 children under the age of 5 died in 2005 as a result of RSV-associated ALRI. As with pneumonia, 99% of these deaths occurred in developing countries.¹⁴ Other infective agents include human meta-pneumovirus (HMPV), influenza, para-influenza, adenovirus, rhinovirus, and less commonly *Mycoplasma pneumoniae*.

Risk factors for bronchiolitis are shown in Box 2.

Morbidity and mortality may be reduced by ensuring a healthy living environment and addressing risk factors in a similar manner to those for pneumonia. Vaccination for RSV is only recommended in high-risk groups (e.g. children with significant cardiac disease), and has been shown to reduce the length of illness, reduce hospital stay and intensive care admissions.^{15,16}

PRESENTATION AND DIFFERENTIAL DIAGNOSIS

The common presenting symptoms and signs of acute lower respiratory tract disease are cough, difficulty breathing and wheeze. The differential diagnoses are summarised in Table 1. Children normally have a higher respiratory rate than adults; normal cardio-respiratory ranges are shown in Table 2. As a rule of thumb, a respiratory rate of more than 50 breaths per minute in a child aged between 2 and 12 months, or more than 40 breaths per minute in a child aged 1-5 years is considered rapid (Table 3). Upper airway conditions are described in detail in another article of this *Update* (page 168).

DIAGNOSIS, TRIAGE, AND MANAGEMENT

Assessment and triage of the seriously ill child are described in detail in 'Recognising the seriously ill child' in this edition of *Update* (page 224). Undertake an ABC assessment and categorise children according to the Emergency Triage Assessment and Treatment (ETAT) need for treatment²⁸: Emergency, Priority or Non-urgent. Emergency cases require immediate attention; priority cases require assessment and rapid attention; non-urgent cases can wait their turn in a queue.

Use a step-wise ABC approach for all critically ill children. You must monitor and record vital signs regularly (oxygen saturation, respiratory rate, heart rate, conscious level and temperature). Any deterioration should prompt full reassessment of the child:

- Re-evaluate the diagnosis
- Look for complications of the disease

Box 2. Risk factors for developing bronchiolitis^{14,15}

- Prematurity
 - Male gender
 - Age <3 months
 - Indoor air pollution (smoking/ biomass fuels)
 - Non-breast feeding
 - Poverty
 - Respiratory disease (chronic lung disease of prematurity, cystic fibrosis)
 - Congenital heart disease with pulmonary hypertension
 - Immunodeficiency
 - Neurodevelopmental delay
 - Upper airway obstruction
- Ask for senior support
 - Consider the need to transfer the child to intensive care facilities.

Pneumonia

The WHO recommends using clinical features of a recent onset cough and rapid breathing to diagnose pneumonia.^{5,28} Normal and abnormal cardiorespiratory values are shown in Tables 2 and 3. Consider other respiratory illnesses than pneumonia if the child has a cough without rapid breathing. Remember, it is not possible to differentiate between viral or bacterial pneumonia clinically as the symptoms are so similar. Some symptoms, such as wheezing do tend to occur more commonly in viral pneumonia, but do not rely on this to direct treatment.

When a child presents with symptoms of pneumonia, triage by the severity of their disease (See Table 4). Seek specific signs and symptoms, in particular

- The 'danger signs' (cyanosis, stridor, somnolence, lethargy or difficulty feeding)
- Lower chest wall in-drawing
- Rapid respiratory rate.

In normal respiration the whole chest expands with inspiration. In severe disease the lower chest wall may be sucked in during inspiration (lower chest in-drawing). In combination with rapid breathing and cough this represents severe clinical pneumonia. Other signs of severe difficulty in breathing are grunting with each breath, nasal flaring and tracheal tug. Children with very severe pneumonia will have additional symptoms and signs such as cyanosis, stridor, the inability to feed or drink, loss of consciousness, hypothermia or convulsions. They need urgent transfer to hospital.

Pulse oximetry correlates well with arterial PaO₂ in adults and is recommended by WHO for detecting hypoxia in children with acute respiratory disease and for guiding oxygen therapy.¹⁹ Target oxygen saturations are shown in Table 2. Where a pulse oximeter is not available, clinical signs can give useful clues to the presence of hypoxia:

- Central cyanosis
- Nasal flaring
- Grunting
- Altered mental state (drowsiness or lethargy)
- Inability to feed due to respiratory distress.

Be aware that no single sign can accurately identify hypoxia, so you should take signs together in context of the overall clinical condition of the child.³¹⁻³³ For example, the blue discolouration of lips or nail beds in central cyanosis can be difficult to identify. There is inter-observer disagreement and assessment is further complicated by the presence of severe anaemia (Hb<7g.dl⁻¹) or in dark skinned children. Central cyanosis is a highly specific sign but with low sensitivity.³² In a critically ill child, severe lower chest wall in-drawing, breathing rate of more than 70min⁻¹ or head bobbing may be more sensitive signs signaling the need for supplementary oxygen.¹⁹

Immediate treatment is outlined in Table 4. Many investigations to direct management (chest X-rays, blood tests, sputum tests) may be unavailable in the low-resource setting thus diagnosis and management is based on clinical symptoms and signs.⁴ All children with severe or very severe pneumonia and infants aged two months or younger require admission to hospital.²

The current recommendation is to administer IV antibiotics for at least three days. As the child recovers, switch to oral antibiotics (amoxicillin or ampicillin), and ensure that the child completes a total of at least five days. Reassess on a regular basis. Clinical deterioration or failure to improve by 48 hours should prompt a change in antibiotics (to chloramphenicol).¹⁸ Parenteral ampicillin plus gentamicin is preferable to chloramphenicol in treating severe pneumonia in children between one month and five years of age in a low-resource setting.³⁴

Complications of pneumonia

Hypoxaemia is the most serious complication of pneumonia.³⁵ It indicates severe disease and has been associated with four times increase in mortality.^{31,36} Most children who require oxygen will have very severe pneumonia, but hypoxia may also be present in children with less severe disease.

Oxygen is an expensive resource. Oxygen concentrators require a reliable supply of electricity and many rural health facilities may need to use cylinder supply. Cylinders are logistically challenging to transport to remote areas so that shortages occur frequently. Monitoring oxygen saturation and providing oxygen to children with severe pneumonia reduces the risk of death by 35%. This is in part due to improved detection of hypoxia and regular reassessment of treatment.³⁵ Neonates are vulnerable to the toxic effects of hyperoxia,

Table 1. Differential diagnosis for children presenting with acute respiratory symptoms^{2,5,15,17-25}

	Presenting Feature
Pneumonia	<ul style="list-style-type: none"> • Most common in 0-5-year-olds • Cough for less than 2 weeks • Rapid breathing rate or difficulty breathing • Fever or chills • Wheeze • Hypoxia (low SpO₂ or clinical signs - see text) • Loss of appetite or unable to feed due to respiratory distress
Bronchiolitis	<ul style="list-style-type: none"> • Age 3-6 months (less than 2 years) • 2-3 day coryzal phase with nasal discharge • Fast or difficult breathing • Harsh cough • Irritability or poor feeding • Wheeze • Fever <39°C • Apnoeas (especially in preterm infants) • Bilateral crepitations • Clinical signs of air trapping
Acute severe asthma	<ul style="list-style-type: none"> • Most common above 5 years of age • Known diagnosis of asthma and exposure to trigger factor • Difficulty in breathing/ respiratory exhaustion • Wheezing or chest tightness • Cough • Fast heart rate • Hypoxia • Hyperinflation of the chest • Confusion or drowsiness
Pleural effusion	<ul style="list-style-type: none"> • Cough • Rapid breathing • Wheeze • Chest pains • Vomiting • Fever (if empyema/ parapneumonic effusion) • Unilateral abnormal air entry • Unilateral dull percussion
Tuberculosis	<ul style="list-style-type: none"> • History of exposure (usually in a confined space) • Stridor • Wheeze • Hypoxia • Difficulty breathing or rapid breathing

Pneumothorax	<ul style="list-style-type: none"> • Sudden onset of difficulty in breathing • Wheeze • Hypoxia • Unilateral abnormal air entry • Unilateral hyper-resonant percussion • Tracheal deviation (indicates tension pneumothorax requiring immediate decompression)
Anaphylaxis	<ul style="list-style-type: none"> • Sudden onset • Airway compromise (swelling, stridor, hoarse voice) • Rapid breathing • Wheeze • Cyanosis/ hypoxia • Cardiovascular collapse (pale, clammy, low blood pressure) • Skin changes/ rashes
Upper airway conditions: e.g. airway foreign body, croup, epiglottitis, tracheitis	<ul style="list-style-type: none"> • Foreign body: sudden episode of coughing or choking, stridor, voice changes • Croup: stridor, barking cough • Epiglottitis/ tracheitis: toxic looking child, drooling
Other medical conditions presenting with respiratory symptoms:	
Gastro-oesophageal reflux disease	<ul style="list-style-type: none"> • Cough worse with feeds or lying flat • Intermittent wheeze • Vagally-mediated reactions (apnoea, bradycardia, laryngospasm)
Severe chronic anaemia	<ul style="list-style-type: none"> • Shortness of breath with exertion • Fatigue, weakness or irritability • Dizziness or syncope • Pallor
Severe sepsis	<ul style="list-style-type: none"> • Rapid breathing • Fast heart rate (or slow heart rate for age) • Fever, or low temperature • Presence of infection • Cardiovascular dysfunction (low blood pressure, prolonged capillary refill time, cold peripheries) • Hypoxia • Altered consciousness
Malaria	<ul style="list-style-type: none"> • Fever • Rapid breathing • Fast heart rate (may have circulatory collapse) • Altered consciousness, convulsions • Jaundice and abnormal bleeding • Children with malaria may also have severe anaemia or concomitant pneumonia
Heart failure and congenital cardiac conditions	<ul style="list-style-type: none"> • Fast breathing rate • Fast heart rate • Clammy, pale, cold peripheries • Weak pulses • Irritability • Limited exertion tolerance • Hypoxia • Enlarged liver

Table 2. Normal values in children^{14,19,26}

	Neonate	Infant	Small child	Adolescent
Heart rate	110-150	100-150	80-120	60-100
Respiratory rate	30-40	25-35	25-30	15-20
Oxygen Saturation	88% at sea level	Altitude greater than 2500m: SpO ₂ > 87% Altitude less than 2500m: SpO ₂ > 90%		
Systolic Blood Pressure (lower limit, mmHg)	65-75	70-80	(65+2 x age)	90

Table 3. Definition of 'rapid breathing' and 'increased heart rate' in children²⁶⁻²⁸

	<2 months	2-12 months	1 -5 years	>5 years
Breathing Rate	>60	>50	>40	>30
Heart Rate (Beats per minute)	>150	>150	>140	>125

Table 4. Assessing the severity of pneumonia^{5,18,19,29}

Signs	Classification	Immediate treatment
Danger signs present: <ul style="list-style-type: none"> • Cyanosis • Stridor (in calm child) • Somnolence • Lethargy • Difficulty drinking liquids 	Very severe pneumonia	If in hospital: Seek urgent senior help Give oxygen Give appropriate antibiotics IV Consider admission to Intensive Care If not in hospital: Seek senior help urgently Give first dose of appropriate antibiotic IV. Refer urgently to hospital for IV antibiotics and oxygen.
Fast breathing Lower chest wall in-drawing	Severe pneumonia	If in hospital: Give first dose of appropriate antibiotic promptly and continue Assess need for oxygen If not in hospital: - Give first dose of appropriate antibiotic promptly - Refer urgently to hospital for antibiotics and oxygen therapy as required
Fast breathing: >50 bpm in 2-12months >40 bpm in 1-5 years	Pneumonia (non-severe)	If no lower chest wall in-drawing or danger signs: Prescribe appropriate antibiotics Does not require hospitalisation Advise mother on supportive measures and when to return for follow-up
No fast breathing	Other respiratory illness	No need for antibiotics Does not require hospitalisation Advise mother on supportive measures and to return if symptoms worsen

so should only receive monitored oxygen therapy to maintain SpO₂ around 87%-94%.¹⁹

Severe pneumonia may be complicated by a collection of either fluid (parapneumonic effusion) or pus (empyema) in the pleural space, adding significantly to morbidity.^{37,22} Exclude these complications in any child who is not responding to treatment after 48 hours. They also necessitate admission to hospital with intravenous antibiotic therapy. Chest imaging with anteroposterior or posteroanterior X-rays and ultrasound can aid diagnosis and guide thoracocentesis. Chest drains should be sited for large collections and will prompt referral to a tertiary centre. Blood, sputum and pleural fluid may be sent for microbiological evaluation if available. Remember that 6% of empyemas globally are due to tuberculosis.³⁷

Asthma

Acute severe asthma and life-threatening asthma are conditions of severe bronchospasm. Inflammation and mucus secretions worsen airway narrowing. It is important to realise that acute exacerbations may be life threatening – do not underestimate their severity.^{20,21} Asthma is uncommon in children under the age of 5 years old.

A child with asthma should be assessed with an ABC approach. Clinical symptoms and signs do not always correlate well with the degree of airway obstruction. Severity classification is shown in Table 5. Pay particular attention to respiratory rate, degree of breathlessness, use of accessory muscles, amount of wheeze, heart rate and degree of agitation or drowsiness. Danger signs include a silent chest, cyanosis or low SpO₂, poor breathing effort or exhaustion, low blood pressure and confusion. As asthma worsens so does the heart rate. A falling heart rate in a child with life-threatening asthma is a pre-terminal sign.^{21,27}

Oxygen saturation monitoring is essential in all wheezy children. Peak Expiratory Flow (PEF) rate is commonly used for disease monitoring in developed countries. If the child is familiar with its use it can also provide additional information in the acute exacerbation, but is not essential.

For children under the age of 2 years the primary cause of intermittent wheeze is usually viral infections. Very few will have wheeze due to asthma.²⁷

The child with an acute severe exacerbation of asthma needs immediate medical attention, assessment and treatment. It can be a frightening situation, not only for the child and their carers, but also for hospital staff, so call for senior help immediately. Keep calm so as not to distress the child further. Allow the child to position itself in the most comfortable posture, which is often sitting upright, to reduce respiratory distress and improve chest wall movement.

Management of asthma is shown in Box 3. Treatment aims to reverse bronchospasm so that normal respiratory gas exchange is restored. Use bronchodilators to relieve bronchospasm, steroids to reduce bronchial oedema, and treat any infection if present. The majority of acute asthma attacks are triggered by viral infection so antibiotics should not be given routinely.²⁷ Avoid any drugs that are known to release histamine, as they will worsen bronchospasm. Consider medications such as magnesium, salbutamol, aminophylline and

ketamine if initial treatments fail and appropriate HDU/ICU level care and monitoring is available. The indications for admitting a child to PICU are shown in Box 4.

Chest Xrays are not routinely necessary. Use them for signs of surgical emphysema, persistent signs of pneumothorax, consolidation or if life-threatening asthma is not responding to treatment.²⁷

For children under two years, response to treatment can be unpredictable. First line treatment of salbutamol given via an inhaler and spacer with a close fitting mask is better than nebulizer.²⁷ Ipratropium may be beneficial to infants with severe symptoms. Steroids should be given (10mg soluble prednisolone for up to three days). After discharge it is not usually necessary to continue bronchodilator therapies in this age group.

Bronchiolitis

Bronchiolitis is diagnosed on the basis of clinical features. The child is usually between 3-6 months old and certainly under the age of two years old. High fever is uncommon and temperatures > 39°C should prompt evaluation for other causes. Danger signs are shown in Table 6.

The principle of managing bronchiolitis is providing supportive care since there are no effective therapies (Box 5). Admission to hospital is often necessary because the baby is unable to feed due to respiratory distress, and for administration of oxygen. As far as possible, give enteral (oral or nasogastric) fluids and feed. If the child is too sick to tolerate enteral feed, give intravenous fluids at 2/3 maintenance rate. For calculation of fluid requirements see 'Recognising the Critically Ill Child' article in this edition of *Update*, page 224.

Approximately 2% of infants will require ventilatory support. CPAP may reduce the need for intubation and ventilation. Look out for any infant who is tiring or has other danger signs indicating the need for intubation or PICU admission. (See Box 6).

Routine antibiotics are not recommended.^{17,38} Children requiring ventilatory support or with more serious illness may have co-existing bacterial infections, so empirical antibiotic cover is reasonable in these children.

