Infantile hypertrophic pyloric stenosis

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Abstract

Infantile hypertrophic pyloric stenosis (IHPS) is the most common cause of gastric outlet obstruction in infants and is one of the most common conditions requiring surgery in the newborn. It arises from marked hypertrophy of the pyloric muscle (primarily circular layer), resulting in partial or complete luminal occlusion. Classically these infants have persistent non-bilious vomiting, are dehydrated with weight loss, and have a hypokalaemic, hypochloraemic metabolic alkalosis. Treatment requires initial careful fluid management before definitive surgical correction. Outcomes are now excellent in developed countries and long-term sequelae are not expected after treatment.

Keywords Newborn; non-bilious vomiting; pyloric stenosis; pyloromyotomy; surgical management

Introduction

Infantile hypertrophic pyloric stenosis (IHPS) was first described in 1717 in a report to the Royal Society. It arises from marked hypertrophy of the pyloric muscle (primarily circular layer), resulting in partial or complete luminal occlusion, and occurs in up to 4 in 1000 live births in the UK and has a male-to-female ratio of 4:1. Most babies present between the age of 2 and 8 weeks, with a peak age at presentation of 6 weeks.

Aetiology

The cause of IHPS remains uncertain. Theories include:

- exposure to erythromycin
- pesticide exposure
- deficiency in nitric oxide pathways
- the presence of milk curds (causing pyloric mucosal oedema)
- gastric hyperacidity
- abnormal pyloric innervation
- diminished pacemaker cells (causing abnormal motility and subsequent pyloric hypertrophy)
- infection by *Helicobacter pylori*.

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Clinical presentation

History

The typical infant with IHPS presents with dehydration and non-bilious, projectile vomiting. This distinction about bilious vomiting in the newborn is critical, because if present this is suggestive of a more serious pathology requiring a time-critical intervention (e.g. malrotation with volvulus). There may be a recent history of fewer wet nappies and weight-loss noticed prior to attendance and the baby may be lethargic and less interactive. A strong family history of IHPS may be elicited, particularly if the mother had IHPS.

Examination

On inspection, the infant may already display signs of dehydration, including sunken eyes, mottled skin and lethargy. Visible peristalsis is sometimes seen migrating from the left upper quadrant inferomedially to the right-hand side. Some parents may have noticed and been alarmed by this sign.

On general examination, a sunken anterior fontanelle, prolonged capillary refill time and cool peripheries suggest significant dehydration.

Careful abdominal examination confirms the diagnosis in most cases. This is performed after a 'test feed' of milk, dextrose or air has been given orally or through the nasogastric tube. The aim of the test feed is to calm and relax the hungry baby to aid examination and to increase gastric peristalsis. Care is taken not to over-distend the stomach, because this can mask palpation of the enlarged pylorus. With the infant in the supine position, the doctor examines the right upper quadrant of the abdomen from the left-hand side. Elevation of the infant's legs can help relax the musculature of the abdominal wall. The hypertrophic pylorus is felt as an olive-shaped mass. The palpably enlarged pylorus is traditionally referred to as the pyloric 'tumour', but it is unwise to use this misleading and alarming term in the presence of the parents. The abdominal examination may need to be repeated several times before the hypertrophied pylorus is palpated.

Investigations

Radiology

The diagnosis of IHPS can usually be made clinically. However, increasingly abdominal ultrasound is used to confirm the diagnosis or has been undertaken prior to referral. Ultrasound is better indicated when the test feed and abdominal examination has not confirmed the diagnosis, but where a strong clinical suspicion still exists. This is ideally performed by a specialist paediatric radiologist to ensure a high specificity and sensitivity from the investigation. Using a high-resolution linear probe, the length of the pyloric channel and pyloric muscle thickness are measured (Figure 1). Channel length >16 mm and pylorus muscle thickness >3 mm are considered diagnostic in this age group. The scan is dynamic and the presence or absence of flow through the pyloric channel is also looked for. If clinical examination and abdominal ultrasound are still inconclusive, an upper gastrointestinal tract contrast study may be indicated.

