Clinical Review

Update in
Anaesthesia

Hypertrophic pyloric stenosis in infants

Maxwell Tobin* and Elizabeth T Drums
*Correspondence email: maxikoto@yahoo.co.uk
doi:10.1029/WFSA-D-20-00007

Abstract

Hypertrophic pyloric stenosis is a common condition, occurring in 1 out of 500 live births. Boys are affected more than girls, with a ratio of 4:1. Presentation is usually between 2 – 6 weeks of life with classical non-bilious projectile vomiting. Examination will reveal a dehydrated child with a palpable olive-shaped mass located between the midline and right upper quadrant. The lesion is commonly delineated by ultrasonograpgy. The exact cause is largely unknown, however several theories have been advanced, which include genetic and environmental factors.

It is not a surgical emergency, so the initial management is aimed at resuscitation that will correct dehydration, electrolyte and acid-base disturbances before proceeding to surgery. Rapid sequence intravenous induction, endotracheal intubation and muscle relaxation is the anaesthesia technique of choice. Post operative apnea is a possibility, and such monitoring should continue well into the postoperative period.

Key words: hypertrophic; pylorus; stenosis; infants; anaesthesia

INTRODUCTION

Hypertrophic pyloric stenosis in infants is the narrowing of the gastric outlet caused by hypertrophy of the muscularis layer of the pylorus. The aetiology is unknown but there is a genetic predisposition. The incidence is approximately 1 in 500 live births, boys are affected more than girls, with a ratio of 4:1. A total of 40–60 percent of cases occur in the first-born children. It is the commonest cause of intestinal obstruction in infancy requiring surgery.

Gastric carcinoma and chronic peptic ulceration can also give rise to acquired pyloric stenosis in adults. However, this article will focus on congenital pyloric stenosis that occurs in neonates or early infancy.

Aetiology

The exact cause is largely unknown, however several theories have been advanced, which include genetic and environmental factors.

Risk Factors

Risk factors for pyloric stenosis include:

- ***Sex***. Pyloric stenosis is seen more often in boys — especially firstborn children — than in girls.
- ***Race***. Pyloric stenosis is more common in whites of northern European ancestry, less common in African-Americans and rare in Asians.
- ***Premature birth***. Pyloric stenosis is more common in babies born prematurely than in full-term babies.
- ***Family history.*** Studies found higher rates of this disorder among certain families. Pyloric stenosis develops in about 20 percent of male descendants and 10 percent of female descendants of mothers who had the condition.
- ***Smoking during pregnancy***. This behavior can nearly double the risk of pyloric stenosis.
- ***Early antibiotic use***. Babies given certain antibiotics in the first weeks of life - erythromycin to treat whooping cough, for example - have an increased risk of pyloric stenosis. In addition, babies born to mothers who took certain antibiotics in late pregnancy may have an increased risk of pyloric stenosis.
- ***Bottle-feeding***. Some studies suggest that bottle feeding rather than breast-feeding can increase the risk of pyloric stenosis. Most of the people who participated in these studies used formula rather than breast milk, so it isn't clear whether the increased risk is related to formula or the mechanism of bottle-feeding.

Dr Maxwell Tobin *MBBS, DA (WASC), FWACS, PEAD ANAESTH CERT*
Association of Professional Women Engineers of Nigeria Port Harcourt Branch Port Harcourt Rivers NIGERIA

Dr Elizabeth T. Drum *MD, FAAP, FCPP, FASA*
Associate Professor of Clinical Anesthesiology and Critical Care Perelman School of Medicine University of Pennsylvania USA

Clinical Presentation

Pyloric stenosis usually manifests within weeks 2 to 6 of life with non-bilious vomiting that is projectile. The infant normally feeds well but vomits after each feed. There may be weight loss.