One or two doses of 3% saline administered through a nebuliser may increase the clearance of mucus in children with non-severe bronchiolitis presenting to hospital. Evidence suggests a shorter hospital stay with no significant adverse effects. Further research is required to determine the safety profile of nebulised hypertonic saline.³⁹

The wheeze in bronchiolitis is mainly caused by debris in the airway, unlike the bronchospasm seen with asthma. Bronchodilators are not recommended due to their lack of efficacy, cost and side-effects (rapid heart rate and shakiness).^{8,17}

Current evidence implies possible positive short-term benefit of steroids for inpatients with bronchiolitis, but long-term outcomes are unchanged.⁴⁰ Although some research indicates that the combination of steroids used with epinephrine nebulisers may reduce outpatient admissions, long term effectiveness and safety still

Table 5. Assessing the severity of asthma in children > 2 years old^{21,27}

Symptoms and Signs	Classification	Immediate treatment
'Danger signs': <ul style="list-style-type: none"> • Silent Chest • Cyanosis or low saturation (SpO₂ <92%) • Poor respiratory effort • Low blood pressure • Exhaustion • Confusion • PEF <33% predicted/best 	Life threatening	Seek urgent senior help Continue management as for severe asthma Consider adjuncts, intubation and ventilation. Needs ICU admission
<ul style="list-style-type: none"> • Can't complete sentences or unable to feed • PEF 33-50% of predicted/best • Fast breathing: <ul style="list-style-type: none"> - >50 bpm 2-12month old - >40 bpm 1-5 year old - >30 bpm >5 year old • Fast heart rate <ul style="list-style-type: none"> - >140 2-5 year old - >125 >5 years year old 	Severe	Seek urgent senior help Check ABC Administer oxygen Nebulised salbutamol 2.5-5mg (or 2 puffs of inhaler via spacer every 2 mins up to 10 puffs) Nebulised ipratropium bromide
<ul style="list-style-type: none"> • Able to talk in sentences or feed well • SpO₂ >92% • PEF >50% predicted/ best • Fast breathing but: <ul style="list-style-type: none"> - <50 bpm 2-12month old - <40 bpm 1-5 year old - <30 bpm >5 year old • Fast heart rate, but: <ul style="list-style-type: none"> - <140 in 2-5 year old - <125 in >5 year old 	Moderate	Administer Oxygen Nebulised Salbutamol 2.5-5mg (or 2 puffs of inhaler via spacer every 2 mins up to 10 puffs) Nebulised Ipratropium

requires further research and adrenaline nebulisers are not routinely recommended.^{17,40,41} There is insufficient evidence to support the use of adrenaline in inpatients, but it may be of value in the outpatient setting.⁴¹

ADVANCED VENTILATORY SUPPORT IN CHILDREN WITH ALRI

Advanced ventilatory support necessitates high-dependency or paediatric intensive care admission. The main options are either non-invasive or invasive ventilation.

Non-invasive ventilation

Non-invasive ventilation relies on augmenting the child's own respiratory effort to reduce the work of breathing. Continuous Positive Airway Pressure (CPAP) acts as its name suggests by administering a

continuous positive pressure of between 5-10 cmH₂O to keep alveoli open, reduce sheering injury and allow improved oxygenation. CPAP can be administered via nasal prongs in infants or through a tight fitting facemask in older children. Low cost systems are available. Non-invasive positive pressure ventilation (NIPPV) administers an alternating inspiratory and expiratory pressure triggered by the patient's own breathing efforts. It is particularly useful for aiding carbon dioxide clearance. Non-invasive techniques may reduce the need for intubation and ventilation.

Invasive ventilation

Invasive ventilation requires intubation with an endotracheal tube. There are numerous ventilation modes and equipment available. The choice of technique will depend on clinician experience, equipment availability and the child's clinical condition. Lung-protective strategies should be used (Box 7). The process of intubating a

Box 3. Management of acute severe asthma^{15,21,27}

Initial management of acute severe asthma:

- Asses ABC and administer high flow oxygen
- Nebulised salbutamol (2.5-5mg) or 10 puffs administered via an inhaler and spacer
- Nebulised ipratropium bromide (250 micrograms)
- Oral prednisolone 20mg (2-5years), 30-40mg (>5year). Use hydrocortisone 4mg.kg⁻¹ IV if unable to take orally
- Review response to treatment. Repeat ABC assessment

If there is no improvement:

- Adrenaline IM 10 micrograms.kg⁻¹ (consider IV adrenaline infusion only if specialized syringe pumps and continuous electrocardiography monitoring are available)
- IV salbutamol bolus dose (15mcg.kg⁻¹ over 10 min)
- IV aminophylline
- IV magnesium sulphate 40mg.kg⁻¹ (max 2g)
- Ketamine or volatile agents may assist in relieving intractable bronchospasm

Subsequent management:

- Rehydrate with 10-20ml.kg⁻¹ crystalloid
- Oral prednisolone 20mg (2-5years), 30-40mg (>5year) for three days
- Repeat nebulisers every 20-30 minutes if required, or 3-4 hourly
- Perform a chest X-ray and arterial blood gases if life-threatening asthma is not responding to treatment
- Consider admission to PICU for non-invasive ventilation or intubation and ventilation

Box 4. Indications for admission to ICU for children with asthma¹⁵

- The child has severe acute- or life-threatening asthma
- The child is not responding to treatment
- Persistent or worsening hypoxia
- Hypercarbia
- Falling pH on arterial blood gases
- Exhaustion, feeble respiration, drowsiness or confusion

critically ill child can provoke cardiorespiratory instability and an initial deterioration. Be prepared for potential complications as well as competent in the management of the paediatric airway. Positive pressure ventilation may also reduce venous return to the heart thus causing low blood pressure.

In cases of severe refractory hypoxia, consider high frequency oscillation (HFO) if available. This acts on the 'open lung strategy' by providing a continuous distending pressure around which oscillations of a frequency of >150 per minute allow gas exchange.¹⁵

Invasive ventilation - special considerations

Asthma

If a child with life-threatening asthma fails to respond to maximal

medical treatment, the decision to intubate and ventilate should be based on the following criteria:

- Availability of Paediatric Intensive Care facilities
- Respiratory arrest
- Hypoxia and rising hypercarbia
- Exhaustion
- Altered mental state.

Intubation risks significant complications, which you should anticipate and plan for. These include worsening of bronchospasm, laryngospasm, worsening hypoxia, pneumothorax and barotrauma, and hypotension due to reduced venous return.

Table 6. Assessing the severity of bronchiolitis^{15,17}

Symptoms and Signs	Classification	Immediate treatment
Danger signs: Breathing Rate >70bpm Periods of apnoea Infant looks ill and exhausted Sweaty and irritable Atelectasis, marked over- inflation or evidence of ARDS on CXR	Severe	Seek urgent senior help Administer oxygen Assess need for ventilatory support or admission to PICU
Fast breathing rate Increased effort of breathing Cough Wheeze Hyperinflation of the chest Fast heart rate	Moderate	Consider admission to hospital Administer oxygen if required Maintain hydration

Box 5. Management of bronchiolitis

- Assess ABC
- Administer oxygen to maintain appropriate saturations
- Maintain hydration:
 - Attempt to hydrate with oral or NG fluids
 - IV fluids may be required if the child is too sick to take oral or NG feed
 - Restrict IV fluids to 2/3 maintenance
- Review indications for ventilation and PICU admission
- Consider nebulised adrenaline

Box 6. Indications for PICU admission in children with bronchiolitis¹⁵

- Increasing oxygen requirement or inability to maintain adequate oxygen saturation in high flow oxygen
- Signs of becoming exhausted
- Progressive rise in PaCO₂
- Apnoeas

Process of intubation:

- Ensure good intravenous access
- Give IV fluid bolus 10ml.kg⁻¹ of crystalloid
- Pre-oxygenate for 3 minutes with tight fitting mask
- Use ketamine for induction as it has bronchodilator effects (2mg.kg⁻¹)
- Use a cuffed endotracheal tube as high inflation pressures may be required
- Avoid rapid ventilation of the child once intubated as this will lead to air trapping, increase the risk of pneumothorax and worsen gas exchange. Use a slow rate with a long expiratory time
- If severe air trapping does occur, disconnect the child from the ventilator and physically squeeze on the chest to assist expiration
- Always perform a chest X-ray after intubation to confirm the correct position of the endotracheal tube and to carefully check for pneumothorax which will worsen with positive pressure ventilation

Table 7. Ventilation in acute severe asthma⁴²

		Rationale
<i>Mode of ventilation</i>	Pressure Controlled (PC)	The decelerating waveform results in lower inspiratory pressures for a given mean airway pressure
	Volume Controlled (VC) Aim for tidal volume 4-8ml.kg ⁻¹	PC is preferable but VC may be used.
<i>Rate</i>	Slow for age 5-15 breaths per minute	A slow rate reduces air-trapping
<i>Inspiratory time</i>	Suggested 1.0-1.5 seconds	
<i>I:E Ratio</i>	1:3 - 1:5	Long expiratory time helps avoid dynamic hyperinflation
<i>Positive Inspiratory Pressures (PIP)</i>	Limit PIP < 35cmH ₂ O Limit inspiratory plateau pressure <30 cmH ₂ O	High inspiratory pressures are likely to be required
<i>PEEP</i>	Avoid high PEEP	High PEEP will worsen hyperinflation and air trapping

Box 7. Principles of lung protective ventilation^{15,42}

The main aim is to optimise lung mechanics to reduce parenchymal damage, accepting altered physiological goals.

Avoid the following:

Barotrauma

High pressures over-stretch healthy lung:

Maintain plateau airway pressures <30cmH₂O

Maintain peak airway pressures <35cmH₂O

Volutrauma

High tidal volumes cause sheering injury to the alveoli:

Maintain tidal volumes 4-8ml.kg⁻¹

Hyperoxia

Hyperoxia worsens atelectasis and inflammatory changes. Use the minimum oxygen required to maintain oxygen saturations:

If possible keep FiO₂ ≤0.4

Consider the following:

PEEP to recruit alveoli

Avoid in conditions which have high intrinsic PEEP such as acute severe asthma

Permissive hypercapnoea

Accept high PaCO₂ if pH normal or near normal (>7.2)

Relative hypoxia (accept SpO₂ 85-90%)

Optimise oxygen delivery (Cardiac output, Haemoglobin) and if possible, use lactate level to assess organ perfusion.

Ventilation of a child with asthma can be difficult. Due to the tight bronchoconstriction, high pressures may be required. Be careful to use techniques to limit barotrauma and air trapping which are common in asthma. Hypercarbia can be tolerated so long as the pH remains >7.2. Table 7 shows suggestions for ventilating such a child.

Continue bronchodilators during ventilation. Administer by nebulised form via the breathing system or as intravenous infusions.

Ketamine (0.5-2mg.kg⁻¹.hour⁻¹) may be used as a sedative, either alone or with morphine/midazolam.

If the patient remains hypoxic, investigate for pneumothorax and signs of infection. Evaluate the use of beta2-agonists, which may worsen VQ mismatch in the presence of hypovolaemia.

Bronchiolitis

In bronchiolitis, the primary difficulty is achieving adequate oxygenation. High inspiratory pressures may be required. Low respiratory rates with long inspiratory times will help to allow adequate tidal volumes while reducing the risk of barotrauma to the lungs. Positive End Expiratory Pressure (PEEP) of 5-10cmH₂O will prevent alveolar collapse and further aid oxygenation.

CONCLUSIONS

Poverty is a substantial risk factor for many diseases, both infectious and non-communicable. Not only are the poor more vulnerable to developing disease, but they are also least likely to be able to access, afford and receive adequate treatment. All children presenting with acute lower respiratory tract disorders should be managed with the same systematic (ABC) approach. Following diagnosis, the key is to deliver prompt treatment, regularly reassess the child's response to treatment and watch for signs of deterioration, which may necessitate admission to intensive care facilities. Development of guidelines will improve the quality of treatment delivered, thereby reducing morbidity and mortality.

REFERENCES

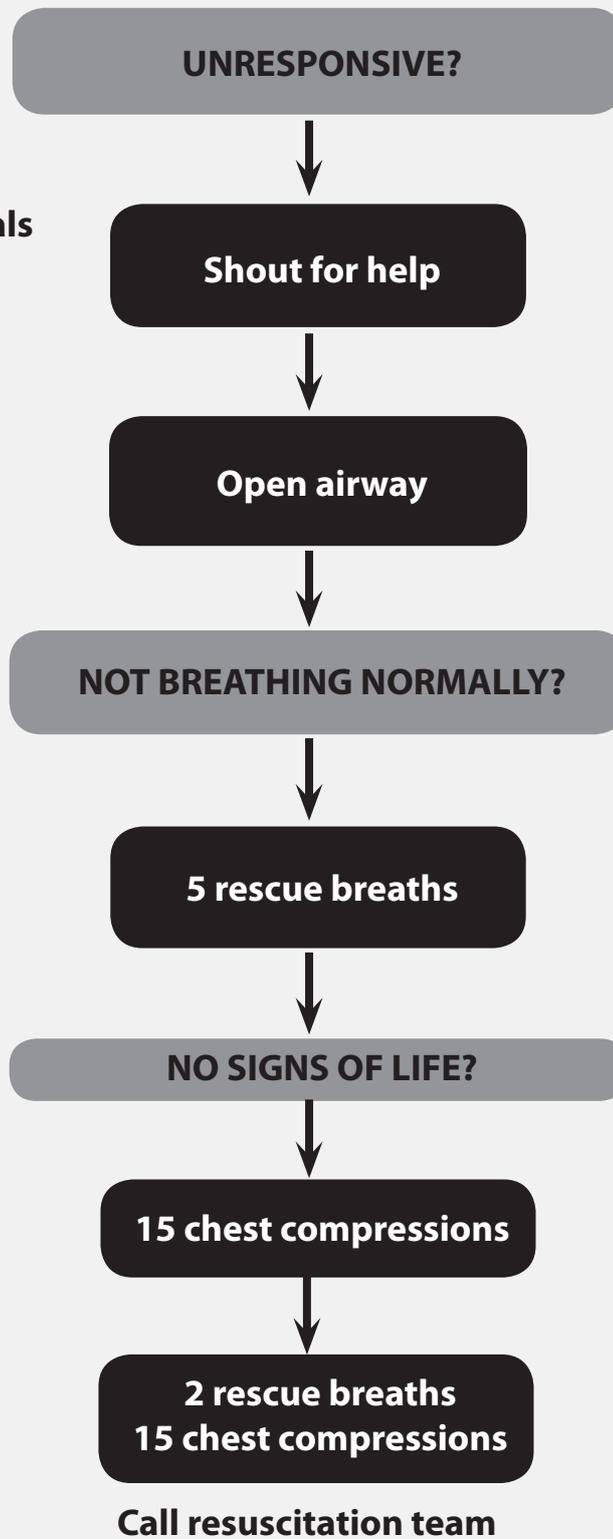
- World Health Organisation (WHO). Child health epidemiology. http://www.who.int/maternal_child_adolescent/epidemiology/child/en/ (accessed 25th August 2014).
- World Health Organisation (WHO). Fact Sheet N° 331: Pneumonia. <http://www.who.int/mediacentre/factsheets/fs331/en/> (accessed 25th August 2014).
- World Health Organisation (WHO). Fact sheet N°178: Children - reducing mortality. <http://www.who.int/mediacentre/factsheets/fs178/en/> (accessed 25th August 2014).
- United Nation Children's Fund (UNICEF). Pneumonia and diarrhoea: Tackling the deadliest diseases for the world's poorest children. New York: UNICEF, 2012.
- United Nation Children's Fund (UNICEF) /World Health Organisation (WHO). Pneumonia: The forgotten killer of children, 2006.
- United Nation Children's Fund (UNICEF). Child info: monitoring the situation of children and women - Pneumonia. <http://www.childinfo.org/pneumonia.html> (accessed 28th August 2012).
- World Health Organisation (WHO)/The United Nations Children's Fund (UNICEF). Global action Plan for Prevention and control of Pneumonia (GAPP). France: WHO, 2009.
- Gadomski AM and Brower M. Bronchodilators for bronchiolitis (a review). *Cochrane Database of Systematic Reviews* 2010; Issue 12.
- Gilani Z, Kwong YD, Levine OS, Deloria-Knoll M, Scott JAG, O'Brien KL et al. Literature Review and Survey of Childhood Pneumonia Studies: 2000-2010. *Clin Infect Dis* 2012; **54**(Supplement 2): S102-108.
- Rudan I, Boschi-Pinto C, Biloglac Z, Mulholland K, Campbell H. Epidemiology and etiology of childhood pneumonia. *Bull World Health Organ* 2008; 86(5):321-416. <http://www.who.int/bulletin/volumes/86/5/07-048769/en/index.html>.
- World Health Organisation (WHO). Fact sheet N°292: Indoor air pollution and health (2011) <http://www.who.int/mediacentre/factsheets/fs292/en/> (accessed 22rd July 2012).
- World Health Organisation (WHO). What are the key health dangers for children? <http://www.who.int/features/qa/13/en/> (accessed 6th September 2012).
- Braman SS. The Global Burden of Asthma. *Chest* 2006; **130** (Supplement 1): 4S-12S.
- Nair H, Nokes JD, Gessner B, Dherani M, Madhi S, Singleton, RJ et al. Global burden of acute lower respiratory infections due to respiratory syncytial virus in young children: a systematic review and meta-analysis. *Lancet* 2010; **375**: 1545-55.
- Chapter 21: Respiratory Disease. In: Barry P, Morris K and Ali T (eds). *Oxford Specialist Handbook in Paediatrics: Paediatric Intensive Care* (1st edition). Oxford: Oxford University Press 2010: 407-450.
- Dudas R and Karron R. Respiratory Syncytial Virus Vaccines. *Clin Microbiol Rev* 1998; **11**(3): 430-439.
- British Thoracic Society (BTS) & Scottish Intercollegiate Guidelines Network (SIGN). 91: Bronchiolitis in Children: a national clinical guideline. Edinburgh: SIGN, 2006.
- Rojas-Reyes M and Granados Rugeles C. Oral antibiotics versus parenteral antibiotics for severe pneumonia in children. *Cochrane Database of Systematic Reviews* 2009, Issue 3.
- World Health Organisation (WHO). Evidence for Technical Update of Pocket Book Recommendations: Recommendations for management of common childhood conditions - Newborn conditions, dysentery, pneumonia, oxygen use and delivery, common causes of fever, severe acute malnutrition and supportive care. Switzerland: WHO, 2012.

