

Infantile Hypertrophic Pyloric Stenosis: Review of Pathophysiology, Clinical Presentation, Treatment and Outcomes

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ABSTRACT

ARTICLE DETAILS

Infantile hypertrophic pyloric stenosis (IHPS) is considered one of the most common pediatric surgical conditions. Presentation is most common between the ages of 2 to 12 weeks with a mean and median age of 5 weeks. IHPS has been reported to be more common in males than females (5:1) and in first-born children (30-40%). Throughout history, the etiology of this condition has been studied, discovering multiple genetic and environmental factors, such as newborns with increased acid secretion, nitric oxide synthase deficiency, poor innervation of the pyloric muscle, smoking during pregnancy, neonatal use of macrolides and cesarean section; however, the exact cause remains uncertain. The classic clinical presentation begins with a recent history of severe, nonbilious projectile vomiting immediately after feeding and a strong, persistent appetite. During abdominal examination, palpation of the "pyloric olive" in the epigastrium by an experienced doctor is considered a pathognomonic sign. Laboratory tests may show low chloride, potassium, and hydrogen ion levels, leading to metabolic alkalosis due to persistent vomiting. Ultrasound is the gold standard imaging study if history and physical examination are insufficient to diagnose IHPS with a reported sensitivity and specificity of up to 98% and 100% by allowing evaluation of pyloric morphology and behavior. Pyloromyotomy is considered the definitive treatment for infantile hypertrophic pyloric stenosis, but should not be performed until the baby is adequately resuscitated. Patients typically experience positive outcomes after pyloromyotomy, with low morbidity and mortality rates. Most cases recover quickly and without complications.

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INTRODUCTION

Infantile hypertrophic pyloric stenosis (IHPS) is considered one of the most encountered pediatric surgical conditions. ^(1,2) It was first described in the early 17th century by Fabricius Hildanus. Although, our contemporary understanding comes from Hirschsprung's work in the late 1800s. ⁽³⁾ Nowadays, IHPS remains a common condition in newborns causing gastric outlet obstruction and leading to a progressive postprandial nonbilious projectile emesis. Presentation is most common between the ages of 2 to 12 weeks with a mean and a median age of 5 weeks. ⁽¹⁾ Initial treatment should be medical, as surgical management is not an emergency and is deferred until the patient is appropriately resuscitated. ^(4,5)

EPIDEMIOLOGY

IHPS has been reported to be more common in males than females (5:1) and firstborn children (30-40%) with a historical incidence of 2 to 5 cases per 1000 live births in the United States. ^(1, 2) On the other hand, Africa and Asia had

reported an incidence of less than 1 case per 1000 live births. ⁽¹⁾

Hypertrophic pyloric stenosis prevalence varies by maternal ethnicity being higher among white and Hispanic mothers and less common in African populations. ^(6,9) The genetic predisposition of IHPS is more common in children of mothers with a history of IHPS, affecting 19% of male children and only 7% of female children; while only 5% of men and 2.5% of women whose father had IHPS developed the condition. ⁽⁷⁾

PATHOGENESIS

The pyloric sphincter is made up of two layers of muscles – an inner circular ring and an outer longitudinal layer. The hypertrophy of these muscular layers leads to the narrowing and elongation of the pyloric channel, gastric outlet obstruction with compensatory dilation, hypertrophy, and hyperperistalsis of the stomach. ^(2,6,8)

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Throughout history, the etiology of this condition has been studied discovering multiple genetic and environmental factors. However, the exact cause remains uncertain. ^(1,2)

One proposed mechanism suggests that newborns with IHPS secrete more acid, causing repeated and increased contraction of the pyloric sphincter leading to hypertrophy development. Another theory identified the nitric oxide pathway as a potential factor; Nitric oxide synthase (NOS) deficiency can lead to impaired physiologic relaxation of the pyloric sphincter, pylorospasm, and hypertrophy. ⁽¹⁾

Other proposed mechanisms include poor pyloric muscle innervation, decreased intestinal pacemaker cells of Cajal, pesticide exposure, milk curds causing pyloric edema, and *Helicobacter pylori* infection. ^(1,2,6) Genetic causes have been described, showing significantly higher rates of disease among family members, with an overall heritability estimate of 87%. These findings suggested that IHPS must be at least partially acquired. ⁽¹⁾

Additional factors include smoking during pregnancy, neonatal exposure to macrolide antibiotics including erythromycin and azithromycin (especially if used before two weeks of age), young maternal age, maternal hyperthyroidism, prematurity, small for gestational age, and cesarian section. ^(1,6) Some genetic syndromes have higher rates of HPS such as Cornelia de Lange syndrome, Smith-Lemli-Optiz syndrome, Apert syndrome, Down syndrome, and Trisomy 18 syndrome ⁽⁹⁾