Examination will often reveal a dehydrated child with a palpable olive-shaped mass, typically 1-2cm in diameter located between the midline and right upper quadrant. The lesion is commonly delineated by ultrasonography or rarely with barium swallow and radiographic examination. Sometimes, this typical olive-shaped mass may not be palpated on examination or easily delineated by ultrasonography, diagnosis in this instance, will be based on high index of suspicion.

Arterial blood gas will show a marked metabolic alkalosis with hypokalaemia and hypochloraemia, especially when the patient presents late. In resource intense areas, this finding is no longer as common, as early suspicion allows diagnosis prior to extensive derangement of electrolytes. Urinalysis will reveal acidic urine with high level of potassium. Hypoglycaemia, mild uraemia, unconjugated hyperbilirubinaemia and haemoconcentration may also be seen.

Pathophysiology of the ABG picture

The blood gas results of hypokalemic hypochloraemic metabolic alkalosis seen in this condition are due to a combination of gastrointestinal, renal and respiratory changes.

Gastrointestinal: The vomitus in pyloric stenosis is mainly gastric fluid rich in hydrochloric acid. It is devoid of bicarbonate mixing due to the pyloric obstruction preventing bicarbonate from the small intestine to mix with the gastric fluid, as seen in normal vomitus. As a result of this only hydrogen and chloride ions are lost.

Renal: Due to these biochemical changes, the kidney is presented with a large bicarbonate load. This exceeds the absorptive threshold of the kidneys leading to alkaline urine that is seen initially. However, prolonged vomiting leads to hypovolaemia and dehydration. This in turn causes the activation of renin-angiotensin-aldosterone axis in an attempt to restore circulating volume. The aldosterone acts on the kidney to retain sodium at the expense of potassium and hydrogen ions. This leads to production of paradoxical aciduria and worsening of hypokalaemia and metabolic alkalosis.

Respiratory: The infant may try to compensate for the metabolic alkalosis by using the respiratory system. They may hypoventilate to produce hypercapnia, but this is never sufficient to correct the alkalosis, as the hypoxic drive will be triggered.

Other biochemical and haematological findings will include; hypoglycaemia, haemoconcentration, mild uraemia and unconjugated hyperbilirubinaemia .

Preanaesthetic consideration

The preoperative considerations can be divided into considerations that are specific to the pyloric stenosis and general consideration of paediatric anaesthesia.

Issues relating specifically to pyloric stenosis include: markedly deranged acid-base status, the effect of dehydration and the increased risk of regurgitation owing to the obstruction. Pyloric stenosis is not a surgical emergency, as such the infant should be fully resuscitated, and electrolyte abnormalities fully corrected before surgical intervention.

The challenges of anaesthetising a small infant include the altered anatomy and physiology, the presence of anxious parents, difficulty obtaining intravenous access and altered drug dosages.

Resuscitation

Resuscitation should take place under the watchful eyes of nursing staff that are well trained to take care of such cases, preferably in special care baby unit. Start by assessing the degree of dehydration (Table 1) and securing intravenous access. At same time take blood sample for full blood count, liver function test, electrolyte, urea and creatinine, as well as blood gases. These will serve as baseline to guide resuscitation and help determine the required potassium supplementation. Insert a nasogastric tube to remove gastric residue and perform a four hourly gastric washout to reduce the risk of aspiration. Regular monitoring which should include: urine output, blood pressure, heart rate, respiratory rate and oxygen saturation, as well as repeat blood gases will help ascertain the success of resuscitation.

Fluid management

Fluid management can be divided into resuscitation fluid, maintenance fluid and ongoing losses.

Resuscitation fluid: 0.9% Normal saline or Hartmann's solution should be used as resuscitation fluid, and it is calculated by assessing the degree of dehydration. The volume of fluid required can be calculated by multiplying the percentage dehydration by the weight of the infant multiplied by ten (% dehydration x body weight(kg) x 10). Half of the fluid deficit should be given within the first 24 hours and the other half given over the second 24 hours.