20. National Asthma Education and Prevention Program. Expert Panel Report 3: Guidelines for the Diagnosis and Management of Asthma. US Department of Health and Human Services 2007 <http://www.nhlbi.nih.gov/guidelines/asthma/asthgdln.pdf>.
21. McIndoe, A. Chapter 35: Anaesthetic Emergencies: Paediatric Emergencies. In: Allman KG and Wilson IH (eds) Oxford Handbook of Anaesthesia (2nd edition) Oxford: Oxford University Press, 2006: 902-915.
22. Langley J, Kellner JD, Solomon N, Robinson JL, Le Saux N, McDonald J et al. Empyema associated with community-acquired pneumonia: A Pediatric Investigator's Collaborative Network on Infections in Canada (PICNIC) study. *BMC Infectious Diseases* 2008; **8**:129.
23. World Health Organisation (WHO). Early detection of tuberculosis An overview of approaches, guidelines and tools. Geneva:WHO, 2011.
24. World Health Organisation (WHO). Tuberculosis. <http://www.who.int/topics/tuberculosis/en/> (Accessed 21st September 2012).
25. Grant GB, Dowell SF, Graham SM, Klugman KP, Mulholland EK, Steinhoff M et al. Recommendations for treatment of childhood non-severe pneumonia. *Lancet Infect Dis* 2009; **9**: 185-96.
26. Fleming S, Thompson M, Stevens R, Heneghan C, Plüddemann A, Maconochie I et al. Normal ranges of heart rate and respiratory rate in children from birth to 18 years of age: a systematic review of observational studies. *Lancet* 2011; **377**:1011-1018.
27. British Thoracic Society (BTS) & Scottish Intercollegiate Guidelines Network (SIGN). 101: British guideline on the management of asthma: A national clinical guideline. Edinburgh:SIGN, 2012.
28. World Health Organisation (WHO). Manual for the health care of children in humanitarian emergencies. Spain:WHO, 2008.
29. WHO (2008) Integrated Management of Childhood Illness Chart booklet Available online [http://whqlibdoc.who.int/publications/2008/9789241597289_eng.pdf].
30. Lifebox (<http://www.lifebox.org>).
31. Subhi R, Adamson M, Campbell H, Weber M, Smith K, Duke T. The prevalence of hypoxaemia among ill children in developing countries: a systematic review. *Lancet Infect Dis* 2009; **9**: 219-27.
32. Ayieko P and English M. In Children Aged 2-59 months with Pneumonia, Which Clinical Signs Best Predict Hypoxaemia? *J Trop Pediatr*, 2006; **52**(5): 307-310.
33. Rojas-Reyes M, Granados Rugeles C, Charry-Anzola LP. Oxygen therapy for lower respiratory tract infection in children between 3 months and 15 years of age (Review). Cochrane Database of Systematic Reviews 2009; Issue 3.
34. Asghar R, Banajeh S, Egas J, Hibberd P, Iqbal I, Katep-Bwalya M et al. Chloramphenicol versus ampicillin plus gentamicin for community acquired very severe pneumonia among children aged 2-59 months in low resource settings: multicentre randomised controlled trial (SPEAR study). *BMJ* 2008; **336**:80-93.
35. Duke T, Wandt F, Jonathan M, Matai S, Kaupa M, Saavu M et al. Improved Oxygen systems for childhood pneumonia: a multihospital effectiveness study in Papua New Guinea. *Lancet* 2008; **372**: 1328-33.
36. Cantin A. Childhood pneumonia and oxygen treatment. *Lancet* 2008 **372**:1279-1280.
37. Balfour-Lynn I, Abrahamson E, Cohen G, Hartley J, King S, Parikh D et al. BTS guidelines for the management of pleural infection in children. *Thorax* 2005; **60**(Supplement 1):i1-i21.
38. Spurling GKP, Doust J, Del Mar CB, Eriksson L. Antibiotics for bronchiolitis in children (review). Cochrane Database of Systematic Reviews 2011; Issue 11.
39. Zhang, L et al 'Nebulised hypertonic saline solution for acute bronchiolitis in infants (Review)' Cochrane Database of Systematic Reviews 2011; Issue 3.
40. Fernandes RM, Bialy LM, Vandermeer B, Tjosvold L, Plint AC, Patel H et al. Glucocorticoids for acute viral bronchiolitis in infants and young children (Review). Cochrane Database of Systematic Reviews 2010; Issue 11.
41. Hartling L, Bialy LM, Vandermeer B, Tjosvold L, Johnson DW, Plint AC et al. Epinephrine for Bronchiolitis (Review). Cochrane Database of Systematic Reviews 2011; Issue 6.
42. Jauncey-Cooke JI, Bogossian F, East CE. Lung protective ventilation strategies in paediatrics - A review. *Aust Crit Care* 2010; **23**: 81-88.



Paediatric Basic Life Support

(Healthcare professionals
with a duty to respond)



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Paediatric life support

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There are some differences between resuscitation techniques for children and adults but there are also many similarities. There is no doubt that a child in cardiorespiratory arrest will be harmed more by doing nothing than by using adult resuscitation guidelines.

Children usually suffer from secondary cardiac arrest – the heart stops secondary to hypoxia or ischaemia caused by respiratory or circulatory failure. The main implication of this is that there is potential to recognise the primary cause early on and prevent its progression to full blown arrest. Respiratory or circulatory failure is initially compensated by the body's physiological mechanisms and the signs are fairly subtle.

Signs of compensated respiratory failure

- Tachypnoea or bradypnoea (e.g. in narcotic overdose)
- Increased work of breathing:
 - Anxious appearance
 - Use of accessory muscles of respiration
 - Noises – stridor, grunting or wheeze
 - Nasal flaring.

Signs of compensated circulatory failure

- Tachycardia
- Slow capillary refill
- Cool peripheries
- Thirst
- Lethargy.

In the compensated phase there are good opportunities to prevent deterioration by the administration of general treatment such as oxygen and fluid (in circulatory failure) and specific treatments such as salbutamol in asthma and antibiotics in pneumonia or sepsis. This compensated phase may progress to decompensation, however and, if immediate action is not then taken, to cardiorespiratory arrest.

Signs of decompensation

Diminishing level of consciousness is an important sign of decompensation and imminent arrest

In addition, for decompensating respiratory failure

- Sudden fall in respiratory rate
- Exhaustion
- Very quiet or silent chest.

Decompensating circulatory failure

- Hypotension
- Sudden fall in heart rate.

Fortunately, the actions required to reverse this process are usually simple and follow the familiar ABC format.

COMMENTARY - BASIC LIFE SUPPORT (Figure 1)

A – Airway

Opening a child's airway is similar to opening that of an adult – a head tilt and chin lift. The most important difference is to avoid pressing on the soft tissues underneath the jaw as this pushes the tongue backwards into the oropharynx and can worsen airway obstruction. Infants have a prominent occiput and simply require the head to be placed in a neutral position – overextension is not helpful. If this simple manoeuvre is ineffective a jaw thrust (performed in the same way to that in adults) usually works.

Sometimes an airway adjunct is required and the most useful is an oropharyngeal airway. The size is selected so that tip of the airway is level with the angle of the jaw when the flange is lined up with the lips. It can be inserted in the same way as for an adult airway (i.e. introduced upside down and then rotated 180 degrees into its final position) but care should be taken not to damage the hard palate.

Successful opening of the airway should be assessed by **looking** (for chest movement), **listening** (for air flow at the nose and mouth) and **feeling** (for expired air on your cheek held close to the child's nose and mouth).

Summary

Children usually suffer cardiac arrest secondary to hypoxia or ischaemia due to respiratory or circulatory failure.

Cardiac arrest is commonly reversed by simple interventions.

Early recognition of a child at risk of deterioration is essential.

Avoid interruptions in chest compressions.

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- If there is chest movement and you can hear and feel expired air then the airway is clear and oxygen (if available) should be given.
- If there is chest movement but no expired air then the airway is still obstructed and it should be repositioned.
- If there is no chest movement, positive pressure ventilation is required.

B – Breathing

Positive pressure ventilation (IPPV) may be given with expired air (mouth to mouth) or bag mask ventilation (BMV) with a self-inflating bag/mask system.

Mouth to mouth ventilation requires no equipment but is inefficient as it only delivers expired oxygen concentrations (about 17%). Nevertheless it can be lifesaving.

The most important points are to open the airway effectively (as above) and to achieve a good seal with your lips over the child's mouth (or nose and mouth in a small infant). You should deliver enough breath to make the child's chest rise as if they had taken a normal breath.

The same principles apply to BMV – the airway should be open and there should be a good seal, this time between the mask rim and the child's face. If this is difficult it may help to have 2 people – one to do a jaw thrust and to achieve a seal with the mask using both hands and the other to squeeze the reservoir bag. Again, the aim is to make the chest rise as if the child has taken a normal breath. Five rescue breaths should be delivered in this fashion and then an assessment of the circulation should be made.

C – Circulation

In diminished level of consciousness due to decompensated respiratory or circulatory failure, failure to respond to positive pressure ventilation by moving, coughing or resuming breathing is a sure sign of absence of an effective circulation and external chest compression (ECC) should be immediately commenced. Prolonged searching for a pulse (>10 seconds) is unnecessary may result in error or delay.

ECC is performed by compression of the chest to a depth of 1/3 to 1/2 of the A-P diameter, at a point just (1 finger's breadth) above the xiphisternum. Don't be afraid of pushing too hard. Compressions should be at a rate of 100 per minute and 2 breaths should be given after every 15 compressions. Compressions should be interrupted as little as possible so, if the trachea is intubated, they should be continuous with about 10 breaths delivered every minute. Generally, people ventilate too vigorously during resuscitation and this has been shown to impede venous return and reduce blood flow.

If a monitor or defibrillator is available it should be applied to check whether there is a shockable cardiac rhythm (ventricular fibrillation or ventricular tachycardia) or not. Adrenaline (10mcg.kg⁻¹) should be given every 3-5 minutes during ECC as it increases cerebral and myocardial perfusion.

COMMENTARY - ADVANCED LIFE SUPPORT (Figure 2)

1. Shockable rhythms - ventricular fibrillation (VF) or pulseless ventricular tachycardia (pVT)

If a shockable rhythm is present defibrillation with 4J.kg⁻¹ should be performed immediately. Chest compression should be restarted immediately even if a rhythm change is seen on the monitor. This is important, as the heart will not be able to support the circulation for a minute or so, even if sinus rhythm resumes. If defibrillation is unsuccessful, CPR should be continued for a further 2 minutes and the defibrillation cycle repeated. If a third shock is necessary, epinephrine (adrenaline) should be given immediately before and an anti-arrhythmic should be used before a fourth shock. Amiodarone (5mg.kg⁻¹), where available, is preferred but lidocaine (1mg.kg⁻¹) is an acceptable alternative.

2. Non-shockable rhythm - asystole or pulseless electrical activity (PEA)

If the rhythm is not shockable, the emphasis is on good quality CPR with minimum interruption in ECC together with adrenaline administration every 3-5 minutes.

3. Reversible causes

In both shockable and non-shockable rhythms treatable causes should be sought and dealt with. Children rarely suffer from primary heart disease, so there is often a precipitating cause and resuscitation will not be successful if this is not removed. Treatable causes can be remembered by the 4Hs and the 4Ts mnemonic.

4Hs	4Ts
Hypoxia	Tension pneumothorax
Hypovolaemia	Cardiac Tamponade
Hypo/hyperkalaemia	Toxicity
Hypothermia	Thromboembolism

4. Other points

Drugs

By far the most important treatment in resuscitation is good quality basic life support with continuous chest compression and effective, but not excessive, lung inflation. The next important action is to remove any reversible precipitating causes. Although drugs are commonly administered, there is little evidence to support routine administration of many of them. As the tracheal route of administration is largely ineffective, circulatory access had to be achieved rapidly; this is most effectively performed by intraosseous cannulation unless a peripheral vein can be accessed immediately.

Oxygen

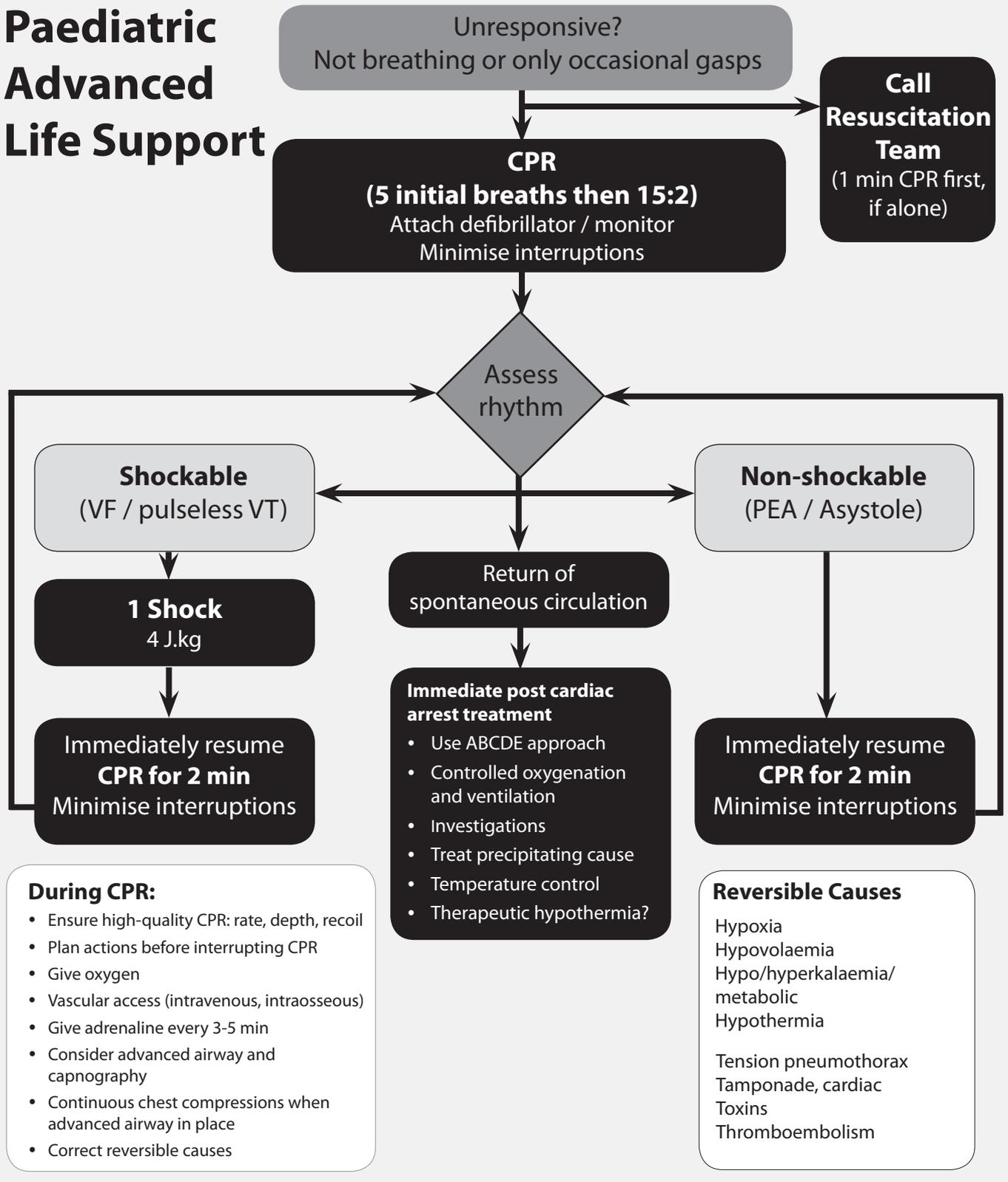
This is the most important drug in paediatric resuscitation as many arrests in children are due to hypoxia. Although high concentrations are often used, effective airway opening and lung inflation are by far the most important steps in achieving adequate oxygenation.

Epinephrine (adrenaline)

Epinephrine been shown to increase the chances of restoring spontaneous circulation and should be administered in a dose of 10mcg.kg⁻¹ every 3-5 minutes during resuscitation. Larger doses have not been shown to be effective and should not be used.



Paediatric Advanced Life Support



During CPR:

- Ensure high-quality CPR: rate, depth, recoil
- Plan actions before interrupting CPR
- Give oxygen
- Vascular access (intravenous, intraosseous)
- Give adrenaline every 3-5 min
- Consider advanced airway and capnography
- Continuous chest compressions when advanced airway in place
- Correct reversible causes

Reversible Causes

- Hypoxia
- Hypovolaemia
- Hypo/hyperkalaemia/
metabolic
- Hypothermia
- Tension pneumothorax
- Tamponade, cardiac
- Toxins
- Thromboembolism

Figure 2. Reproduced by kind permission of the European Resuscitation Council and available at: www.resus.org.uk/pages/palspost.pdf

Sodium bicarbonate (NaHCO₃)

Bicarbonate neutralises acidosis by releasing carbon dioxide. During resuscitation, this cannot be cleared as there is insufficient pulmonary gas exchange, consequently, it has not been shown to be effective and should not be used routinely. NaHCO₃ may be indicated in specific circumstances such as hyperkalaemia or in drug toxicity (e.g. tricyclic antidepressants).

Calcium has not been shown to be effective in resuscitation and it may even be harmful, consequently it should not be used routinely. It may however, be effective in hyperkalaemia, hypocalcaemia and calcium receptor blocker overdose.

Amiodarone (5mg.kg⁻¹) has been shown to be the most effective anti-arrhythmic in resistant VF or pVT but lidocaine is an acceptable alternative. Amiodarone is incompatible with saline and should be diluted in 5% glucose.

OUTCOMES

Although it is often thought that children have extremely poor outcome after cardiac arrest, this is not entirely true. Large North American databases have shown that children that have a full cardiac arrest in a hospital have a 27% chance of survival to discharge and that 75% of these will have a good neurological outcome. Out of hospital resuscitation has poorer survival rates, but these figures are significantly

biased by infants with sudden infant death syndrome (SIDS). Older children and adolescents have survival rates of about 9%.

