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Contemporary management of pyloric stenosis

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ABSTRACT

Hypertrophic pyloric stenosis is a common surgical cause of vomiting in infants. Following appropriate fluid resuscitation, the mainstay of treatment is pyloromyotomy. This article reviews the aetiology and pathophysiology of hypertrophic pyloric stenosis, its clinical presentation, the role of imaging, the preoperative and postoperative management, current surgical approaches and non-surgical treatment options. Contemporary postoperative feeding regimens, outcomes and complications are also discussed.

Introduction

Hypertrophic pyloric stenosis (HPS) is a common surgical cause of vomiting in infants. Our understanding of this condition comes largely from Hirschsprung's seminal work in 1888[1]; although both Hildanus (1627)[2] and Blair (1717)[3] have been attributed with earlier descriptions of HPS.

HPS usually presents in the first 2–12 weeks of life, with a peak incidence occurring during the fifth week of age.[4] Progressive hypertrophy of the pyloric muscle results in obstruction of gastric emptying. This causes the classical symptom of progressively worsening projectile vomiting after feeding. The vomiting is usually non-bilious and if untreated results in dehydration and hypochloraemic, hypokalaemic metabolic alkalosis. The incidence of HPS is reported as between 2 and 5 per thousand live births in the Western world,[5,6] with the European incidence ranging from 0.86 to 3.96 per thousand live births.[7] Males are more commonly affected than females with a ratio of at least 5:1,[4] although the explanation for this remains unclear, and the overall incidence is lower in African and Asian populations.[2] HPS strongly aggregates in families, even amongst distant relatives, and there is a high concordance between monozygotic twins.[7]

Despite extensive research of associated aetiological factors and possible pathophysiological mechanisms, the exact cause of HPS remains unclear. The nitric oxide pathway has been implicated; nitric oxide synthase (NOS) produces nitric oxide contributing to physiological relaxation of the pyloric sphincter. It has been postulated that deficiency of NOS in the pyloric muscle may be associated with pylorospasm and subsequent hypertrophy of the muscle,[8] although Serra et al.[9] found that there was only a marginally higher occurrence of genomic variants in the coding region of neuronal NOS in infants with HPS. More recently, Boybeyi et al.[10] have shown that NOS inhibition with L-NAME (an enteral hormone) appears to be a causative factor of HPS in a murine model, by increasing pyloric muscle thickness. The possibility of an infectious aetiology for HPS has also been recently discussed.[11]

Given the strong familial tendency, attempts have been made to identify genetic loci for HPS; regions on chromosomes 2, 3, 5, 7, 11 and 12 have all been implicated.[12–15] However, variations in the incidence of HPS and the numerous reported environmental associations with HPS suggest that it must, at least in part, be an acquired, as opposed to a congenital, condition. Maternal risk factors that have been reported include hyperthyroidism; nalidixic acid (synthetic quinolone antibiotic) use; young age; smoking; raised pre-pregnancy BMI; and intranasal decongestant use.[16–20] Reported infant risk factors include exposure to erythromycin and azithromycin in the first 2 weeks of life and prematurity.[19,21] HPS incidence has been recently correlated with pesticide use[22] and its occurrence has also been reported as a transient phenomenon in infants receiving prostaglandin infusions.[23] Bottle feeding appears to be independently associated with HPS[24] and data from the Danish National Birth Cohort suggested that bottle-fed infants had a 4.6-fold increased risk of developing HPS compared with infants who were breastfed.[25]

Clinical presentation, diagnosis and imaging

The clinical presentation of HPS is changing. The classical presentation is described as an infant with projectile, non-bilious

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vomiting with a hypokalaemic, hypochloraemic metabolic alkalosis and a palpable, hypertrophied pyloric muscle and an 'olive' in the abdomen. Palpation of the hypertrophied pyloric muscle is deemed diagnostic and has been previously reported as having a 99% positive predictive value.[26] However, the diagnosis is now frequently confirmed using ultrasonography, the sensitivity and specificity of which approaches 100% in experienced hands.[27] The use of this imaging modality has gradually increased since 1977 when Teele and Smith first published their article on the use of diagnostic ultrasonography to diagnose HPS.[28] Although they originally used pyloric diameter as the diagnostic criterion, a pyloric muscle thickness of greater than 3 mm and a pyloric canal length of 15 mm or greater are now generally considered diagnostic.[27] There is a positive correlation between pyloric muscle thickness and patient weight and age, and up to 5% of infants with HPS will have a pyloric muscle thickness of less than 3 mm.[29,30] Interestingly, studies have shown that the accuracy of both emergency physician-performed and surgeon-performed ultrasonography approaches that of radiologists for the diagnosis of HPS.[31–33]

A retrospective review by Glatsein et al.[34] found that only 13.6% of infants had a palpable 'olive' at presentation, compared to more than 50% of infants in older studies. This and other studies also found that fewer infants now present with severe electrolyte abnormalities.[35–37] Tutay et al.[37] found that serum bicarbonate was normal in 62% of infants, serum potassium was normal in 57% and serum chloride was normal in 69% of infants with HPS at presentation. It seems likely that increased use of ultrasound has led to an earlier confirmation of diagnosis with less opportunity for dehydration and electrolyte disturbance than in days gone by. Noteworthy is the fact that bilious vomiting does not exclude a diagnosis of HPS and has been reported to occur in up to 4% of infants with HPS.[35]