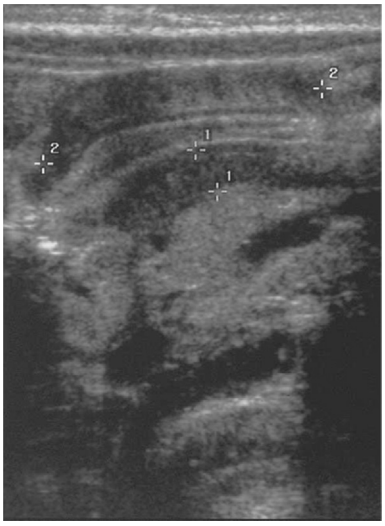

Figure 1 Abdominal ultrasound scan. The thickness of the pyloric muscle (denoted by the distance between the numbers marked '1' on the image) and the length of the pyloric wall (the distance between the numbers marked '2') are measured using a high-resolution linear probe.

Biochemistry

These infants typically develop a hypochloraemic, hypokalaemic, hyponatraemic, metabolic alkalosis due to hydrogen and chloride being lost through persistent vomiting. The kidney attempts to maintain pH by excreting potassium in exchange for hydrogen ion preservation, a process also impaired by the loss of chloride in the vomitus preventing bicarbonate excretion. As the degree of dehydration worsens, aldosterone is stimulated which allows sodium to be retained at the expense of hydrogen ions – leading to a paradoxical acidification of urine. Prompt assessment of serum electrolytes and pH are therefore essential.

In recent years, the diagnosis of IHPS is being made earlier in the disease course (often through an ultrasound performed by paediatricians soon after onset of vomiting) and so the classical electrolyte derangement and significant dehydration of IHPS often has not become established by the time these babies present to the surgical team.

Management

Correction of fluid and electrolytes

Administering a general anaesthetic to an infant before correction of metabolic alkalosis poses significant risks to the infant and can prove fatal. Therefore, the clinical priority is to ensure careful replacement of fluid and electrolytes before surgery is performed. This may take several days. After introduction of a nasogastric tube and withholding of any further feeds, IV fluids are commenced.

Several regimens for fluid resuscitation in babies with IHPS have been described, with most units having a clear departmental policy. Regimens take into account fluids/electrolytes required for resuscitation, maintenance, and ongoing replacement (ml for ml of nasogastric loss). The key thing is to correct each element of the fluid/electrolyte imbalance. Perhaps counterintuitively, but of importance, is that potassium must always be included in the maintenance fluids even if the initial serum potassium is within normal limits. This is because the correction of the alkalosis is dependent on the kidneys reabsorbing hydrogen in exchange for potassium, and despite many of these babies having normal serum potassium, their total body potassium is often depleted. Failure to give potassium can lead to a worsening alkalosis. Naturally, serum electrolytes and acid–base balance need to be monitored regularly. (See also Intravenous fluid and electrolyte management in children and young people, *Surgery* 2019; **37**(4): 189–194.).

Surgical management

Once fluids and electrolytes have been normalized, definitive surgical correction of IHPS is achieved by performing a pyloromyotomy. The principles of pyloromyotomy are still the same as that described by Conrad Ramstedt in 1912. A longitudinal incision is made in the hypertrophied pylorus and the muscle fibres are 'spread', ensuring that the mucosa is left in tact. Traditionally, pyloromyotomy was performed through a right upper quadrant incision enabling access to the pylorus without excessive stretching of the stomach. While this approach is still undertaken by a number of paediatric surgeons today, the long-term cosmetic effect can be poor. An open peri-umbilical approach offers better cosmesis, but delivery of the pylorus can be harder. Laparoscopic pyloromyotomy using diathermy or arthrotome is now used widely in many centres, although the specific benefits, including reduction in recovery time and improved cosmesis, remain debated.

Irrespective of the approach, careful incision of the pyloric serosa is essential. In the open approach the pyloric tumour is held longitudinally between finger and thumb to stabilize the pylorus and to identify the extent of the hypertrophy (Figure 2). A superficial incision of the pyloric tumour is made from the antrum to the start of the duodenum with a scalpel or diathermy point. This superficial incision is opened further by blunt 'spreading' of the muscle with a Dennis–Brown pyloric spreader, a pair of mosquito forceps or the blunt end of a scalpel handle.

Particular care must be taken at the pyloro-duodenal junction where there is an increased risk of mucosal perforation. This junction can be identified by a visible change in the direction of the muscle fibres at this point; the pre-pyloric vein of Mayo is also a helpful landmark. Adequate pyloromyotomy is confirmed by the 'shoe-shine manoeuvre' being able to move either side of the hypertrophied muscle independently in opposite directions (Figure 3).