Children with respiratory arrest only who haven't progressed to full cardiac arrest have an excellent chance of survival with about 70% alive after 1 year.

SUMMARY

The most important intervention in paediatric resuscitation is early recognition of the child at risk of deterioration and the instigation of treatment intended to prevent progression to cardio-respiratory arrest.

Once cardio-respiratory arrest has occurred, early and good quality CPR is the most important step for a favourable outcome. Interruptions in chest compression should be avoided and compressions can be continuous once the trachea is intubated. Reversible causes should be actively sought and treated as many paediatric arrests are secondary to another event.

REFERENCES

Bingham B. Paediatric life support. Update in Anaesthesia 2009 25(2):31-37. Available from: http://www.wfsahq.org/components/com_virtual_library/media/4ffc7f76113dd649ce4ff5a2b1dd607c-Paediatric-Life-Support--Update-25-2-2009-.pdf

Resuscitation Council UK. <https://www.resus.org.uk/resuscitation-guidelines/>

Resuscitation at birth

Reprinted with updates for 2013 algorithms from: Richmond S. Resuscitation at birth. *Update in Anaesthesia* 2009; **25**(2): 65-8.

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INTRODUCTION

Resuscitation of a newborn infant at birth is straightforward, and much more likely to be successful than resuscitation of a collapsed adult. The principles underlying the approach are simple. The issue is not complicated by a need to interpret ECGs or to manage arrhythmias. Babies are well adapted to withstand the periods of intermittent hypoxia which are a feature of normal labour and delivery. At term, their hearts are packed with glycogen and, by switching to anaerobic respiration if necessary, can maintain some circulation for up to about 20 minutes in the face of anoxia.

Of those few neonates who get into difficulties, the vast majority will recover rapidly once their lungs have been successfully inflated. However, it is necessary to be aware of some important differences between babies at birth and adults. It is equally necessary to maintain a logical approach, evaluating and completing each step before proceeding to the next.

NEONATES COMPARED TO OLDER CHILDREN

One obvious difference between babies and older children or adults is that babies are small and have a large surface area to weight ratio. They are also always born wet which means they are particularly prone to rapid evaporative heat loss. The initiating insult will virtually always be an interference with placental respiration but the condition that a baby is born in can vary from healthy to extremely sick and all shades between. Perhaps the most important difference to remember is that a baby at birth is in transition from placental to pulmonary respiration. It will therefore have fluid-filled lungs that have never yet been inflated with gas.

COMMENTARY ON ALGORITHM

Let us now approach the algorithm.¹ This algorithm deals primarily with term infants. To some extent this approach can be extended to preterm infants in similar difficulty. The management of transition in significantly preterm infants is also often referred to, confusingly, as “resuscitation”; it is beyond the scope of this article but is covered in detail in the Newborn Life Support manual.¹

1. Heat loss

The first item addresses the issue of minimising heat loss. The baby should be received into warm towels and rapidly dried. Remove the now-wet towels, cover the baby in warm dry towels and, ideally, place the baby on a flat surface under a radiant heater. This will take 20 to 30 seconds during which time you can also begin to assess the condition of the baby.

2. Assessment

The baby then needs to be rapidly assessed. A healthy baby will:

- Adopt a flexed posture with good tone
- Have a normal heart rate which rapidly rises to above 100 beats per minute (bpm)
- Cry and breathe normally within about 30 seconds of delivery
- Though born blue, will rapidly become pink even though the extremities will remain somewhat cyanosed.

An asphyxiated baby will:

- Be very floppy
- Have a slow or even absent heart rate
- Make no attempt to breathe or may give only a shuddering gasp
- Remain blue, or maybe appear very pale due to restriction of blood flow to the skin in an attempt to maintain central circulation

You will certainly need help if the baby is like this.

Of these four attributes the most indicative of a serious problem is tone.

A floppy baby is in serious difficulty, a baby with good tone is not.

A floppy baby with a low heart rate is in serious difficulty whereas a baby with a slow heart rate but good tone is probably OK.

Summary

A floppy baby is unconscious - a baby with good tone is not.

Good airway management and effective rescue breaths are key to achieving oxygenation of fluid-filled lungs.

Chest compressions and drug administration are rarely needed.

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Newborn Life Support

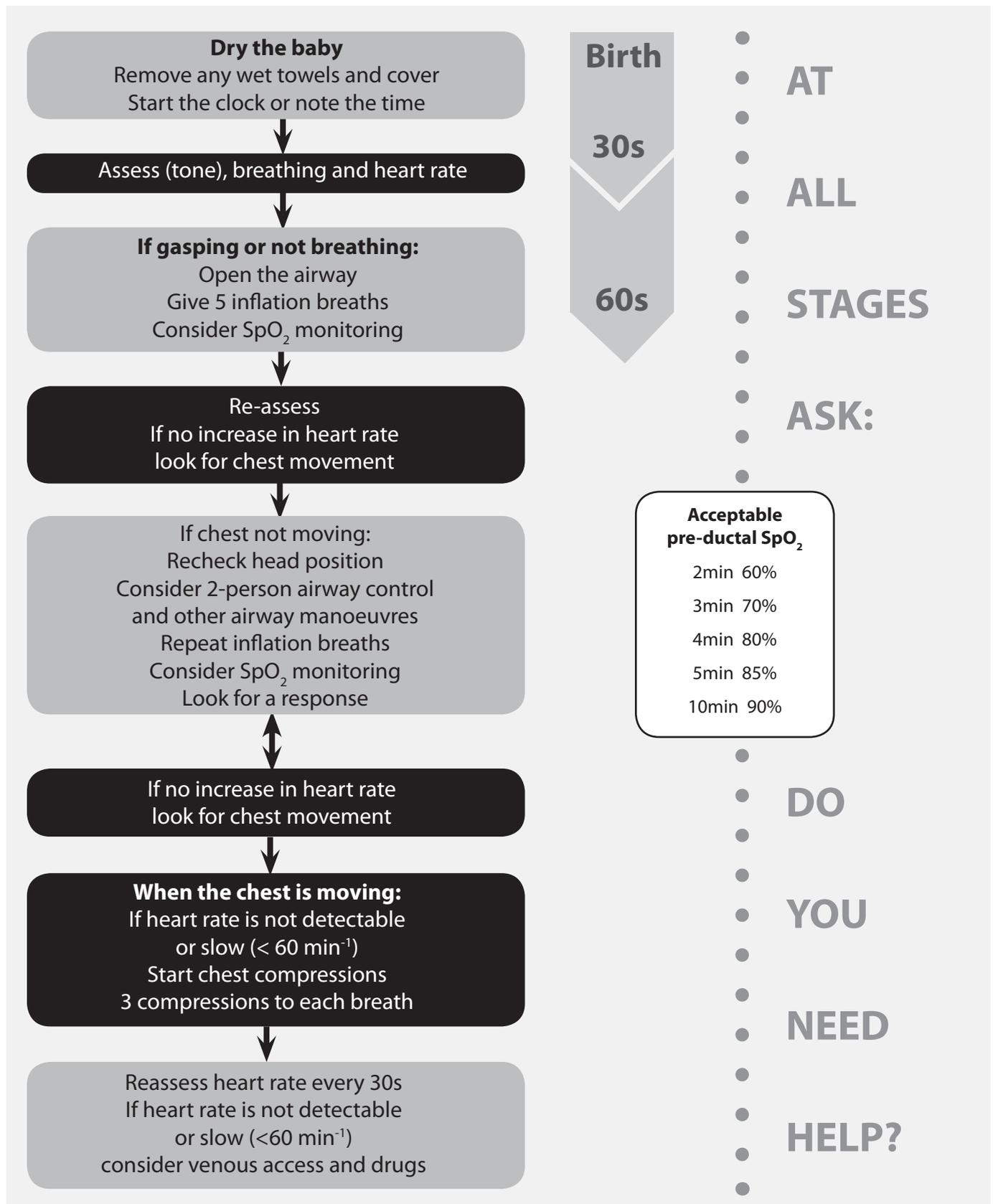


Figure 1. Reproduced by kind permission of the European Resuscitation Council and available at: <http://www.resus.org.uk/pages/nlsalgo.pdf>

The next most important attribute is heart rate. In a baby in difficulty the heart rate will almost instantly respond as soon as oxygenated blood reaches the heart. This will give you the first sign that your resuscitative efforts are having a positive effect. You therefore need to know what the heart rate is at the start so as to be able to judge whether it has later improved.

ABCD

From here on the algorithm follows a familiar pattern – Airway, Breathing, Circulation and Drugs. It is vital that you deal with these items in sequence. In adult collapse ‘compression only’ CPR may be effective because the primary problem is commonly cardiac. In babies the problem is a respiratory one. Applying chest compressions before inflating the lungs merely attempts to circulate blood through fluid filled lungs where it has no hope of acquiring oxygen and is a time-consuming distraction.

3. Airway

An unconscious baby placed on its back will tend to obstruct its airway due to loss of tone in the oropharynx and jaw. This allows the tongue to fall back to obstruct the oropharynx. This tendency is exacerbated by the relatively large occiput of the newborn baby which will tend to flex the neck. In order to open the airway of a baby the head is best held in the neutral position with the face supported parallel to surface on which the baby is lying. Over-extension of the neck is likely to obstruct the airway, as is flexion.

Supporting the jaw and, in very floppy babies, providing formal jaw thrust is sometimes necessary. Given the relatively large size of the newborn baby’s tongue compared to size of the mouth an oropharyngeal airway may also be helpful.

Special case – meconium aspiration

Some babies who get into difficulties before delivery may pass meconium in utero. If insulted further, they may inhale this meconium into the oropharynx or airways during episodes of anoxic gasping before birth. In a baby who is born through heavily meconium stained liquor and who is unresponsive at delivery – and only if unresponsive^{2,3} – it is worth inspecting the oropharynx and removing any thick particulate meconium by means of a large bore suction device. If the infant is unresponsive and you have the appropriate skill then intubating the larynx and then ‘hoovering out’ the upper trachea by applying suction to the tracheal tube during withdrawal may remove a potential blockage. Attempting to remove meconium or other endotracheal blockages by passing a suction catheter down through the endotracheal tube itself is unlikely to be successful as the bore of the catheter will be too small for the purpose.

4. Breathing

If the baby has not yet responded then the next step is to ventilate the lungs. Remember the lungs will be fluid filled if the baby has made no attempts to breathe. Apply a well fitting mask to the mouth and nose and then attempt to inflate the lungs with air at a pressure of around 30 cm of water aiming for an inspiratory time of 2 to 3 seconds. Five such ‘inflation breaths’ will usually be successful in aerating the lung to an extent that will allow any circulation to bring some oxygenated blood back to the heart producing a rapid increase in heart rate.

5. Circulation - re-evaluate heart rate

Having given five inflation breaths you should then assess whether the heart rate has increased. If it has then this is a firm indication that you have aerated the lung. This also tells you that all that is necessary is for you to gently ventilate the baby until it starts to breathe normally. A rate of 30 or so ‘ventilation breaths’ per minute, each with an inspiratory time of around one second, will usually be sufficient to maintain the baby’s heart rate above 100bpm during this period.

If the heart rate has not improved, you need to know whether this is because your attempts at lung aeration have not been successful – the most likely reason – or have you actually succeeded in aerating the lungs but the circulation has deteriorated to such an extent that this alone is not going to be sufficient. The only way to judge this is to see if you can detect passive chest movement in response to attempts at lung inflation. Is the chest moving when you try to inflate it?

Initial chest movement is likely to be subtle and you may have to stoop down and look carefully from the side during further attempts at inflation to be sure on this point. The commonest error is to assume successful chest inflation when it is not present. It is absolutely crucial that this question is answered correctly. If you assume that you have inflated the lungs when you have not, then proceeding to chest compressions will not have any hope of success and you are merely wasting time. Equally, if you assume you haven’t inflated the chest when you have then you will fail to initiate chest compressions when they are necessary and will also waste precious time. The one saving grace is that if you actually have inflated the chest then the rapidly improving chest compliance will make chest movement easier to see with subsequent imposed inflations so chest movement should eventually become obvious.

If chest movement is not seen then the airway is the problem and this must be addressed before going any further. Unless and until the lung is successfully inflated nothing else will have any chance of success. Apart from checking for obvious problems such as failure to switch on the air supply or a big leak from the mask, check the following issues:

Consider:

- Is the baby truly being supported in the neutral position?
- Is jaw thrust necessary?
- Would an oro-pharyngeal airway be helpful?
- Might you achieve better airway control with two people controlling the airway?
- Are you actually delivering an appropriately long inspiratory time?
- Might there be a blockage in the oro-pharynx or trachea?

The presence of meconium on a collapsed baby may give a clue to a blocked airway. It is well known that other less obviously visible substances such as blood clots, lumps of vernix or thick mucus plugs can equally be inhaled and block the airway in exactly the same way.⁶

Once chest movement has been achieved – and only then – consider chest compressions if the heart rate remains slow or absent.

6. Chest compressions

If the heart rate has not responded to lung inflation alone then a brief period of chest compressions may be all that is necessary to bring a little oxygenated blood from the lungs back to the coronary arteries which will then produce a rapid cardiac response. The most effective way to perform chest compressions is with both hands encircling the chest. Place the thumbs together centrally over the lower sternum with the fingers overlying the spine at the back, briskly compress the chest between fingers and thumbs at a rate of about 120 beats per minute. Current advice is that you should intersperse breaths at a rate of one breath for every three beats during this manoeuvre though there is no clear evidence as to the most appropriate compression:inflation ratio.

The need to proceed as far as this is relatively rare – probably around 1 in 1000 births – and the length of time compressions are needed is also relatively short – a few minutes at most.⁷

Having given 30 to 60 seconds of chest compressions you should look for a response. Once again look for an increase in heart rate which indicates successful delivery of oxygenated blood to the heart. Virtually all babies will have responded by this stage. Because this is the expectation it is important to check once again that lung inflation has definitely been successful and that chest compressions are being delivered as expected before deciding that further intervention is needed. However, if the heart rate remains slow – less than about 60 beats per minute - or is absent then consider further intervention.

7. Drugs

What else is available? There is very little published evidence to support any of the drugs which have been suggested for use at this stage. Epinephrine (adrenaline) is traditional in these situations and, if given centrally - ideally via an umbilical venous cannula – does improve coronary artery perfusion pressure in animal experiments. Early animal studies by Geoffrey Dawes and others appear to show a possible place for the use of alkalinising agents such as bicarbonate and dextrose - again given centrally - in boosting a failing circulation at this point. Intuitively one might expect that babies who are seriously hypovolaemic, perhaps from blood loss, would respond to appropriate fluid expansion. If any of these manoeuvres are to be employed then it is necessary to rapidly establish central venous access. This is easily done by inserting a catheter into the umbilical vein.

It must be said that babies who appear to require this degree of help to survive are at very high risk of permanent and severe neurological damage, if they survive at all. Those with the least risk will be those

who have undergone a severe but sudden and recent insult rather than those whose insult has been intermittent and chronic.

8. Post-resuscitation care

Watch babies who have been successfully resuscitated for signs of hypoxic-ischaemic encephalopathy. Those showing such signs should be offered therapeutic hypothermia.⁸

SUMMARY

Resuscitation of babies at birth relies upon good airway management and effective lung inflation with the need to add chest compressions on very rare occasions. Air is all that is necessary for lung inflation and drugs have a very limited place.

REFERENCES

Richmond S. Resuscitation at birth. Update in Anaesthesia (2009) 25(2):65-68. Available from: http://www.wfsahq.org/components/com_virtual_library/media/b4866481ef4d3dec79740f12a3a50482-Newborn-Life-Support-Update-25-2-2009-.pdf

1. Richmond S, ed. Newborn Life Support. Resuscitation at Birth. 3rd ed Resuscitation Council (UK), London, 2011.
2. Wiswell TE, Gannon CM, Jacob J et al. Delivery room management of the apparently vigorous meconium stained neonate: results of a multicenter international collaborative trial. *Pediatr* 2000; **105**: 1-7.
3. Vain NE, Szyld EG, Prudent LM et al. Oropharyngeal and nasopharyngeal suctioning of meconium-stained neonates before delivery of their shoulders: multicentre, randomised controlled trial. *Lancet* 2004; **364**: 597-602.
4. Wyllie J, Perlman J, Kattwinkel J et al. 2010 International consensus on cardiopulmonary resuscitation and emergency cardiovascular care science with treatment recommendations: Neonatal resuscitation. *Resuscitation* 2010; **81S**: e260-87.
5. Dawson JA, Kamlin COF, Vento M, et al. Defining the reference range for oxygen saturation for infants after birth. *Pediatrics*. 2010; **125**:e1340-7.
6. Maskrey S. Neonatal resuscitation. *Clinical Risk* 2008; **14**: 46-8.
7. Perlman JM, Risser R. Cardiopulmonary resuscitation in the delivery room. *Arch Pediatr Adolesc Med* 1995; **149**: 20-5.
8. Perlman JM, Davies P, Wyllie J et al. Therapeutic hypothermia following intrapartum hypoxia- ischemia. An advisory statement from the Neonatal Task Force of the International Liaison Committee on Resuscitation. *Resuscitation* 2010; **81**:1459-61.
9. <https://www.resus.org.uk/resuscitation-guidelines/>

Anaphylaxis; recognition and management

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INTRODUCTION

Anaphylaxis has been defined as 'a serious allergic reaction that is rapid in onset and may cause death'.¹ Rates of allergy and anaphylaxis in low-income countries appear to be low compared to high-income settings, although the incidence appears to be increasing worldwide, and anaphylaxis is becoming more common in children. A survey by the World Allergy Organisation (WAO) found that essential drugs used in the assessment and management of anaphylaxis, with the exception of adrenaline, are not universally available to healthcare providers, and clinical guidelines were in use in only 70% of surveyed nations.² This article will describe recognition and management of anaphylaxis in children, with reference to the UK Resuscitation Council Guidelines.