CLINICAL PRESENTATION

Infantile hypertrophic pyloric stenosis is a condition that usually appears when infants are between 3 to 8 weeks old. ⁽²⁾ The classic onset starts with a recent history of forceful projectile nonbilious emesis immediately after feeding, and a persistent strong appetite. Despite changing feeding regimens or formulations, these symptoms progressively worsen over weeks. ⁽¹⁾ However, projectile vomiting is not always present, especially in early diagnosis, and bilious emesis should not exclude this diagnosis as it has been reported in up to 4% of the cases. ^(1,2)

The prolonged vomiting can lead to weight loss and severe dehydration signs. ⁽²⁾ During abdominal examination, the palpation of the 'pyloric olive' in the epigastrium by an experienced physician is considered a pathognomonic sign. The Olive must be smooth, hard, mobile, oblong, and 1.5 to 2 cm in size. The examiner's ability to identify it ranges from 40-50%, and it's easier to palpate in an infant with a decompressed stomach. ^(5,9)

On the physical exam, peristaltic waves moving from left to right caused by the stomach attempting to pass content throughout the pyloric obstruction after a test feeding may be present. ⁽¹⁾

DIAGNOSIS

Over the course of time, the classic presentation has become less common because of an earlier approach ⁽²⁾ However,

laboratory tests may show low levels of chloride, potassium, and hydrogen ions, leading to metabolic alkalosis due to persistent vomiting. ⁽¹⁾ Elevated blood urea nitrogen (BUN) and creatinine are also helpful makers of dehydration. Ultrasound is the gold standard imaging study if history and physical examination are not sufficient to diagnose IHPS with a reported sensitivity and specificity of up to 98% and 100% by allowing the evaluation of the pyloric morphology and behavior. ^(9,8)

The diagnosis on ultrasound is characterized by pyloric muscle thickness (PMT) > 3 mm and pyloric canal length > 15 mm. The real-time pyloric function should demonstrate the failure of relaxation of the pyloric canal and abnormal flow and peristalsis in an infant with IHPS. ^(1,9) However, premature, or small infants may not fit within standard diagnostic criteria, so it should be taken into consideration when interpreting ultrasound results. ^(9,11) An abdominal X-ray can reveal the presence of an enlarged gastric gas bubble. ⁽⁹⁾

Alternative imaging options for cases of uncertain diagnosis include upper gastrointestinal contrast studies which can show a narrow string. However, it's important to consider that fluoroscopy is time-consuming, involves radiation exposure, and its sensitivity depends on the skill of the examiner. ^(1,6)

DIFFERENTIAL DIAGNOSIS

Gastroesophageal reflux disease (GERD) is a common condition that can be mistaken for IHPS due to the similar symptoms of emesis after feeding. However, while IHPS emesis is severe, GERD-related vomiting is less forceful and smaller in volume and diagnosis can be confirmed through tests like an upper gastrointestinal series, endoscopy, and pH/impedance monitoring. ^(1,9)

Another common differential diagnosis is overfeeding, although patients who are overfed may vomit and lose interest in feeding those with IHPS remain extremely hungry. ⁽⁹⁾ When history suggests hypertrophic pyloric stenosis, but no olive is palpable on examination and ultrasonography fails to detect it, other congenital malformations such as malrotation and Hirschsprung disease should be considered. ⁽⁹⁾

TREATMENT

Preoperative and medical management.

Pyloromyotomy is considered the definitive treatment for infantile hypertrophic pyloric stenosis, but it should not be undertaken until the infant is appropriately resuscitated because the metabolic alkalosis can affect the infantile respiratory drive leading to difficulty in extubation and apnea. ⁽¹⁾

The patient must receive intravenous fluids to restore acid-base balance and establish adequate urine output. ⁽⁵⁾ The recommended pre-operative resuscitation fluid includes an isotonic fluid with 5% dextrose at 1.5 to 2 times the maintenance rate or 150 ml/kg/day. ⁽¹⁾ Patients with severe volume depletion may also require a bolus with 10 – 29 ml/kg

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of 0.9% normal saline before the continuous infusion. ⁽¹⁾ Potassium should be included in a concentration of 10-20 mEq/L until urine output has normalized at 1-2 ml/kg/hour due to the risk of rebound hyperkalemia. Correction of fluid and electrolyte deficits can take days before the patient can go under surgery. ^(1,9)

Adequacy of resuscitation is determined by physical exam and laboratory markers. Improvement should include skin turgor, moist mucous membranes, normal urine output, and resolution of hypotension and tachycardia if present. ⁽¹⁾

Surgical management.

