

## Anaesthesia for Neurosurgical procedures - Ventriculo-peritoneal (VP) shunt and Meningomyelocele repair

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### Abstract

The anaesthetist providing anaesthesia for paediatric neurosurgical procedures should be aware of safe techniques to provide the child with the best chance at good postoperative neurologic outcomes.

Ventriculo-peritoneal shunt (VP shunt) insertion and meningomyelocele repair are two common paediatric neurosurgical procedures. Anaesthesia for these surgeries should prevent further brain damage and preserve or improve the workings of the central nervous system.

**Key words:** ventriculo-peritoneal shunt; meningomyelocele repair; paediatric anaesthesia; hydrocephalus; myelomeningocele repair; neuroanaesthesia; anaesthesia for vp shunt; anaesthesia for meningomyelocele repair; anaesthesia for neurosurgery

### VENTRICULO-PERITONEAL SHUNT

Ventriculo-peritoneal shunt (also known as cerebral shunt) is the passage of a shunt from a ventricle in the brain to the peritoneal cavity. The ventricle may be one of the lateral ventricles, the third ventricle or fourth ventricle. Shunt location is usually based on its indication for insertion. The most common ventricle used is the right lateral ventricle because most persons are right-handed (dominant brain hemisphere on the left), thus the left side of the brain is usually spared by surgeons in case complications arise.

VP shunts are mostly inserted to drain cerebrospinal fluid (CSF) to relieve pressure on the brain in cases of hydrocephalus. Hydrocephalus is excessive accumulation of CSF in the brain caused by a problem in its formation, flow or absorption.

CSF is produced predominantly by ependymal cells in the choroid plexuses of the ventricles of the brain, while absorption takes place in the arachnoid granulations in the subarachnoid space. The normal CSF volume in children is 2 to 4 ml.kg<sup>-1</sup>.

The CSF circulates within the ventricles of the brain with the majority produced within the two lateral ventricles. From here, CSF passes through the interventricular foramina to the third ventricle, then the cerebral aqueduct to the fourth ventricle and then into the subarachnoid space. Any factor that leads to

excessive production of CSF, blockage of CSF flow or disruption of absorption can cause hydrocephalus.

In infants, because of open fontanelles, there may be no change in intracranial pressure (ICP) in the presence of hydrocephalus, however in severe cases (and in older children whose fontanelles are fused) the elasticity of the skull is exceeded. Hydrocephalus can then result in elevated ICP with resultant compromise of cerebral perfusion and risk of brain herniation if severe. Normal ICP is 0-6mmHg in neonates and infants, 3-7mmHg in toddlers and pre-schoolers and 5-15mmHg in older children.

The relationship between cerebral perfusion pressure (CPP) and intracranial pressure (ICP) is governed by the following equation:

$$\text{CPP} = \text{MAP} - (\text{ICP} + \text{JVP})$$

**MAP** = Mean Arterial Pressure

**JVP** = Jugular venous pressure (value is usually zero)

VP Shunts usually have three major parts:

- An inflow catheter which drains CSF from the ventricles. It passes from the brain through a small hole in the skull, and then runs under the skin.
- A valve mechanism which regulates differential pressure or flow through the shunt tubing;

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**Table 1:** Indications for VP Shunt/ Causes of Hydrocephalus

<b>Congenital</b>	<b>Acquired</b>
Aqueduct obstruction/ stenosis	Infection e.g. post meningitis
Meningomyelocele	Tumours leading to CSF blockage
Arnold Chiari malformation	Intraventricular haemorrhage
Dandy-Walker syndrome	Trauma
Benign intracranial cysts	Granulomatous conditions
Vein of Galen aneurysms	Subarachnoid haemorrhage
Congenital central nervous system infections	Cysts e.g. Arachnoid cysts
Craniofacial anomalies e.g. craniosynostosis	Surgery
Inherited (X - linked)	

- An outflow catheter that runs under the skin and directs CSF from the valve to the peritoneal cavity.

The distal end of the shunt is placed where there are epithelial cells to absorb CSF. The peritoneal cavity is commonly favoured for shunts as it is easy to access and has adequate space for absorption of CSF; it is also associated with the least morbidity. A subgaleal (under the scalp) shunt can be used in infants who are too small or premature for other types.

### Types of Hydrocephalus

Hydrocephalus may be:

- Communicating (non-obstructive) - when there is no obstruction to the flow of CSF but the problem is a defect in absorption of CSF, or over production. (E.g. Post-meningitis).
- Non-communicating (obstructive) - due to an anatomical blockage in CSF flow. (E.g. Aqueduct stenosis).