EPIDEMIOLOGY

Accurate information on the prevalence of perioperative anaphylaxis in children is difficult to find. The condition is likely to be both under-diagnosed and under-reported. The incidence of all anaphylactic reactions in children and adolescents has been estimated as 10.5 in 100,000 or higher.³ Perioperative anaphylaxis is thought to occur in around 1 in 10,000 anaesthetics in children. Asthma, family history, multiple surgeries, latex exposure and food allergy are all risk factors.⁴ Mortality rates can be significant, with up to 10% of all reported anaesthesia-related reactions having fatal outcomes, although it is likely that less severe reactions go unreported.⁵ Asthma is an important risk factor for both the occurrence and severity of reaction. Most fatal cases of anaphylaxis are seen in patients with asthma. Variations in diagnostic criteria and reporting rates raise doubts over the true incidence and outcomes in anaphylaxis treatment. Certainly, the incidence of allergy and the number of prescriptions for self-administered adrenaline (e.g. EpiPen) is increasing.^{6,7}

PATHOPHYSIOLOGY

Anaphylaxis is an IgE mediated type I hypersensitivity reaction, which occurs after exposure to a foreign molecule/antigen, and results in mast cell degranulation and histamine release. The clinical syndrome of anaphylaxis is much more complex

and comes from the cascading release of many vasoactive substances including histamine, tryptase, leukotrienes, cytokines, platelet activating factor and prostaglandins.

Initial antigen exposure results in the formation of specific IgE antibodies on mast cells. Second exposure allows binding of an antigen with IgE antibodies on the presensitised mast cells. The resulting antigen-antibody complex leads to the degranulation of mast cells and massive chemical mediator release, which results in the classical features of:

- Airway oedema
- Bronchoconstriction
- Increased vascular permeability
- Vasodilatation/hypotension

Other mechanisms are described, with 'non-IgE mediated' responses often being labelled as anaphylactoid reactions. These reactions do not require antigen pre-sensitization, and can involve direct mast cell/basophil interactions or complement activation, but still result in massive chemical mediator release. IgE and non-IgE reactions are clinically indistinguishable in their presenting features and do not differ in their management. The term 'anaphylactoid' has now largely been abandoned.

COMMON ALLERGENS IN CHILDREN

Food

Food allergy is the commonest cause of anaphylaxis in children.⁸ A 5-year retrospective study in Australia found 85% of paediatric admissions to the emergency department for an allergic reaction were following exposure to a food related allergen.⁹ Peanuts, fish, milk, eggs and shellfish are most commonly identified triggers, although any food can be implicated. Worldwide variation in common food allergens is seen. Of particular interest to the anaesthetist is the association between egg allergy and propofol (discussed below). Some children outgrow their food allergy; hypersensitivity to allergens such as nuts and shellfish remain throughout life and are commonly associated with more severe reactions.¹⁰

SUMMARY

Anaphylaxis is a life threatening condition

Prompt recognition and optimal management reduces adverse outcomes

Follow basic life support and anaphylaxis guidelines

Avoid further allergen exposure, administer oxygen and intravenous fluid and raise the patient's legs. These simple measures are useful in the management of anaphylaxis

Prompt administration of adrenaline is the most effective treatment in anaphylaxis

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COMMON PERIOPERATIVE ALLERGENS

Common allergens encountered in the perioperative period include neuromuscular blocking agents, antibiotics and latex. These account for the majority of perioperative reactions. Radiological contrast, colloid based intravenous fluids, dye and chlorhexidine anti-septic solutions are all potential causative agents.

Neuromuscular blocking agents

Neuromuscular blocking agent (NMBA)-related reactions account for more than 60% of anaphylactic reactions in the perioperative period. All NMBAs are potentially allergenic, and cross-reactivity amongst them is common.¹¹ Suxamethonium is more likely to cause anaphylaxis than any of the non-depolarising agents. The risk of anaphylaxis with different NMBA has been suggested to be as follows¹²:

- High risk: suxamethonium, rocuronium
- Intermediate risk: vecuronium, pancuronium
- Low risk: atracurium and its isomer, cisatracurium

Controversy surrounds the risk of anaphylactic reaction to rocuronium. Some studies claim it to be a high-risk allergen while others suggest that it is an intermediate risk agent and that increased reaction rates merely reflect increased frequency of use.¹³ Non-immune histamine release is seen with atracurium and other benzylquinolonium compounds. Anaphylaxis during first time exposure to NMBAs is also common. Sensitisation is thought to be due to exposure to other compounds with a quaternary ammonium ion, found in common household products such as cosmetics, toothpaste, cough syrup and detergent.

Antibiotics

Antibiotics account for up to 15% of all reactions occurring under anaesthesia and up to a third of all adverse drug reactions in the paediatric population. Rates seem to be increasing.^{12, 14} Penicillins and cephalosporins are commonly used in perioperative care and are the most frequent cause of drug-related hypersensitivity reactions in children. The two agents have a shared β -lactam ring, and cross-reactivity rate of 10% between the two classes of drug is often quoted, but is now discounted by many experts.¹⁵ It is prudent to avoid cross-exposure in those with previously documented anaphylaxis to either agent. Fortunately, anaphylactic reactions to other broad-spectrum antibiotics such as clindamycin and gentamicin are rare.

Induction agents

Propofol is formulated in a lipid emulsion of soya oil, glycerol and egg phosphatide and is a commonly used induction agent. The egg-based constituent of propofol is a highly purified phosphatide, lecithin, whilst the allergens responsible for hypersensitivity reactions are the egg white proteins: ovoalbumin, ovotransferrin and ovomucoid. Propofol allergy is well documented, but a direct causal relationship between propofol and egg allergy has not been demonstrated. Manufacturers suggest a cautious approach is best in those with egg-related anaphylaxis, but propofol has been widely administered to egg allergic patients without incident.¹⁶

Thiopentone related anaphylaxis is recognised and becomes more

likely with repeated usage. Etomidate hypersensitivity is exceedingly rare. Ketamine use is increasing in hospital and pre-hospital settings and has been a common sole anaesthetic agent in the developing world for many years. Allergic reaction to ketamine is also rare. Thus both ketamine and etomidate provide a good anaesthetic option for patients with previous anaphylaxis from an unidentified agent.

Other anaesthetic agents

Local anaesthetic allergy is rare. Reported reactions are more likely to be caused by accidental intravascular injection or reaction to preservative. Inhalational anaesthetic agents are the only agents in perioperative practice not to have associated immune-mediated allergic reactions.

Latex

Latex hypersensitivity is increasingly reported, and as a result, many healthcare institutions have moved to a latex free clinical environment. Perioperative reaction rates are now falling in areas where this has been achieved. The following groups of children are at high risk for latex allergy:¹⁷

- Multiple operations
- Surgery in the neonatal period
- Atopic children
- Spina bifida
- Cerebral palsy.

There is also recognised cross reactivity between latex and food such as kiwi, banana and avocado.¹⁸ Hospitals should have clear policies for latex allergic patients. Staff should have good knowledge of latex products and the latex-free alternatives. Medical staff should use latex free products where possible to avoid sensitisation of themselves and their patients.

Chlorhexidine

Chlorhexidine is a chemical antiseptic used for skin preparation in surgery, and is also present in a number of different household products such mouth washes, antiseptic wipes, eye drops, and as a coating for medical devices such as urinary catheters, central lines and antiseptic dressings. Anaphylaxis to chlorhexidine has been reported in those with a known allergy to chlorhexidine, but where the presence of chlorhexidine was not recognised, for instance in a medical device.

RECOGNITION OF ANAPHYLAXIS

Anaphylaxis is an acute, severe multisystem disorder. It varies in its presentation and severity and so a high index of suspicion is required. Clinical diagnosis is aided by history and clinical context. It is important to recognise that anaphylaxis can occur on the first exposure to a drug and within seconds of administration, particularly if given by the intravenous route. The vast majority of anaphylactic reactions occur around induction of anaesthesia. Symptoms and signs evolve within seconds or minutes of allergen exposure. The chief difficulty in managing perioperative anaphylaxis has often been to distinguish it from other serious adverse reactions during surgery and anaesthesia, shown in Table 1. Anaphylaxis must always

be considered in the event of perioperative airway obstruction, bronchospasm and hypotension.

Dermatological signs may be the first to appear and can include pruritis, urticaria, erythema, flushing or angioedema. Over 80% of paediatric reactions have skin manifestations. In the absence of skin manifestations the diagnosis can be overlooked in favour of an alternative event. Even when present, these classical signs may be missed as access for examination is limited by surgical drapes or impeded by poor theatre lighting. Signs may also be less obvious in pigmented skin.

Respiratory manifestations occur in over 90% of children and are the most worrying. Laryngeal swelling and bronchospasm may rapidly cause hypoxia. If the child is conscious, they may initially develop hoarseness or complain of a tingling throat. They can rapidly develop stridor and upper airway obstruction.

Cardiovascular effects such as hypotension are less common signs in children, only found in between a quarter and a third of cases. Tachycardia and hypotension indicate more severe reactions. Cardiovascular collapse is a late sign, which occurs peri-arrest. It is usually due to hypovolaemia due to both profound vasodilatation and increased capillary permeability leading to fluid leakage.

Gastrointestinal symptoms such as abdominal pain, nausea, vomiting and diarrhoea may also be seen in non-anaesthetised children.

A diagnostic tool has been proposed to improve identification of patients with anaphylaxis.¹ This is outlined in Table 2. It is proposed that following these criteria will identify over 90% of reactions, leading to early treatment and thus improved outcome.

ALGORITHMS AND GUIDELINES

There are many published guidelines and management algorithms in the literature. Many nations adopt those produced by their own national societies and expert panels. In 2011, the World Allergy Organisation (WAO) Anaphylaxis guideline was created following a lack of a single global template for anaphylaxis management

Table 1. Differential diagnosis of anaphylaxis

-
- Depth of anaesthesia (too deep i.e. hypotension)
 - Depth of anaesthesia (too light i.e. bronchospasm)
 - Inhaled foreign body
 - Acute asthma
 - Drug induced histamine release (opioids, atracurium etc)
 - Flushing syndrome (red man syndrome with vancomycin)
 - Pulmonary /air embolism
 - Vagal syncope
 - Hypovolaemic shock
 - Septic shock
-

Table 2. Clinical criteria for diagnosing anaphylaxis (adapted with permission from Sampson HA,¹ included with permission of Sampson HA & Elsevier)

Anaphylaxis is highly likely when any ONE of the following three criteria is met:

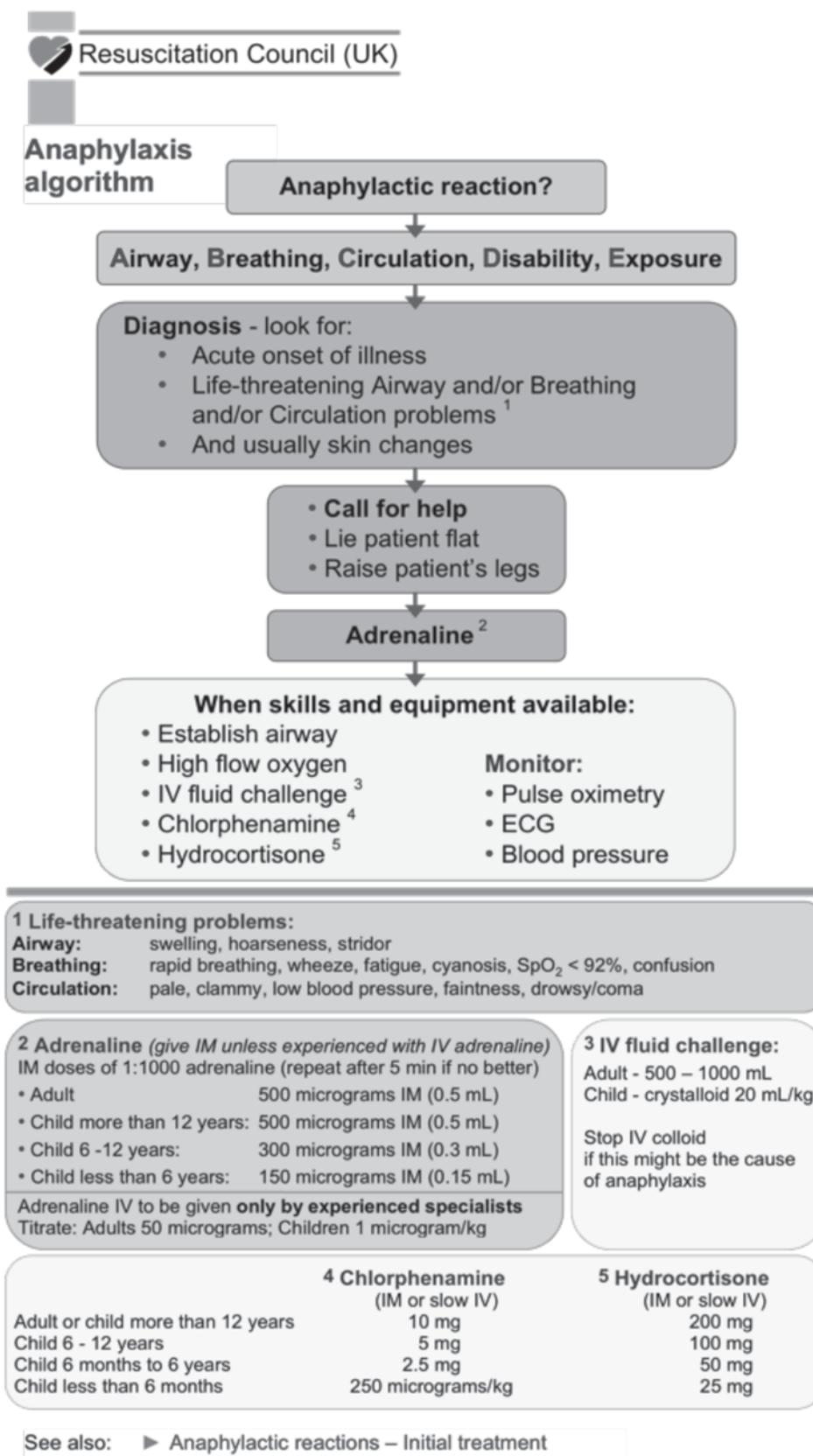
1. Acute onset of illness (minutes to several hours) with involvement of the skin, mucosal tissue, or both (e.g. generalised urticaria, itching or flushing, swollen lips-tongue-uvula) and at least ONE of the following:
 - Respiratory compromise (e.g. dyspnoea, wheeze, bronchospasm, stridor, reduced PEF, hypoxaemia)
 - Reduced blood pressure or associated symptoms of end organ dysfunction (e.g. hypotonia [collapse], syncope, incontinence)
 2. Two or more of the following that occur rapidly after exposure to a likely allergen for that patient (minutes to several hours):
 - Involvement of skin-mucosal tissue (e.g. generalised urticaria, itch-flush, swollen lips-tongue-uvula)
 - Respiratory compromise (e.g. dyspnoea, wheeze, bronchospasm, stridor, reduced PEF, hypoxaemia)
 - Reduced blood pressure or associated symptoms (e.g. hypotonia [collapse] syncope, incontinence)
 - Persistent gastrointestinal symptoms (e.g. abdominal pain, vomiting)
 3. Reduced blood pressure after exposure to a known allergen for that patient (minutes to several hours)
 - Infants/ children: low age - specific systolic blood pressure or greater than 30% decrease in systolic pressure
-

and is an excellent resource.¹⁹ The algorithm produced by the UK Resuscitation Council is well presented and concise, making it ideal for display in clinical areas.²⁰ This is shown in Figure 1. The choice of guideline in itself is not important. Of greater importance is that clinical staff are aware and have access to the guideline. They must also have opportunity to rehearse critical incident scenarios in the management of anaphylaxis.

Immediate management

Use an ABCDE approach for assessment. This approach, combined with knowledge of presenting symptoms/signs and implementation of diagnostic criteria, aids a prompt diagnosis. Once anaphylaxis has been recognised, you should also follow an ABCDE approach in management, combined with the rapid implementation of basic measures of care as outlined below (see Figure1). For life threatening or severe reactions, you must do this together with the prompt administration of intramuscular adrenaline. A rapid decision is needed as to whether the surgical procedure is able to continue. It is recognised that many healthcare providers may have limited resources and limited access to drugs and monitoring equipment. It is important to recognise that many reactions can be treated successfully with implementation of simple measures and the early administration of adrenaline alone.

FIGURE 1. Treatment algorithm for anaphylaxis (with permission from Resuscitation Council UK²⁰)



Basic measures

Stop further administration of potential causative agents, administer supplementary oxygen and place the patient supine with legs raised. These are simple measures to implement. These can be instituted whilst extra help, equipment and adrenaline is obtained. If not already in place, obtain appropriate airway management and vascular access. Treat cardiac arrest using the standard resuscitation protocols.

Early adrenaline

The early use of adrenaline is the most important factor in achieving a good outcome. Adrenaline acts on alpha and beta adrenoreceptors and increases systemic vascular resistance, coronary perfusion pressure, cardiac contractility whilst causing bronchodilatation and inhibiting inflammatory mediator release.