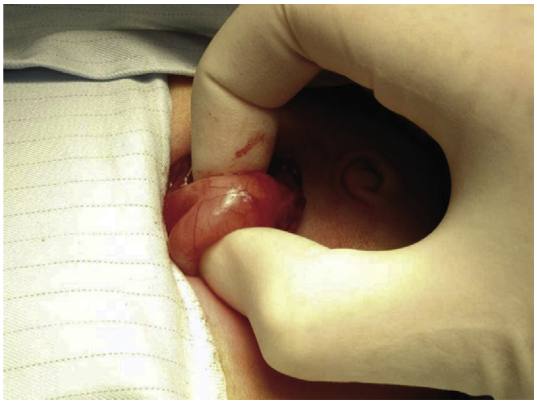

Figure 2 The pyloric tumour is held longitudinally between the finger and thumb to stabilize the pylorus and to identify the extent of the hypertrophy.

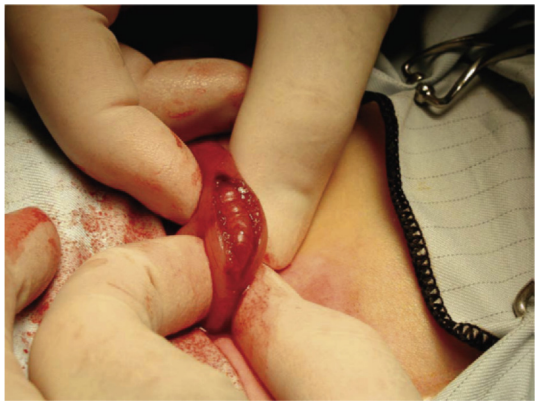

Figure 3 Adequate pyloromyotomy is confirmed if both sides of the hypertrophied muscle can move independently in opposite directions.

After pyloromyotomy, it is important to assess for any mucosal perforation by inspecting for signs of air or bile leaking from the pyloromyotomy site. Alternatively, the anaesthetist may inflate the stomach (via the nasogastric tube), with signs of air leakage looked for. This can be observed more readily by dripping saline onto the exposed mucosa during gastric air insufflation. If a perforation is identified, it is oversewn using an interrupted 5/0 absorbable suture or an omental patch loosely sutured over the perforation.

Postoperative management

Providing that a perforation has not been identified during the operation, the nasogastric tube can be removed at the end of the procedure. If a perforation has occurred, the nasogastric tube should be kept in place for 48 hours and the baby kept nil-by-mouth and given antibiotic cover. There are a number of different protocols used for the timing of reintroduction of feeds postoperatively, but there is no evidence whether early or later feeds are more beneficial. The incidence of vomiting increases with earlier feeding regimens, though early feeding does not appear to influence the total length of hospital stay.

Many centres in the UK restart feeds 4–6 hours after the operation. Regardless of feeding protocol, parents should be warned that some postoperative vomiting is common (particularly if the history of IHPS has been prolonged) and is not usually a feature of concern after initiating feeds. Babies usually go home within 1–2 days of surgery.

Complications and outcome

The majority of babies undergoing pyloromyotomy in the developed world today will have no long-term sequelae. Current data show that in the developed world, mortality from IHPS is almost unknown. To put this in context, the mortality was approaching 50% when Ramstedt reported his operative procedure. This significant mortality reduction has largely been achieved by a better understanding and availability of fluid/electrolyte correction, and the development of better paediatric anaesthesia.

Minor complications (including wound infections and dehiscence) occur in about 1% of patients. Postoperative vomiting is not unusual and is usually secondary to the decreased gastric motility that temporarily persists after surgical intervention. Normal gastric motility is usually fully re-established 1 week after the operation. Persistent vomiting can occur with incomplete myotomy, but is more often caused by concomitant gastro-oesophageal reflux, which may also need treatment. After discharge it is good practice for the health visitor to oversee adequate weight gain is taking place and if so, no further surgical follow up is required. Several studies of adults who have undergone pyloromyotomy for IHPS have demonstrated normal gastric emptying and normal pyloric measurements on ultrasound compared with age-matched, gender-matched controls. ◆

FURTHER READING

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