### Epidemiology

The mean global prevalence of isolated hydrocephalus in the paediatric population ( $\leq 18$  years) is 71.9/100,000. When spina bifida-associated hydrocephalus is included, the prevalence increases to 87.8/100,000. Africa has almost double the prevalence of North America because of untreated or poorly treated neonatal meningitis and ventriculitis.

### Clinical features of hydrocephalus

The clinical features depend on the age of the child.

0 - 2 years: enlarged head, bulging fontanelles, bulging scalp veins

**Table 2:** Types of cerebral shunts

<b>Type of Cerebral Shunt</b>	<b>Location of CSF Drain</b>
Ventriculo-peritoneal (VP) shunt	Peritoneal cavity
Ventriculo-atrial (VA) shunt	Right atrium
Ventriculo-pleural (VPL) shunt	Pleural cavity
Ventriculo-cisternal (VC) shunt	Cisterna magna
Ventriculo-subgaleal(SG) shunt	Subgaleal space
Lumbar-peritoneal (LP) shunt	Peritoneal cavity



**Figure 1:** Advanced hydrocephalus with huge head and bulging scalp veins. Picture, courtesy of Dr Yusuf AS, Consultant Neurosurgeon, National Hospital Abuja, Nigeria.

(especially on crying), irritability, lethargy, fever, vomiting, failure of sutures to close.

‘Sun-setting eyes’ develop as the case worsens- the eyes are displaced downward and the child is not able to look up; causing the eyes to resemble a sun setting on the horizon.

Impaired vision - caused by compression of the optic chiasma from a dilated 3rd ventricle, occurs in advanced cases.

Nystagmus and random eye movement - result from abducent nerve paresis from stretching of periventricular structures.

Increased deep tendon reflexes and muscle tone in lower extremities (in advanced cases).

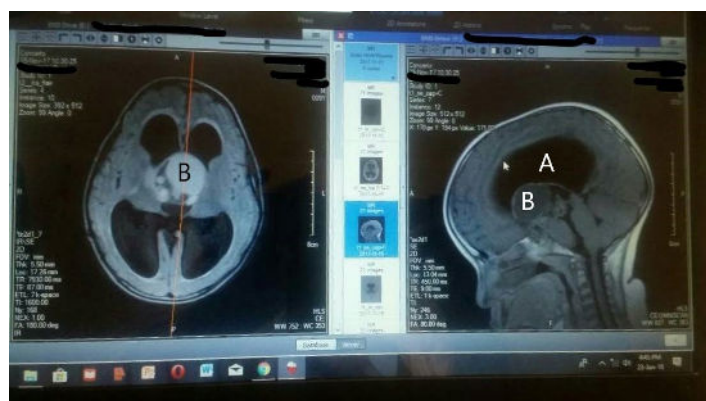
*Others are:* failure to thrive, delayed neurological development, limited control in the head and trunk, high pitched cry, seizures, coma.

*Older children:* When hydrocephalus occurs in a child after fontanelle closure, the symptoms are somewhat different. They present with normal head size (or mild enlargement), headache, vomiting, irritability, visual impairment, impaired eye movement, lower limb hyperreflexia, urinary incontinence, learning difficulties, seizures, lethargy and altered consciousness.

Untreated or mismanaged hydrocephalus, can lead to personality changes, intellectual disability and gait disturbances.

### Investigations for VP shunt insertion

- *Skull X-rays:* enlarged head, craniofacial disproportion, elongated inter-digitation of suture lines (raised ICP in older children).
- *Cranial/ Head/ Brain Ultrasound:* examines the size of the ventricles in very young babies. (Abdominal ultrasound in the pregnant mother can also diagnose hydrocephalus before birth).
- *Computed Tomography (CT) scan:* shows enlarged ventricles and periventricular oedema in hydrocephalus. Enlargement of the 4th ventricle suggests a communicating hydrocephalus. A relatively small 4th ventricle implies obstructive hydrocephalus.
- *Magnetic Resonance Imaging (MRI) of the brain:* shows size of the ventricle. Useful in detecting the cause of the hydrocephalus. E.g. aqueductal stenosis, tumour, Chiari malformations.
- *Transcranial Doppler:* Non-invasive measurement of the middle cerebral artery flow velocities and pulsatility index. This is used to analyse the cerebral circulation; increase in resistance of the cerebral arteries due to increased intracranial pressure is reflected in changes in blood flow velocity.
- *Lumbar puncture/ CSF tap:* for cell counts, protein concentration, and to exclude residual infection (e.g. post meningitic hydrocephalus). A protein concentration  $> 4\text{g.L}^{-1}$  will clog up most VP shunt valves.