Adrenaline 1:1000, at a dose of 0.01ml.kg⁻¹ intramuscularly (IM), is the drug of choice and should be injected into the antero-lateral thigh. Some algorithms have simplified adrenaline dosing to include EpiPen use, with a range of 150micrograms (0.15ml 1:1000 adrenaline) to 500micrograms (0.5ml 1:1000 adrenaline) depending on age (see Figure 1). The IM route is preferred as it confers a better safety profile in the hands of most health professionals.

The intravenous (IV) route should be used with caution. Arrhythmias can be induced if adrenaline is given IV (VF/VT), so ECG monitoring is essential. The intraosseous (IO) route can also be used, using the same dose as the IV route. IO adrenaline should be followed by a saline flush. Ongoing clinical assessment is essential. If ineffective, the IM adrenaline can be repeated at 5-minute intervals with further doses indicated until clinical improvement is achieved.

Airway

It is essential to maintain a clear airway and give oxygen. Early endotracheal intubation is advised if there is any suggestion of upper airway obstruction developing. A range of endotracheal tube sizes should be available to allow for any developing laryngeal oedema and intubation difficulty. Surgical cricothyroidotomy may be required if there is severe oedema or if mask ventilation is not possible.

Breathing

Bronchospasm may be alleviated by IM adrenaline through its action on beta-2 adrenoreceptors. Treatment with a nebulised beta-2 agonist, such as salbutamol 2.5-5mg is useful, although this should not delay administration of adrenaline if it is required. Administration to an anaesthetised patient is described elsewhere (page 61 and reference 21).

Circulation

Obtain vascular access, if not already secured, and begin fluid resuscitation. Change to the IO route if IV access is difficult. Give 20 ml.kg⁻¹ IV bolus of crystalloid (0.9% saline or balanced salt solutions) if the child is hypotensive. Give further fluids titrated to blood pressure, urine output and heart rate. Position the child head down if hypotension persists. This increases venous return, and is useful if IV access has yet to be achieved or if access to IV fluids is limited. If more than 40ml.kg⁻¹ IV fluid is required, consider inotropic support and invasive ventilation.

Manage fluid resistant hypotension with an adrenaline infusion rather than continuing IM injections or intermittent IV boluses of

adrenaline. Titrate IV adrenaline to effect, starting from 0.1mcg.kg⁻¹.min⁻¹ (range 0.1-1.0mcg.kg⁻¹.min⁻¹) to achieve a normal blood pressure. Although adrenaline can be infused peripherally initially, it should be administered via a central venous catheter if possible. An adrenaline infusion can be made by adding 0.3mg.kg⁻¹ adrenaline to 50ml of 0.9% saline or 5% dextrose; an infusion of 1ml.hr⁻¹ = 0.1mcg.kg⁻¹.min⁻¹.

Dopamine, noradrenaline and phenylephrine are acceptable alternatives; noradrenaline has a powerful alpha-receptor agonist effect and should be considered if hypotension is unresponsive to adrenaline. Specialised equipment, monitoring and appropriately trained staff are required if a vasopressor infusion is used: the child should be looked after in an intensive care unit or high dependency unit if possible. Mortality can be high in this patient group, even in well-resourced clinical settings.

Secondary management

Adrenaline is the drug treatment of choice for severe anaphylaxis. Antihistamines and steroids are useful adjuncts for the management of anaphylaxis, but their administration should not delay the use of adrenaline. There is concern that inclusion of agents other than adrenaline in guidelines risks their use as inappropriate first line agents.²³

Histamine (H1) antagonists such as chlorpheniramine (2.5-10mg IM or slow IV, see Figure 1) are useful in minor allergic reactions but their speed of action means they are not appropriate as first line agents. Some guidelines omit them entirely as there is a lack of strong evidence for their use, their effect on outcomes, or in prevention of biphasic reactions.

Steroids, such as hydrocortisone (25-200mg IV depending on age, see figure 1), are often given IV in the treatment of anaphylaxis, but offer little benefit in the acute phase. Intravenous methylprednisolone (1mg.kg⁻¹) has been used in less severe reactions, or where the oral route is still available, prednisolone 1mg.kg⁻¹ PO. Steroids are thought to reduce the risk of biphasic reactions. Biphasic reactions can occur in up to 20% of cases, with most occurring in the first 6 hours. A period of close observation is recommended in a well-staffed and monitored environment. They are more often seen in those patients who have delayed administration of adrenaline, or in those who require repeated doses, so a period of observation is required after stability is achieved.²⁰ A recently published systematic review showed that there is no good quality evidence to support the use of glucocorticoids in this setting, although use in patients with coincidental asthma is still advisable.²³

Investigations and follow-up

Correct identification of triggers for anaphylactic reaction in the perioperative period can be difficult as patients are exposed to multiple drugs and potential causative allergens in a brief period.

Specialist laboratory assays are required to confirm the diagnosis. At present serum tryptase is the only useful blood test commonly available in most modern laboratories.

The half-life of tryptase is approximately 2 hours; levels increase after mast cell activation, peaking rapidly and falling again. It is important that a sample of clotted blood is taken as soon as possible during

the reaction and a second sample 1 to 2 hours later to show the rise and fall in serum tryptase. A third sample is taken at 24 hours to determine baseline tryptase levels and allow interpretation of the earlier results. It is essential to record the times that samples are taken for analysis purposes. Samples that require transfer to another centre for analysis should be refrigerated at +4°C.

Patients who have experienced anaphylaxis under anaesthesia should undergo investigation prior to repeated exposure to anaesthesia. Make detailed records of all drugs, timings and events surrounding the reaction. Ideally, the child should undergo further investigation and immunological testing to identify the causative agent under the guidance of a specialist allergist. This may include skin testing utilising dilute concentrations of drugs. Skin pricks or intradermal injections can be used to look for signs of sensitisation. Specific immunological assays, looking for antigen-specific IgE, are available for a number of drugs. Tests are available for suxamethonium, latex and many commonly used antibiotics, but this is often only available in specialist laboratories. For many this is not achievable due to lack of resource and access to a certified allergist. The Association of Anaesthetists of Great Britain and Ireland (AAGBI) have produced guidelines for anaesthetists detailing the immediate investigation and more specialist tests that may be warranted.⁵

A strategy for any future anaesthesia is very important as patients are at increased risk of another reaction. Future operations should occur in a latex free environment where possible. Regional anaesthesia is ideal and avoidance of NMBAs and any drug previously used or those with recognised cross-reactivity is strongly recommended.

REFERENCES

1. Sampson HA, Munoz-Furlong A, Campbell RL et al. Second symposium on the definition and management of anaphylaxis; summary report: Second National Institute of Allergy and Infectious Disease/Food Allergy and Anaphylaxis Network Symposium. *J Allergy Clin Immunol.* 2006; **117**:391-397.
2. Simons FER, for the World Allergy Organisation. World Allergy Organisation survey on the global availability of essentials for the assessment and management of anaphylaxis by allergy/immunology specialists in healthcare settings. *Ann Allergy Asthma Immunology.* 2010; **104**:405-412.
3. Bohle K, Davis, RL, DeStefano F et al. Epidemiology of anaphylaxis among children and adolescents enrolled in a health maintenance organisation. *J Allergy Clin Immunol.* 2004; **113**: 536-542.
4. Tam JS, Kelly K, Pongracic JA. A seventeen-year review of peri-operative anaphylaxis in a paediatric hospital. *J Allergy Clin Immunol* 2011; **127**: AB247.
5. Association of Anaesthetists of Great Britain and Ireland Safety Guideline. Suspected anaphylactic reactions associated with anaesthesia. *Anaesthesia* 2009; **64**: 199-211 available from http://www.aagbi.org/sites/default/files/anaphylaxis_2009.pdf [accessed 3/6/15].
6. Leiber P, Camargo CA, Bohle K, Jick H, Miller RL, Sheikh A, et al. Epidemiology of anaphylaxis: findings of the American College of Allergy, Asthma and Immunology Epidemiology of Anaphylaxis Working Group. *Ann Allergy Asthma Immunology.* 2006; **97**: 596-602.
7. Sheikh A, Hippisley-Cox J, Newton J, Fenty J. Trends in national incidence, lifetime prevalence and adrenaline prescribing for anaphylaxis in England. *J R Soc Med.* 2008; **101**:139-143.
8. Bock SA, Munoz-Furlong A, Sampson HA. Fatalities due to anaphylactic reactions to food. *J Allergy Clin Immunol* 2001; **107**:191-193.
9. de Silva IL, Mehr SS, Tey D, Tang MLK. Paediatric anaphylaxis; a 5-year retrospective review. *Allergy* 2008; **63**:1071-1076.
10. Sampson HA. Anaphylaxis in children. *Paediatrics* 2003; **111**(6 Pt3): 1601-1608.
11. Krila C, Brunet-Langot C, Labbez F et al. Anaphylaxis during anaesthesia: results of a 12 year survey at a French paediatric center. *Allergy* 2005; **60**: 828-834.
12. Mertes PM, Laxenaire MC, Alla F and GERAP. Anaphylactic and anaphylactoid reactions occurring during anaesthesia in France 1999-2000. *Anaesthesiology* 2003; **99**: 536-545.
13. Rose M, Fisher M. Rocuronium; high risk for anaphylaxis? *Br J Anaesth* 2001; **86**: 678-82.
14. Le J, Nguyen T, Law AV, Hodding J. Adverse drug reactions among children over a 10-year period. *Paediatrics.* 2006; **118** (2): 555-562.
15. Anne S, Reisman RE. Risk of administering cephalosporin antibiotics to patients with histories of penicillin allergy. *Ann Allergy Asthma Immunol* 1995; **74**: 167-70.
16. Murphy A, Campbell DE, Baines D, Mehr S. Allergic reactions to propofol in egg allergic patients. *Anesth Analg* 2011; **113**(1): 140-4.
17. Kagy L, Blaiss MS. Anaphylaxis in children. *Paediatric Ann,* 1998; **27** (11): 727-734.
18. Queiroz M, Combet S, Berard J et al. Latex allergy in children: modalities and prevention. *Paediatric Anaesthesia.* 2009 **19**: 313-319.
19. Simons FER, Arduoso LRF, Bilo MB et al. World Allergy Organization Anaphylaxis Guidelines: Summary. *J Allergy Clin Immunol.* 2011; **127**(3): 587-93.
20. Resuscitation Council UK. Emergency treatment of anaphylactic reactions. www.resus.org.uk/pages/reaction.pdf.
21. Wilson I. Asthma and anaesthesia. Tutorial of the Week 2005;1 available from <http://www.anaesthesiauk.com/documents/Asthma%20and%20anaesthesia%20iii.pdf> (accessed 17/10/14)
22. Ellis AK. Biphasic anaphylaxis: a review of the incidence, characteristics and predictors. *The Open Allergy Journal.* 2010; **3**: 24-28.
23. Choo KJL, Simons FER, Sheikh A. Glucocorticoids for the treatment of anaphylaxis. *Cochrane Database of Systematic | Reviews* 2012, Issue 4. Art No: CD007596.

Accidental poisoning in children

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INTRODUCTION

Poisoning is a significant global public health problem. According to World Health Organisation data, an estimated 350 000 people died from unintentional poisoning in 2002. Accidental poisoning occurs most often in the age group 1-5 years, although less than one per cent of poisoning in children is serious. More than 94% of fatal poisonings occur in low-and middle-income countries. It remains a challenge to identify those at risk at an early stage.

Education of the general public and also the provision of child-proof containers for household chemicals and medicines play a key role in prevention.

(Toxic plants, insect and snake bites are not discussed in this article, but see www.rch.org.au/poisons for more information)

ACCIDENTAL POISONING IN CHILDREN: GENERAL PRINCIPLES

A history of poisoning usually makes the diagnosis obvious but poisoning should be considered in any child presenting with altered consciousness, cardiorespiratory depression, hypothermia, seizures or gastrointestinal symptoms. The possibility of more than one poison should always be considered.

1. Ascertain the nature and time of poisoning.
2. Contact the local Poison Centre for advice (if available). Sources on information are common and include internet-based information databases such as the International Programme on Chemical Safety (www.inchem.org), the UK-based TOXBASE (www.spib.axl.co.uk), or in the USA, TOXNET (toxnet.nlm.nih.gov).
3. Assess the patient following the principles of 'ABC' (Airway, Breathing, and Circulation). If airway protection is impaired, the patient requires intubation.
4. Induced emesis is no longer recommended and is contraindicated with volatile substances.
5. Consider gastric emptying and administration of activated charcoal. Gastric lavage is recommended

if life-threatening poisoning has occurred within the previous hour, but contra-indicated if the airway cannot be protected. Gastric lavage is also contra-indicated in the ingestion of hydrocarbons (risk of aspiration and chemical pneumonitis) and corrosives. Multiple-dose activated charcoal (1g.kg⁻¹ every four hours) is used for substances with a long half life (anticonvulsants, digoxin, and theophylline). The complications of gastric lavage include aspiration pneumonia, hypoxia, mechanical injury to the gut and the induction of hypo- or hypernatraemia. It may also induce charcoal bezoars that may cause intestinal obstruction. Activated charcoal can cause serious complications if aspirated.

6. Remove contaminated clothing and wash patient with soap and water.
7. Send samples for lab investigation (urea, electrolytes, blood glucose.) Urine and gastric aspirates should be saved for later toxicology analysis (where available).
8. Measure relevant drug levels (paracetamol and salicylate and others if available).
9. ECG – important in cases of tricyclic antidepressants and unknown poisons. QRS prolongation is an early sign of cardiovascular involvement
10. Specific antidotes should be given as per instruction by Poison Centre.

SPECIFIC POISONS

Household chemicals

Children frequently ingest household substances and most of these are nontoxic. However, some products contain alcohol which may cause seizures and hypoxia.

Household solutions of bleach contain approximately 10% hypochlorite. They are rarely ingested as they are extremely unpalatable. Commonly, they cause nausea, vomiting and diarrhoea. Less than 100ml of household bleach is unlikely to cause serious problems. Fluids should be encouraged, particularly milk. Oesophageal

Summary

Prevention of childhood poisoning is vital. prevention requires adequate supervision, safe placement of medications, child safe cabinets and containers, blister packaging and education. Most paediatric cases are not severe. Recognition of potentially life-threatening ingestions is important so that appropriate early treatment can be instituted. Aggressive supportive care is essential.

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damage occurs rarely and is associated with concentrated solutions. There is a worrying recent increase in oesophageal injury associated with dishwasher tablets. Removal of these under anaesthesia is a medical emergency.

Ingestion of small quantities of strong alkalis such as drain cleaner containing sodium hydroxide may cause devastating injuries. Oesophageal injury is most common and evolves over the course of a few days. Ingestion causes immediate burning pain, swelling of the lips, and depending on the quantity ingested, salivation, haematemesis, dyspnoea, stridor or shock.

The burn injury is classified by endoscopy:

- mucosal (first degree burn)
 - usually resolves after 2-3 days
- submucosal (second degree burn)
 - requires total parenteral nutrition
- full thickness (third degree burn)
 - shocked, will require intensive care.

Third degree burns may lead to gastrointestinal perforation or airway complications or long term, severe oesophageal stricture causing absolute dysphagia which evolves over a period of several months.

Treatment is supportive; emesis is contraindicated and nasogastric aspiration unhelpful as the injury is oesophageal. Patients with first degree burns may be discharged after 2-3 days once tolerating oral fluids. Patients with third degree burns will be in shock and will require intensive care: tracheostomy is indicated for patients with evidence of supraglottic/epiglottic burns. Steroids may reduce the risk of stricture formation if given within 12-24 hours but their use is controversial. Antibiotics are indicated for perforation. Long term gastrostomy or oesophageal replacement surgery (gastric pull-up) may be required for oesophageal stricture.

Hydrocarbons (paraffin, white spirit)

Most of the hydrocarbons are petroleum distillates, containing a variable amount of saturated and unsaturated aliphatic (open-chain) and aromatic (cyclic) hydrocarbons. The aliphatics are not readily absorbed from the gastrointestinal tract and therefore cause minimal systemic toxicity. Paraffin, petrol, thinners, diesel and benzene are low viscosity aliphatic-based petroleum distillates. The aromatic hydrocarbons are well absorbed, and therefore may cause systemic toxic effects but are less inclined to aspiration-related complications. The main hazard of accidental ingestion of the aliphatic hydrocarbons (paraffin) is that of chemical pneumonitis characterised by ventilation/perfusion imbalance and hypoxia. The aspirated hydrocarbons inhibit surfactant and also cause direct broncho-alveolar injury. This can occur even in the absence of vomiting or impaired consciousness and as little as 1ml aspirated hydrocarbon can result in a chemical pneumonitis.

Paraffin is often sold or stored in unlabeled containers which may be within children's reach. Volumes ingested tend to be small because of a burning sensation in the mouth and the bad taste of the liquid.

Nausea and vomiting are common. Aspiration pneumonitis occurs in 12-40% of patients. Signs and symptoms appear 30 minutes after ingestion but may be delayed for 8 hours. A non-productive cough, tachypnoea and tachycardia are signs of a developing chemical pneumonitis. There may be wheeze, coarse crackles as well as signs of respiratory distress (intercostals and subcostal retraction). Hypoxia and cyanosis may be present in severe cases. Patients may be lethargic or irritable. A fever may also be present. The clinical picture generally deteriorates over the first 24 hours. A temperature that persists or only develops after 24 hours usually suggests a secondary infection.