Table 1: Assessment of degree of dehydration

History	Examination	Degree	Clinical presentation
Frequency of vomiting, how much the infant takes orally, frequency of wet diapers, any associated diarrhoea and fever	Look for dry mucous membrane, a sunken fontanelle and eyes, tachycardia, hypotension, prolonged capillary refill and decreased level of consciousness	5%	Dry skin and mucous membrane
		10%	Cool peripheries, depressed fontanelle and oliguria
		15%	Hypotension and changes in level of consciousness

Alternatively, the resuscitation fluid can be given based on the initial values of serum electrolytes, especially serum chloride (Cl-) as follows:

- Cl- value < 85mmol/l give three boluses of 20ml/kg separated one hour apart
- Cl- value ≤ 97mmol/l give two boluses of 20ml/kg separated one hour apart

Maintenance Fluid: in addition to the resuscitation fluid, maintenance fluids are given in the form of Hartmann's solution or 0.45% saline with 5% dextrose, using the 4-2-1 rule (as described in table 2) to determine the hourly maintenance fluid.

Ongoing losses: Nasogastric drain or aspirate should be estimated and replaced using Hartman's solution.

Potassium replacement

Provided renal function test is within normal limit, 3mmol/kg/24 hour of potassium should be added to the maintenance fluids, as guided by regular blood gas analysis.

Intraoperative consideration

The anaesthetist should aim at achieving a urinary output of 1–2ml/kg/hr, if patient is catheterized, or the infant has had two wet diapers over 4–8 hours; as well as the following biochemical parameters;

- serum chloride concentration of above 97mmol/l
- bicarbonate of below 30mmol/l
- PH less than 7.5 with a base excess less than 6mmol/l and
- potassium concentration that is within normal limit, before proceeding with the case.

The procedure can be done laparoscopically or open and entails making an incision into muscularis layer of the pylorus making sure not to breech the mucosa. It lasts for between 30 minutes to 1 hour depending on speed and expertise of the surgeon. Prior to induction, the nasogastric tube should be aspirated, attempting to suction the four quadrants of the stomach by turning the infant accordingly. Standard monitors should be attached as per AAGBI guideline.

Rapid sequence induction with or without cricoid pressure is the induction technique of choice, using ketamine, sodium thiopentone or propofol depending on haemodynamic parameters. However, inhalational induction using sevoflurane or halothane, after an ultrasound confirmation of an empty stomach, has also been reported. The main reason that was advanced by the proponents of inhalational induction is avoidance of hypoxia in the infant that already has reduced oxygen reserve due to reduced functional residual capacity.

Tracheal intubation is facilitated with 1–2mg.kg^{-1} of suxamethonium. The airway should be secured using an appropriate sized endotracheal tube, throat packed, tube secured with adhesive tape and the infant connected to the machine via an Ayre's T-piece or circle system using pressure controlled ventilation mode. An intermediate or long-acting non-depolarizing neuromuscular blocker is given when the effect of suxamethonium has worn off. Fentanyl, paracetamol and local anaesthetic skin infiltration can be used for analgesia. The use of opioids is controversial, especially, if there is significant electrolyte disturbances on presentation. Even when these derangements are corrected, there is still a higher incidence of respiratory depression. Anaesthesia should be maintained with volatile agent/N_2O/O_2 combination (avoid nitrous if surgery is to be done laparoscopically).

Isotonic fluid (0.9% saline or Ringers lactate/Hartmann's) at 10ml/kg should be used intraoperatively to maintain circulating volume. However, if the infant is on glucose containing fluid preoperatively, that should continue in the intraoperative period with regular blood glucose monitoring to ensure normoglycaemia.

At the end of surgery, residual muscle paralysis is reversed and the infant extubated awake in a left lateral position.