**Figure 1:** MRI of a 5 year old male showing a huge mass (B) in the region of the Foramina of Monro causing severe obstructive hydrocephalus (A). Picture, courtesy of Dr Yusuf AS, Consultant Neurosurgeon, National Hospital Abuja, Nigeria.

Other investigations are

- *Full blood count (Complete blood count):* leucocytosis if there is infection. Anaemia may be present if the child has feeding problems.
- *Electrolytes, urea and creatinine:* There may be dyselectrolytemia due to vomiting caused by raised ICP.

Treatment of hydrocephalus is mostly surgical. Medical treatment is reserved for selected cases and is usually temporary. The three surgical options are:

- VP shunt
- Endoscopic Third Ventriculostomy (ETV) - A procedure especially suited for obstructive hydrocephalus. It involves the creation of an opening in the floor of the third ventricle to allow CSF drainage.
- Endoscopic Third Ventriculostomy with Choroid Plexus Cauterization (ETV/CPC)

VP shunt insertion may be done as an elective or emergency procedure depending on the presentation and associated pathology.

Contraindications to shunt placement:

- Infection over the entry site.
- CSF infection.
- Bloody CSF: potential for clots to block the shunt. Initial External Ventricular Drainage (EVD) can be performed to treat raised ICP until it is reasonable to shunt.
- High CSF protein (relative contraindication): can also potentially lead to shunt blockage.
- Coagulopathy (relative contraindication).

### Anaesthetic considerations for VP shunt placement

The following must be considered for successful outcome of anaesthesia:

- Coexisting congenital anomalies and syndromes need to be considered when planning anaesthesia.
- Large head may be wobbly and heavy. An assistant or head roll can help to stabilise the head.
- A large head may also prevent optimal positioning for laryngoscopy, thus ramping may be required. A difficult airway tray should be available.
- Risk of airway occlusion from over-flexion of the neck due to large head falling forward.
- Raised ICP: aim to prevent further rise in ICP at induction and intraoperatively.
- Positioning - supine, slight head up and with head tilt, usually left tilt.
- Restricted access to patient intraoperatively requires adequate intravenous access for fluid administration and intubation to secure the airway.

- The patient can be unpleasantly stimulated while the surgeon advances the trocar for tunnelling. Ensure good analgesia/ deep anaesthesia and muscle relaxation prior to tunnelling to avoid stimulation, pain and movement.
- Risk of postoperative nausea and vomiting (PONV) which requires use of antiemetic particularly in children older than 2 years.
- Intraoperative bleeding may be significant especially in small infants and they may require blood transfusion.

### Preoperative management

The goals of preoperative anaesthesia management are to establish a rapport with the patient/ parents, allay anxiety, do a proper review of the patient and laboratory investigations, optimise the patient's physical condition if possible and to plan a safe anaesthetic technique.

The child may have coexisting congenital anomalies like congenital heart disease or spinal bifida. Other children may be syndromic (e.g. Down's syndrome). These conditions may have implications for anaesthesia. The cause of the hydrocephalus must be considered when planning anaesthesia. History should include symptoms suggestive of raised ICP such as seizures, vomiting and neurologic deficits, a drug history may reveal the use of anticonvulsants. The child may have had previous surgeries including previous VP shunt insertions and be coming for a revision or reinsertion.

Physical examination with special emphasis on cardiac, respiratory, neurologic and airway examination is performed. The level of consciousness should be determined. The potential for difficult airway and intubation is assessed and adequate preparations made.

Radiological and laboratory investigations listed earlier are reviewed as well as other investigations related to comorbidities e.g. echocardiography for congenital heart disease. Anaemia, dyselectrolytaemia and infections should be treated prior to surgery (this may require blood transfusion if the child is severely anaemic) and the child optimised as best as possible. The parents (and child) must be properly counselled on the procedure (expectations should be managed ideally by the surgical team) and informed consent obtained. The weight of the child may be erroneously high because of the large head. Standard fasting guidelines are communicated to the care-givers. The child should not be fasted for too long as dehydration can complicate anaesthesia. Maintenance intravenous fluids should be commenced preoperatively to prevent hypovolaemia; younger infants may require dextrose-containing IV fluid to prevent hypoglycaemia.