Abnormalities on the chest film may be seen within 30 minutes after aspiration even in the absence of clinical signs or symptoms. Common radiological findings of chemical pneumonitis include bilateral perihilar infiltrates which progress to form patchy infiltrates and later become large areas of consolidation.

Management of hydrocarbon poisoning

- ABC principles.
- Observe all asymptomatic patients for at least 8 hours. Carefully examine patients for the development of respiratory signs. A chest film at 6 hours is recommended.
- If asymptomatic after 8 hours and the chest film is normal, the patient may be discharged.
- Symptomatic patients should be X-rayed on admission.
- Emesis and gastric lavage is contra-indicated as it increases the risk of aspiration. The use of milk is not recommended, but clear fluids are not contra-indicated.
- Oxygen should be administered to all patients with respiratory signs or symptoms. Other respiratory support modalities should be instigated as required, according to standard indications.
- Corticosteroid therapy may increase the risk of secondary bacterial infection and has not been shown to be of any benefit.
- Antibiotics should be given if there are suggestions of a secondary infection (persistent fever or a fever that develops after 24 hours.)
- Prophylactic antibiotics have not been shown to prevent secondary infection and are not advocated.

PESTICIDES

Paraquat

Paraquat ingestion remains a problem in a large number of countries. Most of these cases are intentional suicide attempts (73% in a Malaysian study) or occupational exposure and therefore not that frequently seen in the paediatric population. It is the most toxic herbicide known, producing multi-organ failure. After oral ingestion, patients develop a severe gastroenteritis with oral, oesophageal and gastric ulceration. Depending on the dose ingested, multi-organ failure may develop within 48-72 hours. As little as 10ml may be fatal. Those who survive the initial phase develop pulmonary fibrosis

as a response to the acute alveolitis in the first phase. This leads to respiratory failure and patients die of anoxia.

Management of paraquat poisoning

- Any history of oral exposure should be considered potentially fatal. Treatment strategies are aimed at prevention of absorption, enhancement of elimination and prevention of pulmonary damage.
- Treat immediately with Fuller's earth (Fuller's earth 300g and magnesium sulphate 50g in a litre of water). Give 5-10ml.kg⁻¹ orally every 2-3 hours for 2-3 days.
- Activated charcoal may be used if Fuller's earth is not available. Mix with saline and a laxative such as lactulose to prevent constipation. Repeat every 4 hours until paraquat is no longer detected in the urine.
- If neither is available, careful gastric emptying may be done but be aware of possible pharyngeal and oesophageal ulceration and perforation.
- Haemoperfusion or dialysis may be used (if available).
- Appropriate management of respiratory complications includes PEEP/CPAP. Low FiO₂ should be used as high concentrations of oxygen lead to worse pulmonary toxicity. Paraquat accumulates in the lung where it generates superoxide anions through the reaction with oxygen.

Organophosphate poisoning

Organophosphate poisoning (OP) remains an issue in developing countries. In one study, 35% of OP poisoned victims were children.

The organophosphates and carbamates are cholinesterase inhibitors, thereby eliciting cholinergic signs and symptoms. They can be absorbed by ingestion, inhalation or via the skin. The organophosphates form an irreversible complex with cholinesterase but the carbamyl-enzyme complex is reversible. Thus a less severe clinical picture of shorter duration is presented. It also causes fewer CNS effects because of less penetration of the blood brain barrier.

OP poisoning causes: SLUDGE; Salivation, Urination, Lacrimation, Defecation, Gastric, Emesis and may progress quickly to CNS symptoms with seizures, especially in children.

- Muscarinic effects – hypersecretion, vomiting, diarrhoea, constricted pupils, bronchoconstriction and urinary incontinence
- Nicotinic effects – muscular weakness, fasciculations (rarer in children) and respiratory muscle weakness (may override muscarinic effect and cause tachycardia, hypertension and mydriasis)
- CNS effects – irritability, seizures, coma (and accompanying respiratory depression).

Management of organophosphate poisoning

- ABC
- Patient may require intubation (avoid suxamethonium because of prolonged effect)

- Gastric lavage, activated charcoal
- Remove all contaminated clothes (take care not to get exposed yourself)
- Wash patient with soap and water (mild alkaline soap helps deactivate organophosphates)
- Start intravenous atropine administration as soon as possible. Initial test dose of 0.01mg.kg⁻¹. Then 0.05mg.kg⁻¹ every 15 minutes until full atropinisation is reached. Maintenance therapy is usually around 0.05mg.kg⁻¹.h⁻¹. Adequate therapy is achieved when there is control of excessive bronchial and oral secretions. Don't worry about the tachycardia – it is well tolerated. Eye signs aren't a good marker of progress. When the patient improves, the dose should be tapered slowly over 24 hours. Atropine should never be stopped abruptly. Close observation is required as rebound organophosphate toxicity may occur due to their lipid solubility.
- Watch out for chemical aspiration pneumonia as a lot of the organophosphate pesticides have a petroleum base.

INHALED POISONS

Carbon monoxide

Carbon monoxide is produced through incomplete combustion processes. It is colourless, tasteless and non-irritant. The commonest sources of carbon monoxide poisoning are smoke inhalation, poorly maintained domestic gas appliances, and deliberate inhalation of car exhaust fumes (the latter less common in children). Carbon monoxide causes tissue hypoxia by the interruption of electron transport in the mitochondria. It also reduces oxygen delivery by competing with O₂ for binding to Hb and altering the shape of the HbO₂ dissociation curve (making the curve less sigmoidal, a shift to the left). Its affinity for Hb is >200-fold that of O₂.

Carbon monoxide causes injury by hypoxia, with symptoms referable to tissues with greatest oxygen consumption, notably the heart and brain. Patients present with hypoxia without cyanosis. Skin and mucosal surfaces may appear cherry red. Symptoms correlate roughly with the COHb level:

Carboxyhaemoglobin (COHb) level	Symptoms
<30%	dizziness and headache
50-60%	syncope, tachypnoea, tachycardia and fits
>60%	increased risk of cardiorespiratory failure and death

Anaemia, increased metabolic rate (e.g. children) and underlying ischaemic heart disease all increase susceptibility to CO. Neurological recovery depends on the duration of hypoxic coma.

Management of carbon monoxide poisoning

- Pulse saturation monitoring does not distinguish between HbO₂ and COHb and will read falsely high.
- Arterial blood gases should be done if available. PaO₂ may be normal but any evidence of metabolic acidosis indicates serious poisoning (useful even in the absence of COHb measuring facilities.) COHb measurements are diagnostic but not always available.
- Treat according to ABC principles.
- Apply tight-fitting mask with 100% oxygen. This reduces the half life of COHb from 320 mins (in room air) to 80 minutes. Intubation and ventilation may be necessary in severe cases.
- Hyperbaric oxygen is useful to further reduce the COHb half life, but access and transfer times to a hyperbaric chamber make it an impractical option.
- Check 12 lead ECG – if there are signs of myocardial ischaemia, the patient needs intubation/ventilation.
- Supportive care should include continuous cardiac monitoring, treatment of arrhythmias and correction of acid/base and electrolyte abnormalities.
- Lab bloods should be done for creatinine and electrolytes, FBC, creatine kinase and cardiac enzymes.
- Brain hypoxia leads to cerebral oedema and fits. Diazepam 5-10mg iv is used to control the fits.
- Initial chest Xray may be normal even in severe smoke inhalation.

Patients need to be followed up as some of the neuropsychiatric complications may take weeks to develop.

Combustion of household materials can also generate a number of other toxic substances such as sulphur dioxide, nitrogen dioxide, acrolein, cyanide and various acids from PVC and polyurethane. This may cause direct lung, skin and conjunctival injury.

Cyanide poisoning

Most commonly seen in victims of smoke inhalation as a combustion product of polyurethane foams. Cyanide derivatives are also used in industrial processes and fertilizers. Children may ingest amygdalin, a cyanogenic glycoside contained in kernels of almonds and cherries. Cyanide acts by irreversibly blocking mitochondrial electron transport.

HCN gas can lead to cardiorespiratory collapse and arrest within a few minutes. Patients surviving to reach hospital are unlikely to have suffered significant poisoning. Early signs include dizziness, chest tightness, dyspnoea, confusion and paralysis, followed by cardiovascular collapse, apnoea and seizures.

Management of cyanide poisoning

- Do not attempt mouth to mouth resuscitation as the skin will be contaminated.

- Apply 100% O₂ with a tight fitting mask. Treatment is supportive.
- Decontaminate the skin with soap and water.
- Gastric lavage could be attempted where there are no signs of cyanide poisoning.
- Dicobalt ededate, sodium thiosulphate and sodium nitrate are used as antidotes.

MEDICINES

Children frequently ingest medicines. Dangerous substances for children include salicylates, paracetamol, iron, theophylline and tricyclic antidepressants.

Aspirin / salicylate poisoning

Commonly ingested by children. Also probably the commonest drug to be ingested deliberately in overdose. Oil of wintergreen is 98% methyl salicylate. Its primary toxic effect is by uncoupling of oxidative phosphorylation.

Patients present with restlessness, hyperventilation, tinnitus, deafness, tachycardia, nausea, vomiting, sweating, hyperthermia and dehydration. Pulmonary oedema, acute renal failure, hypokalaemia, hypoglycaemia and hypothrombinaemia may also develop. In adults there is an early increase in respiration rate causing a respiratory alkalosis that precedes the later development of metabolic acidosis. However this is not seen in children.

Management of aspirin / salicylate poisoning

- Gastric lavage should be attempted, even up to 12 hours or longer post ingestion. In overdose, salicylates may form concretions in the stomach and delay absorption.
- Activated charcoal orally every four hours.
- Severity of the poisoning can be determined by measuring blood salicylate levels 6 or more hours after ingestion but may be misleading in severe acidosis. Ideally should be done on admission and 4 hours later to assess continued absorption. Therapeutic levels of salicylate are generally less than 300mg.l⁻¹. Levels more than 750mg.l⁻¹ represents severe poisoning.
- Check electrolytes. Blood glucose monitoring should be done every 2 hours.
- Dehydration, acidosis, hypoglycaemia and electrolyte disturbances should be corrected. Pay particular attention to potassium. Care should be taken to avoid fluid overload, and renal function should be closely monitored.
- Alkalinisation of the urine (pH 7.5-8.5) by administering sodium bicarbonate (either orally or by infusion) is recommended to increase excretion of salicylates.
- Haemodialysis or charcoal haemoperfusion may be required in severe poisoning (levels > 1000mg.l⁻¹ or 7.25mmol.l⁻¹) or in patients with decreased urine output, pulmonary oedema or progressive deterioration.

Paracetamol overdose

Paracetamol ingestion is common but seldom leads to severe toxicity in children due to the diluted concentration of the paediatric syrup formulation. The children at risk are those with glutathione depletion, such as children with cystic fibrosis, adolescents with eating disorders and those also on enzyme inducing agents such as anticonvulsants. The liver is the main target organ in paracetamol poisoning. Patients are generally asymptomatic up to 24 hours post ingestion. Mild nausea, vomiting and anorexia may occur. Hepatic necrosis becomes apparent in 24-36 hours with right subchondral pain and tenderness, jaundice, vomiting and acute liver failure. Confusion and encephalopathy develop over 36-72 hours. Oliguria and renal failure may occur from acute tubular necrosis in the absence of liver failure. Lactic acidosis may be seen early or late.

Management of paracetamol poisoning

- Patients presenting within 4 hours of ingestion should undergo gastric lavage.
- Activated charcoal should not be given orally if the specific antidote is also given orally.
- Measure paracetamol levels at least 4 hours post ingestion and plot on 'plasma paracetamol concentration – time graph' to determine treatment line.
 - If the initial levels indicate no treatment, repeat after 4 hours to check for delayed absorption.
 - All patients on or above the 'Normal treatment line' should be given N-acetylcysteine.
 - Patients on enzyme-inducing drugs should be treated if above the 'Enhanced risk treatment line'.
- Where levels are not available, assume liver damage after ingestion of a single dose of more than 150mg.kg⁻¹ paracetamol, or a staggered overdose (spread over several hours) of the same amount.
- Acetylcysteine is the antidote of choice and is usually given iv. Oral methionine may be used if the patient is allergic to acetylcysteine and is also a suitable alternative in remote areas if vomiting is not a problem. Oral acetylcysteine and carbocysteine have also been used.
- Monitor urea and electrolytes, PT and LFT.
- Give vitamin K but avoid giving fresh frozen plasma unless there is active bleeding. The PT is the best indicator of the severity of liver failure.
- All patients with encephalopathy or a rapidly rising PT should be referred to a liver unit (where available).
- Supportive treatment and treatment of liver and renal failure as indicated.

Iron poisoning

Ingestion of more than 20mg.kg⁻¹ of elemental iron is considered potentially toxic, and the lethal dose is estimated at about 180mg.kg⁻¹. Iron is extremely irritant. The clinical features can be divided into

three phases (not always very clear). Patients present with vomiting, diarrhoea, abdominal pain, haematemesis and rectal bleeding in the early phase (0-2 hours). This is followed by a period of stabilisation (up to 12 hours) during which a deceptive recovery occurs. This is followed by a life-threatening period during which coma, fits, jaundice, hepatic failure renal failure, clotting abnormalities, hypoglycaemia and cardiovascular collapse may occur.

Patients alive 72 hours after ingestion usually make a full recovery. Late complications of gut stricture, gastric fibrosis and pyloric obstruction have been reported.

Management of iron overdose

- The stomach should be emptied with gastric lavage.
- Blood samples for FBC, glucose, UE, iron levels and ABG where available.
- Serum iron only peaks four hours after ingestion.
- An abdominal X-ray helps to determine the number of tablets ingested and also the success or failure of gastrointestinal evacuation.
- A patient with serum iron >90mmol.l⁻¹ may be treated with a chelating agent.
- If <20mg.kg⁻¹ are ingested, treatment is supportive. Patients with iron level below 54mmol.l⁻¹ and who remains asymptomatic 6 hours post ingestion would not be expected to develop significant toxicity and require no active treatment.
- For ingestion of between 20-60mg.kg⁻¹ elemental iron, gastric emptying may be considered if within one hour of ingestion. Whole bowel irrigation may be used in patients with ingestion of more than 60mg.kg⁻¹ and more than one hour post ingestion. It is especially useful if a slow release preparation has been ingested. Charcoal is of no benefit as iron does not bind to it.
- Desferrioxamine chelates iron and is the recommended treatment.
 - Give 1 gram im every 6-12 hours for children (2g for adolescents). (100mg of desferrioxamine binds 8.5mg of elemental iron)
 - If the patient is hypotensive, give desferrioxamine iv at a rate of 15mg.kg⁻¹.hr⁻¹, until the serum iron falls (maximum daily dose 80mg.kg⁻¹)
- Haemodialysis is indicated in very high serum iron levels that respond poorly to chelation therapy or if the urine output is not maintained. (the iron-chelate is excreted entirely in the urine)

Theophylline poisoning

This type of poisoning is rare but serious. Most preparations are slow release so that problems develop 12-24 hours after ingestion. Features of acute ingestion reflect the local irritant GI effects – nausea, vomiting, haematemesis and diarrhoea. The child may be hyperactive with dilated pupils, hyperreflexia, hypotonia and myoclonus. There may also be severe hypokalaemia, arrhythmias, metabolic acidosis, hyperglycaemia, hypotension and seizures.

Management of theophylline poisoning

- Gastric lavage should be considered if ingestion occurred within 1-2 hours.
- Repeated administration of activated charcoal to prevent further absorption and enhance systemic clearance. (May be difficult in the presence of nausea and vomiting).
- Whole bowel irrigation is considered if a slow-release preparation is ingested.
- Haemoperfusion should also be considered.
- Seizures are treated with diazepam.
- Assess hydration and correct hypokalaemia and electrolyte disturbances.
- Cardiac monitoring to detect arrhythmias.
- Verapamil and propranolol may be required to treat supraventricular and ventricular tachyarrhythmias. Lignocaine appears to have little effect on ventricular ectopy.

Tricyclic antidepressants

Cardiovascular toxicity is the main cause of death in tricyclic antidepressant overdose. It is caused by the blockade of noradrenaline uptake as well as the anticholinergic, membrane-stabilising and alpha-blocking effects. Nervous system toxicity includes drowsiness, agitation, hallucinations, hyperreflexia, myoclonus, rigidity, convulsions, respiratory depression and coma. Anticholinergic effects include flushing, dry mouth, dilated pupils, hyperpyrexia and bladder/bowel paralysis. Cardiovascular toxicity includes sinus tachycardia, hypotension, conduction abnormalities and arrhythmias. Respiratory complications include respiratory depression, aspiration pneumonia, ARDS and pulmonary oedema.

Management of tricyclic overdose

- Gastric emptying is delayed by tricyclic antidepressants, and therefore gastric lavage should be attempted as late as 12 hours post ingestion
- Due to CNS depression and inadequate ventilation, intubation may be required.

- Continue cardiac monitoring for up to 48 hours.
- Inotropic support should be given as necessary.
- Correct metabolic acidosis to enhance protein binding of the drug.
- Control seizures with diazepam.

SUMMARY

Prevention of childhood poisoning is vital. There must be adequate supervision, safe placement of medications, child safe cabinets and containers, blister packaging and education. Most paediatric cases are not severe. Recognition of potentially life-threatening ingestions is important so that appropriate early treatment can be instituted. Aggressive supportive care is vital.