The first successful surgery for IHPS was a gastroenterostomy performed in 1898 by Lobker. This procedure was the standard approach for 10 years and was associated with high mortality (61%). In 1903 Dent performed the first pyloroplasty, and over the course of time, different surgeons independently performed an extra mucosal pyloroplasty transversally until 1911 when Ramstedt did the first extra mucosal longitudinal pyloromyotomy using a transverse right upper quadrant abdominal laparotomy. ⁽¹²⁾ The main goal of pyloromyotomy was to divide the pylorus longitudinally down to the submucosal layer from the circular fibers of the stomach wall. ⁽¹⁾

The concept of pyloromyotomy has remained, although the surgical approach has evolved over time. The right upper quadrant open approach was updated to a circumumbilical incision by Tan and Bianchi. With this approach, a semicircular supra-umbilical incision is placed in the natural umbilical skin fold. A longitudinal midline incision of the linea alba is made, and the peritoneal cavity is entered. If greater access is needed small transverse skin extensions can be made ^(10,12)

Besides the type of incision, the pylorus must be identified and delivered outside the abdomen. The prepyloric vein of a Mayo marks the pyloroduodenal junction and hence the distal end of the pylorus. Then, the surgeon makes a longitudinal seromuscular incision along the length of the hypertrophic muscle. Some surgeons would test adequacy as evidenced by the independent movement of the pyloric muscle lips called the 'shoe-shine maneuver' ⁽¹²⁾

In modern times, it is possible to do a laparoscopic approach to perform the pyloromyotomy with minimal invasion. This requires introducing a 3 mm or 5 mm port in the umbilical fold. once a capnoperitoneum is established, two stab incisions in the upper abdomen are made by introducing 3 mm instruments without trocars. The surgeon grasps the duodenum just distally or at the level of the hypertrophied pylorus using atraumatic forceps and performs the longitudinal pyloromyotomy in the avascular plane. ⁽¹²⁾ Adequacy can be tested by distension of the stomach with air which should pass into the duodenum. ^(1,2) Whether the laparoscopic approach is superior to open pyloromyotomy has been debated for a long time, but a consensus has not been reached. ^(5,10,12)

Postoperative management.

After pyloromyotomy, patients require at least 24 hours of inpatient observation due to the risk of postoperative apnea and the need for pain management. ⁽¹⁾ Patients may be discharged once they're feeding at goal volumes. However, patients with severe metabolic derangement are associated with longer time to full feeds. ⁽²⁾ When infants experience persistent vomiting beyond 7 to 10 days after surgery, a contrast study should be performed to evaluate an incomplete pyloromyotomy. ⁽²⁾ Non-surgical options for patients who cannot undergo surgery include atropine sulphate as a pyloric antispasmodic agent or endoscopic balloon dilatation ⁽¹⁾

OUTCOMES

Patients commonly experience positive outcomes following pyloromyotomy, with low morbidity and mortality rates. Most of the cases recover quickly without complications. ^(1,2) After surgery, postoperative ultrasound studies have shown that the pyloric muscle heals and returns to normal thickness and function within a month. ⁽⁷⁾ Several complications include mucosal perforation, incomplete pyloromyotomy, and surgical site infection with an overall rate between 4.6 and 12% ^(2,1)

Postoperative emesis is common with an incidence of 25 to 90%, it usually resolves without any additional intervention. If emesis persists concomitant gastroesophageal reflux should be suspected. ⁽²⁾

CONCLUSION

Infantile hypertrophic pyloric stenosis is a condition that affects infants and can lead to clinical deterioration of the pediatric patient if there is no adequate diagnostic suspicion. It is important to suspect the disease in those patients between 3 and 8 weeks of age, who present visible clinical deterioration due to dehydration, a strong appetite but with intolerance to oral intake, in addition to non-bilious projectile vomiting, as well as identify the pathognomonic sign known as "pyloric olive".

Indicate imaging studies such as abdominal ultrasound, which has adequate sensitivity and specificity, and is very useful in case of diagnostic doubt. Remember that the first line of treatment will be life support, monitoring of the respiratory pattern, management of hypochloremic metabolic alkalosis, correction of hydroelectrolyte disorders, and adequate resuscitation with intravenous fluids. Once the patient is stabilized, consider pyloromyotomy as a definitive treatment for this condition, with strict monitoring 24 hours after the surgical procedure, and inform the parents of the possible post-surgical complications that could occur, although their frequency is generally less than 12%.

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