The anaesthetic technique of choice for VP shunt insertion is general anaesthesia with muscle relaxation. The positioning of the patient's head for the surgery makes the airway inaccessible to the anaesthetist intraoperatively, furthermore tunnelling of the VP shunt from the brain to the peritoneum is stimulating and thus requires a still patient.

### Intraoperative management

Transportation to theatre should avoid airway occlusion and hypothermia. The head is prevented from falling forward as this can

compromise the airway, a chin lift can help solve this. The patient is kept warm and properly covered to prevent hypothermia. This is especially important for the large head because of its increased surface area.

Difficult airway equipment such as laryngeal mask airway, stylette, gum elastic bougie and a video laryngoscope (if available) should be available at induction of anaesthesia.

Basic monitoring (pulse oximetry, continuous electrocardiogram, non-invasive blood pressure, capnography and temperature) is usually sufficient for VP shunt insertion unless the child has a serious condition that requires more advanced monitoring.

Induction technique is based on the peculiarity of each case. Intravenous induction is applicable to patients without suspected difficult airway. Propofol or Thiopentone and rocuronium/ atracurium/ vecuronium are appropriate. Ketamine and suxamethonium should ideally be avoided as they cause a rise in ICP. However, in situations with a risk of aspiration at induction, suxamethonium use is appropriate as the rise in ICP is transient. In patients with raised ICP, pre-treatment of suxamethonium can be done with a non-depolarising muscle relaxant. In cases of suspicious airway, inhalational induction is preferred using Sevoflurane (or halothane if sevoflurane is unavailable). Mask ventilation may be a challenge because the head can fall forward and occlude the airway therefore proper stabilisation should be ensured using a head roll. Getting a proper mask seal can occasionally be problematic, thus different types and sizes of face masks should be provided. In cases of raised ICP, care should be taken with the use of volatile agents especially halothane, because they increase cerebral blood flow (CBF) which could lead to further rise in ICP. Isoflurane and Sevoflurane are associated with the least increase in CBF, keeping the minimum alveolar concentration (MAC) below 1 also lessens the risk of rise in CBF. Ramping may be required for optimal laryngoscopic view and easier tracheal intubation. Ramping involves placing supports (a ramp) beneath the head, neck and shoulders to bring the ears and sternum to the same horizontal plane/ level. Laryngoscopy and tracheal intubation may sometimes be easier in the lateral position.

Maintenance of anaesthesia is achieved using volatile agent, oxygen and air. Nitrous oxide is avoided as it increases CBF. Total intravenous Anaesthesia can be used in older children.

Normal Saline or Ringer's Lactate is employed for IV fluid maintenance; hypotonic solutions are best avoided to prevent further damage to the brain from cerebral oedema. Fentanyl, paracetamol and diclofenac suppository (in children >10kg) will provide adequate analgesia. Hypercapnia must be prevented as it causes cerebral vasodilatation and increased cerebral blood flow which causes an increase in ICP and resultant reduction in CPP. Prolonged hypocapnia on the other hand causes cerebral vasoconstriction which can lead to ischaemia. Normocapnia is preferred (ETCO<sub>2</sub> of 35-45mmHg). The theatre suite should be kept thermo-neutral and the child actively warmed. Adequate antibiotic coverage is necessary to prevent infection.

Positioning is usually supine with a head tilt and slight head up. The surgeon may infiltrate the scalp with local anaesthetic and adrenaline



mixture to reduce blood loss and provide analgesia. The dose of local anaesthetic and adrenaline used must be within safe limits for the child's age and weight. Prior to tunnelling of the shunt from the head down to the neck and abdomen, adequate analgesia and muscle relaxation is ensured. Antiemetic e.g. ondansetron 0.1mg.kg<sup>-1</sup> may be administered to prevent postoperative nausea and vomiting in older children.

At the end of surgery, the oropharynx is suctioned and residual neuromuscular block antagonised with neostigmine and glycopyrrolate/ atropine, or sugammadex. Extubation may be performed following confirmation of return of good muscle function and tidal volume. Extubation should be performed when the child is fully awake.

### Postoperative management

In the Post Anaesthesia Care Unit (PACU), the child's head is properly positioned with slight head-up tilt and chin lift to avoid airway occlusion which can lead to hypoxia and cardiac arrest.

Depending on the patient's clinical condition, some children may require direct admission to the ICU, while others may be observed in the PACU and sent to the ward.