Ribbens S. Accidental poisoning in children. Anaesthesia Tutorial of the Week 95(2008) available from: http://www.wfsahq.org/components/com_virtual_library/media/0bcfbec632c1b044303ba371f9491c6-a96849513ae3b1ef36a9fb85127f0b50-95-Accidental-poisoning-in-children.pdf

USEFUL WEBSITES

- World Health Organisation – The International Programme on Chemical Safety : Poisoning Prevention and Management (www.inchem.org)
- TOXBASE.org www.spib.acl.co.uk (NHS only, requires registration)
- Agency for toxic substances & disease registry: (no password or registration required) <http://www.atsdr.cdc.gov/substances/toxsubstance.asp?toxid=19>
- TOXNET (toxnet.nlm.nih.gov)
- Royal Children's Hospital Melbourne: Victorian Poisons Information Centre (www.rch.org.au/poisons)

Further resources available online or to download

Rachel Homer, Edition Editor

This is not intended to be an exhaustive list, but I've tried to bring together cheap or (majority) free resources which I've found useful or interesting, or which have been recommended to me.

WEBSITES

Safety

<http://www.who.int/patientsafety//safesurgery/en/index.html>

The 2009 edition of WHO surgical safety checklist available to download free in English, French, Arabic, Chinese, Russian or Spanish. Also links to

- The checklist's implementation manual in the same languages
- WHO guidelines for safe surgery (English)
- Pilot study showing mortality benefit [Haynes et al]
- Examples of locally adapted checklists
- Draft translations into further languages
- Other resources (also free).

<http://www.lifebox.org/education/>

Training materials, user manual and trouble-shooting guide developed to support the Lifebox foundation's distribution of free (donated) rugged pulse oximeters to widen access to this critical patient safety monitor. Free to download in English, Arabic, Chinese, Portuguese, French, Spanish, and Russian. Also links to application/needs assessment for a Lifebox pulse oximeter if your hospital does not yet have one.

<http://www.phoneoximeter.org/the-phone-oximeter/>

[Hudson et al] Prototype of software activated by connecting hardware (pulse oximeter probe, cable) to a mobile phone. First use of mobile phone technology for continuous pulse oximetry monitoring. Cost: perhaps \$20 USD; not yet available as at June 2015.

<http://www.saferintubation.com/>

Intubation Checklist pdf is designed to help the whole team, who may be unfamiliar, share a mental model for emergency intubation under stress and in less familiar conditions, such as a major trauma patient in

the emergency department. May also aid preparations in theatre for urgent cases requiring intubation.

<http://www.wakeupsafe.org/index.iphtml>

This is an incident reporting system run by the US Society for Pediatric Anesthesia. Participating institutions [who have to pay a fee to join] share critical incidents so that lessons on safer practice can result. Findings are free to download, and although not designed for a resource-poor setting it may still be helpful to read about common problems and how they have been solved or avoided.

<http://www.aagbi.org/safety/salg/salg-incident-summaries>

The Safe Anaesthesia Liaison Group [SALG] is jointly hosted by the UK's Royal College of Anaesthetists and Association of Anaesthetists of Great Britain and Ireland. Safety incidents across the breadth of anaesthesia [not just paediatrics as for Wake Up Safe] are shared from across England and Wales. SALG produce a quarterly report of recurring themes, as well as safety briefings on specific topics, which again assume a high-income environment but still contain useful learning points. All are free to download.

Education

Not specific to paediatric anaesthesia, but have paediatric content:

<http://www.anaesthesiologists.org/>

World Federation of Societies of Anaesthesiologists homepage. Links from here to *Update in Anaesthesia*, *Tutorial of the Week*, and many other resources.

<http://www.rcoa.ac.uk/e-learning-anaesthesia/sample-sessions>

E-Learning for Anaesthesia (e-LA) is an extensive online resource designed for anaesthetists who work for the UK's National Health Service. It follows the UK Royal College curriculum in anaesthesia. Free registration is only available if you have an NHS email address. Otherwise the programme is available to purchase annual access at high cost [full programme £600/year]. These sample sessions can be completed free, and some other sessions are available on the e-SAFE DVD.

<http://www.rcoa.ac.uk/e-SAFE>

The Royal College of Anaesthetists, Association of Anaesthetists of Great Britain and Ireland and e-LfH [e-learning for healthcare] have developed e-SAFE, an e-learning DVD to support education in anaesthesia throughout the developing world. This DVD contains over 100 interactive e-learning sessions and an e-Library with over 500 articles covering basic science and clinical anaesthesia, including many e-Learning for Anaesthesia sessions [see above]. Online form to request a copy.

<http://www.anaesthesiacases.org/>

Library of online case reports. Requires free registration [link on home page] to read or submit cases.

<http://www.logbook.org.uk/>

Keeping a logbook of your cases can help you reflect on and improve your practice, as well as being a requirement in some countries. This free software works on a variety of systems [take care to download the correct version]. There is also a troubleshooting [help] guide.

<http://www.nysora.com>

Illustrated online tutorials on a range of regional anaesthesia techniques. Tutorials include nerve stimulator and landmark techniques as well as ultrasound-guided for those possessing this technology. Most of these nerve blocks are useful in paediatric patients.

<http://www.biodigitalhuman.com/>

Interactive human anatomy. Requires free registration [email address or facebook account]. Works with Google Chrome and Firefox browsers but not Internet Explorer.

Specifically paediatric anaesthesia

[http://onlinelibrary.wiley.com/journal/10.1111/\(ISSN\)1460-9592/issues](http://onlinelibrary.wiley.com/journal/10.1111/(ISSN)1460-9592/issues)

The journal *Paediatric Anaesthesia* offers many articles and entire issues free; the remainder require a subscription.

Examples of free issues [accessed 08/08/2013] include:

- Anything published in or before 2008
- Volume 19, Issue Supplement s1 (July 2009) Special Issue: The Paediatric Airway
- Volume 19, Issue 10 (October 2009): range of articles
- Volume 22, Issue Supplement s1 (July 2012) Special Issue: Good Practice in Postoperative and Procedural Pain Management, 2nd Edition
- Volume 23, Issue 1 (January 2013): range of articles.

<http://apagbi.org.uk/publications/apa-guidelines>

lists guidelines and advice from the Association of Paediatric Anaesthetists of Great Britain and Ireland. Accessed on 08/08/2013, contains free-to-download guidelines on:

- Pain management
- Difficult airway

- Fluid management
- Nausea and vomiting prevention
- Immunisations.

<http://www.euroespa.com/science-education/specialized-sections/espa-guidelines/> English-language guidelines set compiled by the European Society for Paediatric Anaesthesiology. Divided into Emergency Care, Treatment, and Safety sections. Some are online, others free to download.

<http://www.sickkids.ca/Anesthesia/Pediatric-anesthesia-forum/index.html>

Paediatric anesthesia forum is a discussion board run by Toronto's Sick Kids Hospital and open to anyone worldwide who registers [email address required].

<http://medstation.yale.edu/pedres/files/Resident%20Education/PED/ShockAfricaNEJM.pdf>

The FEAST (Fluid Expansion as Supportive Therapy) trial, a large multicentre randomised trial, examined the effectiveness of fluid resuscitation of children with severe infections in Africa. The results - that children given fluid at maintenance rate did better than those given boluses - surprised both the investigators and the wider paediatric medicine community.

<http://www.virtualpediatrichospital.org/>

APPS

iPhone and iPad users have a wide choice of medical apps. Android phone apps are more limited, but Android versions of some of these are in development. iTunes versions can be downloaded to both Mac and PC computers.

Free copies of latest guidelines available from the iTunes website include:

iRESUS: Latest Resuscitation Council (UK) guidelines and algorithms for adult and paediatric resuscitation, currently 2010. Automatic updates when new guidelines released. Review and screenshots at <http://www.imedicalapps.com/2011/04/title-enjoy-free-quick-access-resuscitation-guidelines-iresus-app/>

iDAS: Adult algorithms produced by the UK Difficult Airway Society; assume ready availability of a wide variety of airway adjuncts.

The following listings contain further ideas, all free to download:

<http://www.imedicalapps.com/2013/06/free-iphone-medical-apps-physicians/>

<http://www.imedicalapps.com/2011/01/top-free-android-medical-apps-healthcare-professionals/>

"Calculate by QxMD"

<https://itunes.apple.com/us/app/calculate-medical-calculator/id361811483?mt=8> is useful for paediatric drug dose calculations.

MediBabble

<http://www.medibabble.com/features.html>

This is a medical translator app which does not require an internet connection once downloaded. Language choice is currently limited but more languages are being added.

Bhansali and Armstrong recently reviewed paediatric anaesthesia apps. They found the following worth considering:

Peds ED

<https://itunes.apple.com/us/app/peds-ed/id326615524?mt=8> and

Paeds ED Lite

<https://itunes.apple.com/us/app/paeds-ed-lite/id331122046?mt=8>

are quick reference guides to resuscitation and simple PICU calculations, and both are currently [31/08/13] free to download.

AnaPaed

<https://itunes.apple.com/us/app/anapaed-peds-anesthesia/id393135847?mt=8>

contains a wealth of useful information on airway device sizing and regional anaesthesia techniques as well as routine and emergency drug dose calculations.

This app has relatively low cost at the time of writing.

The Open Anaesthesia Self-Study app

<https://itunes.apple.com/us/app/openanesthesia-self-study/id657712202?ls=1&mt=8>

is a question/model answer bank aimed at resident physicians and nurse anaesthetists preparing for US credentialing examinations. It covers the whole of anaesthesia, not just paediatric patients, and has a strong first-world emphasis. Basic concepts are well covered. Basic app free to download, updates/additional questions rather costly.

REFERENCES

Sheraton TE, Wilkes AR, Hall JE. Mobile phones and the developing world. *Anaesthesia* 2012; **67**(9) : 945-50.

Hudson J, Nguku SM, Sleiman J, et al. Usability testing of a prototype Phone Oximeter with healthcare providers in high- and low-medical resource environments. *Anaesthesia* 2012; **67**(9) : 957-67.

Haynes AB, Weiser TG, Berry WR, et al, for the Safe Surgery Saves Lives study group. A Surgical Safety Checklist to Reduce Morbidity and Mortality in a Global Population *N Engl J Med* 2009; **360** : 491-499. This article is free to download; <http://www.nejm.org/doi/full/10.1056/NEJMs0810119#t=articleTop>.

Bhansali R, Armstrong J. Smartphone applications for pediatric anesthesia. *Paed Anaesth* 2012;22: 400-4 – this article is free to download; <http://onlinelibrary.wiley.com/doi/10.1111/j.1460-9592.2012.03805.x/full>.

Update in Anaesthesia

Guide for Contributors

Update in Anaesthesia is primarily an educational journal, which aims to provide ongoing learning and support for anaesthetists working in situations with limited resources.

Update is sent to over 3000 English-speaking anaesthetists, and read by many others including surgeons, nurses and medical students. *Update* is also translated into different languages including Spanish, Russian, French and Mandarin. After being produced in the paper format, *Update* is published on the internet (www.worldanaesthesia.org) and read by 90 people a day from more than 130 countries. *Update* is also distributed in the form of a CD-ROM, produced by the Association of Anaesthetists of Great Britain and Ireland.

Articles for consideration by the Editorial Board should be submitted as Word documents (Rich Text Format is preferred) to the Editor-in-chief, Bruce McCormick, by email at Bruce.McCormick@rdefn.nhs.uk or post on CD-ROM or paper copy to Dr Bruce McCormick, Department of Anaesthesia, Royal Devon and Exeter Hospital, Barrack Road, Exeter, EX2 5DW, UK.

CLINICAL OVERVIEW ARTICLES

General considerations

- Papers must not have been published in whole or any part in another journal.
- Papers are subject to editorial revision.
- On acceptance for publication copyright becomes vested in the journal.
- Original textual matter quoted from other authors must have formal citation and be appropriately referenced.
- Some readers' first language may not be English. Please keep your text straightforward and avoid long sentences and complex terminology. Explain words and abbreviations that may not be universally standardised. Aim to include the full range of therapies available worldwide, but provide most detailed descriptions of those therapies available in resource-poor settings (see 'Management of sepsis with limited resources' in *Update 23* – www.worldanaesthesia.org/component/option,com_docmanta,cat_viewgid,67/Itemid,49/). Discuss older drugs as well as newer ones; halothane, thiopentone, ketamine and ether are widely used around the world.
- The article should be long enough to cover the topic in reasonable detail. Many readers will not have access to texts or journals to supplement their reading. Include text boxes and teaching points to make the layout interesting. Avoid long numbered lists with complex subdivisions. Check that your text is correct, particularly drug doses, as many readers will not be able to verify them.

Authors' details

Please supply the full forename and surname of all authors, stating their title (Anaesthetic Clinical Officer, Dr, Professor etc) and the name and address of their institution. One author should be identified for correspondence, with an email address provided.

Drug doses

Please use the international units, e.g. mg.kg⁻¹ rather than mg/kg. Use SI notation for g, mg, mcg etc. Please use internationally accepted non-proprietary drug names, e.g. furosemide, epinephrine and avoid trade names.

Headings

Three levels of heading may be used CAPITALS, **bold** and *italic*. Please do not employ different fonts within the text. Bullet points can be helpful.

Illustrations / figures

These may be sent to us as drawings (black on white), which we will scan into the text, or as picture files in jpg (JPEG) format. Black and white photos are also suitable. If you do not have facilities to produce drawings, contact the editor for help. If you copy illustrations from another publication please obtain copyright permission from the publishers or author. If patients appear in a photo please ensure that they have consented to this. Text accompanying illustrations should be supplied on a separate piece of paper.

Tables or figures reproduced from other published texts should be accompanied by a statement that permission for reproduction has been obtained from the author or publisher. An acknowledgment should be included in the caption and the full reference included in the reference list.

Tables

These should be prepared using the Microsoft Word table facility whenever possible.

Graphs

Graphs should be supplied using the Microsoft graph-compiling feature within Microsoft Word, or as a figure on paper.

References

A minority of Update readers have access to journals and therefore references should in general be limited to those that would be considered as 'further reading'. Please format your references as shown. Number the references in the order they appear, using the reference number as a superscript at the relevant point in the text.

References should include: names and initials of all authors (unless more than 6, when only the first 6 are given followed by 'et al.'), title of the paper; Medline abbreviation of the journal title (in *italic*); year of publication; volume number; first and last page numbers.

Papers accepted but not yet published should be included in the references, with the abbreviated journal name, followed by '(in press)'.

Those in preparation (including any submitted for publication), personal communications and unpublished observations should be referred to as such in the text.

1. Reynolds F, O'Sullivan G. Lumbar puncture and headache. 'Atraumatic needle' is a better term than 'blunt needle'. *Br Med J* 1998; **316**: 1018.
2. Costigan SN, Sprigge JS. Dural puncture: the patients' perspective. A patient survey of cases at a DGH maternity unit 1983–1993. *Acta Anaesthesiol Scand* 1996; **40**: 710–14.
3. Spriggs DA, Burn DJ, French J, Carlidge NE, Bates D. Is bedrest useful after diagnostic lumbar puncture? *Postgrad Med J* 1992; **68**: 581–3.

References to books should give book title, place of publication, publisher and year; those of multiple authorship should also include chapter title, first and last page numbers, and names and initials of editors. For example:

1. Roberts F. Chapter 22: Ear, nose and throat surgery. In: Allman KG, Wilson IH, eds. *Oxford handbook of Anaesthesia* (1st edition) Oxford: Oxford University Press, 2001: 506-39.

UPDATE SHORT REPORTS

The scope for publication of articles describing original research and audit conducted in, and specifically relevant to, poorly-resourced settings is limited. Successful publication in major journals is rare and the distribution and accessibility of the national and regional journals that currently publish these articles is often poor. As the official journal of the World Federation of Societies of Anaesthesiologists, *Update in Anaesthesia* is the appropriate forum for publication of these manuscripts and offers a wide distribution.

The guidance above for clinical overview articles applies, with the following additional considerations.

Legal considerations

- Papers based on clinical investigation on humans should include the consent of patients and a statement of approval from an appropriate Ethics Committee. In those institutions where Institutional Review Board consent is required for the performance of audits, this should be obtained and referred to in the text.
- Avoid use of identifiable names, initials and hospital numbers of patients.
- Human subjects of case reports, research or audits should not be identifiable. Manuscripts should not disclose patients' names, initials, hospital numbers (or other data that might identify the patient(s)).
- Guides for use of tables, figures and illustrations are as described above for Clinical Overview articles.

Brief Communications

- Original investigative articles or audits of patient outcome or clinical techniques.
- Up to 1500 words (approximately 2 pages of *Update in Anaesthesia*).
- Subdivided into:
 - Summary (maximum five sentences) and key words
 - Introduction
 - Patients and methods
 - Results
 - Discussion
 - Acknowledgements
 - References – maximum 15
 - Tables and/or figures - limited to two per article.

Case Reports

- Suitable for presenting descriptive studies (a series of cases), personal experience or individual case reports of particular interest.
- Up to 800 words. Three tables or figures is allowed in addition to text.
- A summary may be included (up to five sentences). Division into sections is optional.
- Up to seven references may be given.

Correspondence

- Welcomed on any subject, including editorials or articles that have appeared in *Update in Anaesthesia*.
- Letters may also be a suitable vehicle for presenting items of experience or observation that are too brief for Brief Communications.
- Papers describing procedures, techniques or equipment adapted by readers to their own conditions of work are welcomed.

Proofs

- Proofs are sent to the author designated to receive them. Corrections should be kept to a minimum and the proofs returned within 7 days of receipt.

The editorial team will be delighted to help with the preparation of articles. The best way of doing this is via email - Bruce.McCormick@rdefnhs.uk

Dr Bruce McCormick
Editor-in-chief
Update in Anaesthesia, July 2008

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