Postoperative consideration

During the postoperative period, vital signs monitoring must continue, including SpO_2 and apnoea monitoring due to the possibility of postoperative apnoea up to 60 weeks post-gestational age, as well as increased incidence of apnoea in the face of electrolyte derangement, even after it has been corrected. Supplemental oxygen is required and intravenous fluid continued until feeding is established, in a graded manner, usually within 6 hours post operatively, unless the bowel mucosa was breached. Analgesia can be achieved with paracetamol, given intravenously at a dose of 7.5mg.kg^{-1} every 4–6 hours and converted to oral when oral intake is established. Alternatively, suppository paracetamol can also be used at a dose of 30mg.kg^{-1} as a loading dose followed by 20mg.kg^{-1} every 8 hours to a maximum of 60mg in 24 hours. However, the smallest suppository is available in 60mg, 125mg and 250mg formulations, meaning that the infant should be at least 2kg to receive the smallest rectal dose.

Table 2: Fluid replacement

	Type of fluid	**Volume of fluid**
Resuscitation fluid	Hartmann's solution or N/saline	% dehydration x Kg body weight x 10
Maintenance fluid	Hartmann's solution or 5% Dextrose in 0.45% saline	4- 2-1 rule 4ml/kg for 1st 10kg 2ml/kg for 2nd 10kg 1ml/kg for each following 10kg
Ongoing losses	Hartmann's solution	As estimated

The practice of cutting the suppository should be avoided because the active drug may not be evenly distributed throughout the wax.

Conclusion

Hypertrophic pyloric stenosis is a common condition, occurring in 1 out of 500 live births. Presentation is usually between 2 and 6 weeks of life with classical non-bilious projectile vomiting. It is not a surgical emergency, so the initial management is aimed at resuscitation that will correct dehydration, electrolyte and acid-base disturbances before proceeding to surgery. Rapid sequence intravenous induction, endotracheal intubation and muscle relaxation is the anaesthesia technique of choice. Post operatively, the infants generally do well. However, postoperative apnea is a possibility, and as such monitoring should continue well into the postoperative period.

REFERENCES

1. Bachiller PR, Chou JH, Romanelli TM, Roberts Jr JD. Neonatal Emergencies. In: Cotè CJ, Lerman J, Anderson BJ, editors. A Practice of Anesthesia for Infants and Children; 5th edition. Philadelphia: Elsevier Saunder, 2013; 746 – 765
2. Bell SF, Jane CSG. Pyloric Stenosis. In: Leslie RA, Johnson EK, Thomas G, Goodwin APL, editors. Dr Podcast Scripts for the Final FRCA. New York: Cambridge University Press. 2011; 337 – 341.
3. Miozzari HH, Tönz M, von Vigier RO, Bianchetti MG. Fluid resuscitation in infantile hypertrophic pyloric stenosis. *Acta Paediatr*. 2001; **90(5)**:511-4.
4. Sathya C, Wayne C, Gostsch A, Vincent J et al. Laparoscopic versus Open Pyloromyotomy in Infants: a systematic review and meta-analysis. *Pediatr Surg Int*. 2017; **33(3)**: 325 – 333.
5. Gagey AC, de Queiroz Siqueira M, Desgranges FP, Combet S etal. Ultrasound Assessment of the Gastric Contents for the Guidance of the Anaesthetic Strategy in infants with Hypertrophic Pyloric Stenosis: a prospective cohort study. *Br J Anaesth* 2016 May; **116(5)**: 649 – 54.
6. Mahon BE, Roseman MB, Kleiman MB. Maternal and infant use of erythromycin and other macrolide antibiotics as risk factors for infantile hypertrophic pyloric stenosis. *J Pediatr* 2001;**139**:380-4
7. Krogh C, Biggar RJ, Fischer TK, Lindholm M, Wohlfahrt J and Melbye M. Bottle feeding and the Risk of Pyloric Stenosis. *Pediatrics* October 2012, **130 (4)** 943-949.
8. Dalton BG, Gonzalez KW, Boda SR, Thomas PG, Sherman AK, St Peter SD. Optimizing fluid resuscitation in hypertrophic pyloric stenosis. *J Pediatr Surg*. 2016 Aug; **51(8)**:1279-82