INFANTILE HYPERTROPHIC PYLORIC STENOSIS ANAESTHESIA TUTORIAL OF THE WEEK 276

26TH NOVEMBER 2012

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QUESTIONS

Before reading this tutorial, please answer the following question (true or false). The answers and an explanation are at the end of the tutorial.

1. The following fluids have a role in infantile hypertrophic pyloric stenosis (true or false)
    a. 0.9% saline
    b. Hartmann's solution
    c. Isotonic bicarbonate
    d. 0.45% saline with 5% glucose

2. Which one of the following statements is false?
    a. The usual biochemical picture is that of hypochloraemic metabolic alkalosis
    b. Potassium chloride may be added to intravenous fluid once urine output is established
    c. Upper GI imaging and barium studies are the gold-standard for diagnosis
    d. Up to 80% of infants continue to regurgitate following corrective surgery

3. Regarding pyloromyotomy, which of the following statements is true or false?
    a. Pyloromyotomy is always an emergency procedure
    b. Pyloromyotomy is associated with severe postoperative pain
    c. Babies should be nil by mouth for a minimum of 12 hours following surgery
    d. Pyloromyotomy is curative and is associated with low mortality

INTRODUCTION

Pyloric stenosis, or infantile hypertrophic pyloric stenosis (IHPS), is a condition characterised by hypertrophy of the two muscle layers of the pylorus. The pyloric canal lengthens, the whole pylorus thickens, and the mucosa becomes oedematous causing functional obstruction of the gastric outlet.

EPIDEMIOLOGY

Incidence varies between regions, but ranges from 2-4 per 1000 live births. The condition is more commonly seen in males (4:1). Presentation is usually early in life, between 3 and 5 weeks of age, and approximately 95% of cases are identified in those aged 3-12 weeks.

Mortality is low. An eight-year cohort study of 350 patients admitted to hospital in the Republic of Ireland with IHPS identified only one death, which was attributed to an underlying myopathy.

AETIOLOGY

Whilst the exact causative mechanisms of IHPS are unknown, there are several established associations.

A Danish study identified a nearly 200-fold increase in incidence of IHPS among monozygotic twins, suggesting familial factors are significant. Several genetic loci have been implicated, including IHPS1, where a deficient production of neuronal nitric oxide synthase causes impaired relaxation of pyloric smooth muscle.

It has been suggested that a high gastric parietal cell mass may result in impaired gastrin control and hence, duodenal hyperacidity. This hyperacidity may produce IHPS as a result of repeated contraction of the pylorus.

The use of macrolide antibiotics such as erythromycin in the first two weeks of life appears to convey a risk of IHPS, and an association has also been shown between macrolide antibiotic use by pregnant or breast-feeding mothers and the development of IHPS in their offspring.

PRESENTATION

The classical presentation is that of a 3-5 week old infant with non-bilious projectile vomiting immediately after feeding, and where the infant is hungry soon after vomiting. An olive-like mass can be felt at the lateral edge of the rectus abdominis muscle in the right upper quadrant of the abdomen. The best time to feel this is immediately after a test feed, after the child has vomited.

The child may present with clinical signs of dehydration if the diagnosis if delayed and vomiting prolonged for a number of weeks. Signs of dehydration include a sunken fontanelle, dry mucous membranes, poor skin turgor and lethargy. Signs of malnutrition and poor weight gain may also be apparent.

Laboratory Investigations

The classical picture is hypochloraemic metabolic alkalosis due to loss of hydrochloric acid in vomited gastric fluid. This may be associated with hypokalaemia as the kidneys compensate for the underlying alkalosis by preferentially excreting potassium in order to retain hydrogen ions. This is usually seen as a result of persistent vomiting (often more than three weeks). Persistent vomiting and dehydration may also result in either hyper- or hyponatraemia. Pre-renal failure can occur.

DIAGNOSIS

Diagnosis is usually made on the basis of a typical history and palpation of the olive-like mass. In the early stages, IHPS can be difficult to distinguish from gastro-oesophageal reflux disease or even sepsis. In such situations, diagnosis may be confirmed radiologically.

Ultrasound

This is the diagnostic test of choice, although the accuracy is operator-dependent. The size of the pylorus is measured and compared to standard values. Pyloric muscle thickness >4mm, pyloric muscle length >14 – 20mm and pyloric diameter >10-14mm is diagnostic in term infants, with both a high sensitivity (90-99%) and high specificity (97-100%). Ultrasound is less accurate in premature infants.

Upper GI Imaging

Barium studies show classical signs of an elongated pyloric canal ('string sign') or thickened pyloric mucosa ('double-track sign'). Due to concerns about radiation exposure, upper GI imaging is only used when ultrasound is inconclusive.

MANAGEMENT

Resuscitation

Initial management should follow an 'ABC' approach, and fluid management should focus on correction of underlying dehydration, as well as electrolyte and acid-base abnormalities. An initial bolus of 20ml/kg 0.9% saline should be used if the infant is dehydrated. Thereafter, maintenance fluid should be started, the purpose of which is to maintain adequate hydration whilst protecting against hypernatraemia and hypoglycaemia. An appropriate maintenance fluid is 5% glucose/0.45% saline, provided the plasma sodium is not low; 5% glucose/0.9% saline should be used in this situation. Potassium chloride should be added to the fluid as required once urine output has been established. An accurate fluid balance chart should be kept and urinary catheterisation should be considered if the child is dehydrated. Serial electrolyte, acid-base and blood glucose measurements must be performed. A nasogastric tube should be passed to decompress the stomach.