Good shunt monitoring is very important for early detection of complications so that they are quickly resolved before the patient deteriorates.



**Figure 3:** A Right sided Ventriculo-Peritoneal shunt in place. Picture, courtesy Dr. M. R. Mahmud, Associate Professor, Ahmadu Bello University, Zaria, Nigeria.

### Outcome/ Prognosis

VP shunts can be life-saving; however, the outcome depends on the indication for the shunt. Patients with benign disorders have better outcomes than those with malignant tumours. In children, up to 15 - 50% of VP shunts fail within two years and repeat surgeries are often required.

### MENINGOMYELOCELE

Meningomyelocele (Myelomeningocele) is a type of spina bifida that is characterised by the protrusion of the meninges and spinal cord through a vertebral defect into a sac. It occurs due to failure of closure of the neural tube during embryonic development (fourth-week post-fertilization). There are three main types of spina bifida - spina bifida occulta, meningocele and meningomyelocele, with meningomyelocele being the most severe.

The commonest location for meningomyelocele is the lumbosacral region (lower back), though it may rarely occur in the thoracic region and neck. The incidence is 1 in 1000 livebirths, with some geographic variation. It is 3 to 7 times more common in females than males. There is higher prevalence in whites and Hispanics than in blacks. Low socioeconomic status also increases the risk.

Meningomyelocele is a condition with serious neurologic implications. It may occur with other congenital anomalies such as facial clefts, cardiac anomalies and urogenital anomalies. Associated problems include lower limb paresis/ paralysis, bladder or bowel incontinence, Arnold-Chiari malformation, hydrocephalus, a tethered spinal cord and latex allergy.

### Causes and Risk factors

The cause of meningomyelocele is still not very clear, but it is associated with nutritional, environmental and genetic factors. A maternal deficiency in folic acid has been demonstrated to play a role in the pathogenesis. Environmental factors such as maternal diabetes mellitus, obesity as well as exposure to folic acid antagonists and

**Table 3:** Complications of VP shunt insertion

Intra-operative	Post-operative
Bleeding from subcutaneous vessels during the tap	Infection - Shunt infection, ventriculitis, meningitis, peritonitis
Ventricular collapse from rapid aspiration of CSF	Shunt obstruction/ Kinks
Intracranial haemorrhage if CSF is drained too rapidly	Over-drainage, under drainage
CSF leak from puncture site	Shunt failure (from obstruction, infection, shunt component separation or migration, pseudocyst formation, bowel perforation, etc.)
Pneumothorax	Misplaced shunt
Infection from skin flora entering the shunt	Abdominal complications: peritoneal pseudocysts, lost distal catheters, bowel perforations and hernias.



**Figure 4:** A large lumbosacral meningocele. Picture, courtesy of Dr Osazuwa UA, Consultant Neurosurgeon, University of Benin Teaching Hospital, Benin City, Nigeria.



**Figure 5:** Ruptured meningocele. Picture, courtesy of Dr Timothy S, Senior Registrar, Neurosurgery Department, Ahmadu Bello University Teaching Hospital, Zaria, Nigeria.

anti-epileptics like carbamazepine, valproic acid and methotrexate have been implicated. 5, 10 Methylene tetrahydrofolate reductase (MTHFR) 677TT genotype is a risk factor for meningocele. There is also increased risk for subsequent babies to have meningocele after an index case in a family.

#### *Clinical features:*

The most common presentation of meningocele is a swelling at the back with or without associated features of hydrocephalus, noted at birth or on antenatal ultrasound. The child may also present with lower limb paresis/ paralysis. The presenting neurologic deficits depend on the level of the lesion. When meningocele is noticed at birth, the neuroplaque is at risk of trauma and subsequent CSF leakage and infection. The sac may be intact or have ruptured at presentation. It should be covered with a warm soaked gauze and the neonate transferred to a specialised centre.

#### **Investigations**

*Prenatal:* Screening for elevated maternal alpha-fetoprotein level during the first trimester is diagnostic in 85% of cases. Amniocentesis can be performed to test for alpha-fetoprotein level. Ultrasound is very effective in screening for neural tube defects.

*Postnatal:* CT scan, MRI.

#### **Treatment**

Surgical repair is the mainstay of treatment of meningocele which involves closure of the neural defect. It is a surgical emergency and primary closure is done within 24 – 48 hours of birth to reduce the risk of rupture of the neuroplaque and further neurological damage.