Preoperative management

The surgical management of HPS is not an emergency and should be deferred until the infant is appropriately resuscitated. Fluid resuscitation should be initiated in the referring hospital and should be based upon the degree of electrolyte abnormality and the level of dehydration. Vomiting of gastric contents leads to depletion of sodium, potassium, chloride and hydrogen ions, eventually resulting in the classical hypochloraemic, hypokalaemic metabolic alkalosis. The usual ability of the kidneys to maintain a normal pH by excreting bicarbonate is impaired by chloride depletion. Excess bicarbonate is instead reabsorbed in an attempt to maintain electrochemical neutrality. This exacerbates the alkalosis. Furthermore, the kidneys conserve potassium at the expense of hydrogen ions, leading to paradoxical aciduria, exacerbating the alkalosis further.[2]

The aim of initial fluid resuscitation in infants with HPS is to correct dehydration and these biochemical changes. An example of a suitable, initial fluid regimen is as follows: 0.45% or 0.9% sodium chloride with 5% dextrose and 10–20 mmol/L of potassium chloride at a rate of 150 mL/kg/d. This can be reduced to 100 mL/kg/d when the serum bicarbonate is <25 mmol/L. Volume correction can be administered as required (10–20 mL/kg of 0.9% sodium chloride). If a nasogastric (NG) tube has been placed, aspirates can be replaced mL for mL with 0.9% sodium chloride with 13.5 mmol of potassium chloride.

Although severe metabolic alkalosis can be potentially life-threatening, there is limited evidence in the literature to suggest what level of alkalosis is acceptable for an infant to safely undergo a general anaesthetic.[38] Metabolic alkalosis can potentially affect the respiratory drive of an infant and has been associated with apneas and extubation difficulties.[39] Surgery is usually performed once the serum bicarbonate and serum chloride are within the normal range and this will often occur within 24 h of commencing resuscitation.[38]

For infants with HPS, it is widely accepted that the stomach should be emptied using an NG tube immediately prior to the general anaesthetic, to reduce the risk of aspiration of gastric contents. However, there is debate about whether an NG tube is truly necessary prior to this. Some paediatric surgeons advocate the use of an NG tube to decompress the stomach as much as possible in the preoperative period; whereas others believe that infants with HPS can tolerate their gastric secretions and that an NG tube may exacerbate the underlying electrolyte abnormalities.[40] Following a retrospective review, Flageole et al. recently performed a prospective, randomised controlled pilot trial and found that the presence of a preoperatively placed NG tube had no effect on postoperative emesis or length of hospital stay. They 'cautiously' state that a preoperative NG tube is unnecessary; although they admit that their study was underpowered to demonstrate this definitively.[41]

Operative management

Rammstedt[42] described the longitudinal, extramucosal division of the pyloric muscle in 1912, having performed the procedure on the child of a physician during the previous year. Interestingly, an operation note describing a very similar procedure, performed by Stiles in 1910, can be found in a recent article by Keys et al.[43] Although the approach to the abdomen has continued to evolve, the pyloromyotomy itself has remained relatively unchanged over the past century. An adequate pyloromyotomy extends from the vein of Mayo at the duodenal end to the circular fibres of the stomach wall proximally.[39] Following an adequate myotomy, the separated edges of the pyloric muscle should move independently, and filling the stomach with air demonstrates passage of gas into the duodenum. This manoeuvre also allows the myotomy site to be checked for a leak.

Although the traditional right upper quadrant approach provides excellent access to the pylorus, it does leave a scar that can become quite significant, as it grows with the patient. In the circumumbilical approach, introduced by Tan and Bianchi,[44] a semicircular incision is made in a supraumbilical skin crease and a skin flap raised along the linea alba, which is then opened longitudinally. The pylorus is delivered into the wound and the pyloromyotomy is performed. Although cosmetically superior, the downside to this approach occurs when difficulty in delivering the pylorus is encountered. There is a risk of serosal tear and the skin incision may need to be extended. Successful intracavitary pyloromyotomy has also been described and negates the need to extend the wound to accommodate the pylorus.[45,46]

The laparoscopic pyloromyotomy (LP) was introduced by Alain et al.[47] Conventionally, a 3–5-mm laparoscopic port and laparoscope are used in the umbilicus alongside a 'stab' incision in each hypogastrium. The pyloromyotomy may be performed with electrocautery or an arthrotomy knife. Neonatal laparoscopic surgery is known to be safe and induction of carbon dioxide pneumoperitoneum in neonates has been recently shown to have no impact on brain oxygenation.[48] LP has become increasingly popular over time, although the risks and benefits of this procedure, when compared with open pyloromyotomy (OP), are still widely debated. It is accepted that, as with all laparoscopic procedures, there is a learning curve for LP and this has been estimated at 35 procedures.[49]

There have been several randomised controlled trials (RCTs) and meta-analyses comparing various outcomes of LP with OP. Although these studies offer a good overview of complication rates, time to achieve full enteral feeding and length of