Conservative Management

IHPS can be managed conservatively by naso-duodenal feeding. This has to be continued over several months to allow the obstructive process resolve and the infant to gain weight. Treatment with atropine has also been described. The safety and effectiveness of surgery means that conservative management is reserved for those in whom surgery is contra-indicated.

Surgery

This is not an emergency procedure, and surgery may proceed only when fluid balance, acid-base status and electrolyte levels have been restored to normal. The baby is at risk of postoperative apnoeas if the metabolic alkalosis is not corrected prior to surgery. The classical operation is a Ramstedt pyloromyotomy; a longitudinal incision of the pylorus with blunt dissection to the level of the sub-mucosa, thus relieving the gastric outlet obstruction. This operation is considered curative with minimal associated mortality.

Pyloromyotomy may also be performed laparoscopically. Prospective randomised controlled trials comparing open and laparoscopic techniques have suggested that laparoscopic surgery results in reduced post-operative emesis, reduced analgesia requirements, a more rapid return to enteral feeding and shorter hospital stay.

Endoscopic balloon dilatation has been used, but is not as successful as surgical pyloromyotomy. This technique is reserved for those in whom surgery poses a significant risk.

ANAESTHESIA

The nasogastric tube should be aspirated before induction of anaesthesia to minimise the risk of pulmonary aspiration of gastric contents. A "4 quadrant aspiration" (turning the infant through a full rotation and aspirating the stomach at each quarter turn) has been suggested as being effective.

Anaesthesia may be induced by inhalation with sevoflurane (or halothane), or rapid sequence induction with either thiopentone or propofol. This depends on personal preference and whether the stomach can be safely considered empty following suctioning, or if the child should still be considered at a high risk of aspirating on induction. Good intubating conditions are obtained with 1-2mg/kg of suxamethonium,

and prolonged paralysis using non-depolarising neuromuscular blocking drugs. Classic RSI with application of cricoid pressure is not possible in infants as the child will desaturate rapidly whilst apnoeic; a modified RSI with gentle positive pressure ventilation is commonly used.

Anaesthesia should be maintained with volatile agents with or without nitrous oxide (avoid nitrous in laparoscopic surgery). Full intra-operative monitoring as per AAGBI guidelines is strongly advised. Patient temperature should be monitored and the child should be maintained in a warm environment to avoid cooling. Antibiotic prophylaxis (e.g. co-amoxiclav 30 mg/kg IV or cefuroxime 50mg/kg IV) should be given prior to skin incision to reduce postoperative wound infection.

Boluses of an isotonic fluid (saline 0.9% or hartmann's solution) 10ml/kg may be given intra-operatively to correct circulating volume if required. Glucose containing maintenance solutions may be continued in theatre, but must NOT be used for bolus fluid replacement. If glucose containing maintenance fluids are discontinued intra-operatively, the blood glucose should be checked regularly to ensure normoglycaemia throughout the perioperative period.

It used to be common practice to inject air via the nasogastric tube at the end of the procedure to ensure pyloric patency, but this is no longer recommended.

Postoperative pain is not usually severe, and good analgesia may be achieved intra-operatively with an intravenous opioid such as fentanyl 1 mcg/kg and paracetamol (7.5 mg/kg IV or 30-40mg/kg PR). The wound should be infiltrated with bupivacaine 0.25% 2 mg/kg (0.8 ml/kg) where possible.

At the end of surgery, extubation should be performed in the left lateral position once neuromuscular blockade is fully reversed, and the infant is awake.

POSTOPERATIVE CONSIDERATIONS

Analgesia should be provided with paracetamol as required and non-steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen 5mg/kg if pain is worse than anticipated. The nasogastric tube may be removed at the conclusion of the procedure, and maintenance intravenous fluids should continue until feeding is re-established. Modern strategies focus on early implementation of feeding from four hours after surgery. Up to 80% patients continue to regurgitate after surgery, but this almost always improves in the ensuing days.

The infant can be considered fit for discharge when well hydrated and tolerating feeds. Prognosis following surgery is good and complete recovery and catch-up growth is the norm.

SUMMARY

- Infantile hypertrophic pyloric stenosis is a common condition, occurring in 2 – 4 per 1000 live births
- Presentation is usually early, between 3 – 5 weeks of age
- Initial management is aimed at resuscitation, correcting of dehydration, alkalosis and electrolyte disturbances before corrective surgery can occur
- A variety of anaesthetic techniques have been used successfully, although rapid sequence intravenous induction, endotracheal intubation and maintenance with an inhalational agent is common and safe.

ANSWERS TO QUESTIONS

1.
    a. T – the resuscitation fluid of choice
    b. T – may be used if monitoring blood glucose regularly
    c. F – may worsen alkalosis
    d. T – maintenance fluid of choice, provided the child is not hyponatraemic
2.
    a. T
    b. T
    c. F – on the occasions the disorder cannot be diagnosed clinically, ultrasound is the choice
    d. T
3.
    a. F – should be delayed until fluid balance and acid-base status are corrected
    b. F – although adequate analgesia should still be ensured
    c. F – early feeding, from 4 hours post-op, has been shown to be of benefit
    d. T

REFERENCES AND FURTHER READING

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