**Figure 6:** An MRI showing long segment syrinx with meningocele in a child, sac contains nerve roots and CSF. Picture, courtesy of Dr Ugwuanyi C, Consultant Neurosurgeon, National Hospital Abuja, Nigeria.

#### **Anaesthetic considerations for meningocele repair**

- Prone positioning is required for surgical access
- Protection of meningocele during induction of anaesthesia requires a doughnut roll or induction in the lateral position.
- Temperature control, avoid excessive heat loss.

- Risk of Latex sensitisation and allergy because of repeated surgical procedures and urethral catheterisations. Where possible, anaesthesia and surgery should be latex-free.

If there is associated hydrocephalus, the anaesthetic considerations will include those already discussed above.

### Pre-operative management

The preoperative review and investigations are generally as highlighted earlier for VP shunt.

If rupture of the neuroplaque has occurred, the child may be acutely ill with hypothermia and infection, thus will need resuscitation and optimisation before surgery.

### Intra-operative management

Anaesthetic technique of choice is general anaesthesia with tracheal intubation. For surgical access, the child will be placed prone on the operating table. Intravenous or inhalational induction is employed after preoxygenation. Anticholinergic prophylaxis with atropine or glycopyrrolate may be indicated in neonates. Tracheal intubation is done with the patient supine or in lateral position, the tracheal tube must be fixed securely to minimise dislodgement during positioning. In the supine position, the meningomyelocele should be placed in a 'doughnut' to avoid unnecessary pressure on it and accidental rupture. An armoured tracheal tube is ideal to prevent kinking in the prone position, if not available a regular tracheal tube can be used. However, care must be taken to prevent kinking of the tube while in the prone position. The eyes should be well covered by pads and tape to avoid injury in the prone position. A head ring will be required to place the head in a comfortable position and excessive rotation of the head is avoided. All pressure areas are padded and the abdomen ensured to be freely mobile for adequate ventilation and to prevent engorgement of epidural vessels from increased intra-abdominal pressure which increases blood loss. It is imperative to re-confirm correct tracheal tube placement after positioning as it can be easily dislodged. This is reliably done with capnography or auscultation. Fentanyl or remifentanyl and paracetamol are adequate for intra-operative analgesia. Volaeamic status is maintained to ensure adequate spinal cord perfusion pressure and minimise spinal cord ischaemia. Fluid replacement is with an isotonic solution and 3rd space loss may be high. Blood glucose monitoring is essential. Children with meningomyelocele have defective autonomic control below the lesion and easily become hypothermic. Active warming methods must therefore be deployed.

If the surgeon intends to do direct muscle stimulation to spare neurologic tissue, ultrashort acting muscle relaxants are used. A VP shunt may be inserted if the child has associated hydrocephalus. Blood loss is usually minimal except for large lesions requiring extensive flap cover. In addition, the surgical site may be infiltrated with adrenaline to reduce blood loss in these situations. Local anaesthetic can also be infiltrated at the end of surgery to provide immediate pain relief.

At the end of surgery, the patient is returned to the supine or lateral position for emergence and extubation. Adequate intravascular volume must be ensured before returning to the supine position to prevent hypotension.

### Post-operative management

Postoperatively, the child is nursed in the Post Anaesthesia Care Unit, the High Dependency Unit or the Intensive Care Unit depending on perioperative condition. Neonates less than 60 days post-conceptual age have a high risk of post-operative apnoea and should be appropriately monitored. Symptoms of raised ICP can occur post-operatively in patients with associated hydrocephalus if a shunt was not placed. Other immediate post-operative complications include brain stem herniation presenting as bradycardia, apnoea, cyanosis and respiratory arrest.

Meningomyelocele repair can also be done in utero, this will involve general anaesthesia for the mother. Foetal repair has been shown to decrease the need for shunt placement and is associated with better outcomes and less morbidity. However, foetal repairs are presently available only in quaternary health systems with extensive training and resources.

### Postoperative complications

- Apnoea
- CSF leakage
- Wound dehiscence
- Infection
- Hydrocephalus/ Raised ICP

### Outcome/ Prognosis

Physiotherapy will be required to optimise limb function. Survival rates have improved due to recent advances, however urinary tract infection can still be a problem due to repeated need for catheterisation.

### CONCLUSION

Understanding of the pathophysiology of these paediatric neurosurgical conditions help to plan safe anaesthesia in order to prevent worsening of morbidity and also to help improve surgical outcomes.

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