Infantile Hypertrophic Pyloric Stenosis (IHPS) — Causes and Symptoms

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Pyloric stenosis, also known as infantile hypertrophic pyloric stenosis (IHPS), is a condition that is characterized by pyloric muscle hypertrophy and hyperplasia, which leads to gastric outlet obstruction. Infants usually present in their third week of life with repeated, projectile vomiting that is often associated with an olive-like palpable mass in the abdomen. The clinical presentation is usually adequate to make the diagnosis; ultrasonography can help confirm the diagnosis of pyloric stenosis in most patients. Surgical correction with pyloromyotomy is the treatment of choice.

Definition of Pyloric Stenosis

Infantile hypertrophic pyloric stenosis (IHPS) is a condition caused by hypertrophy and hyperplasia of the muscularis propria of the pylorus in infants. The pylorus opens and closes as needed, to allow food to pass through the stomach into the intestine. This hypertrophy/hyperplasia eventually results in gastric outlet obstruction.
Epidemiology of Pyloric Stenosis

Pediatric pyloric stenosis is a relatively common disorder in the United States with an estimated incidence of up to **4 per 1,000** live births. Fortunately, this condition is not associated with significant mortality.

For reasons poorly understood, pyloric stenosis seems to be slightly more common among Caucasians than other ethnic groups. Additionally, pyloric stenosis is **four times more common in males**, and 30 to 40% more common in first-born males.

Most patients are diagnosed at the age of three weeks. By 18 weeks, almost all patients with pyloric stenosis would have already presented to a medical practitioner.

Etiology of Pyloric Stenosis

The etiology of IHPS appears to be **both genetic and environmental**. For instance, exposure to **bottle-feeding** and **macrolide antibiotics** has been reported to increase risk. Additionally, infants who have **abnormal innervation of the myenteric plexus** or **deficiency of nitric oxide synthase** are at risk of developing IHPS.

The etiology is unclear but there appears to be an association with the following factors:

- Maternal smoking
- Bottle feeding
- Genetic component
- Use of erythromycin/azithromycin
Pathophysiology of Pyloric Stenosis

Hypertrophy and hyperplasia of the two layers (circular and longitudinal) of the pylorus lead to the development of symptoms in infants, which eventually results in obstruction of the gastric outlet causing abdominal distension. If left untreated, it may progress to gastric dilatation.

Signs and symptoms of pediatric pyloric stenosis are as follows:

- Nonbilious projectile vomiting after three weeks of age
- “Happy vomiter”
- Palpable olive-shaped mass in the right upper quadrant of the abdomen
- Gastric peristaltic waves
Causes of pyloric stenosis

Although the causes of pyloric stenosis are unknown, environmental and genetic factors might play a role. IHPS is usually not congenital and probably develops later. Several mechanisms and hypotheses have been suggested to explain why hypertrophy and hyperplasia of the pylorus occur in the first place. Some of the causative factors are as follows:

- Patients with a family history of IHPS and low nitric oxide synthase levels
- Exposure to antibiotics, such as macrolides and erythromycin, at 2–6 weeks of age
- Low serum-lipid levels
- Persistent duodenal acidity

Nitric oxide plays a vital role as a neurotransmitter in the gastrointestinal tract and relaxes sphincters by inducing the relaxation of smooth muscle. Patients who are deficient in nitric oxide synthase have an increased tone of the lower esophageal sphincter causing achalasia, increased tone of the pylorus causing pyloric stenosis, and impaired gastric motility that can lead to gastroparesis.

There is no single gene that can be directly linked to pediatric pyloric stenosis; however, familial aggregation is usually evident in most cases.

Clinical Presentation of Pyloric Stenosis

Since obstruction is at the gastric outlet, patients typically experience non-bilious vomiting, which can be projectile. After vomiting, infants get hungry again. Despite increased feeding, they show poor weight gain due to repeated vomiting after almost every feed.

This presentation is usually evident at three weeks of age. If an adequate fluid replacement is not attempted, the infant can develop dehydration and lethargy.

A physical examination can reveal an olive-like, non-tender mass in the upper right quadrant of the abdomen. Additionally, in cases of complete gastric obstruction, gastric peristaltic waves are visible before vomiting. Fortunately, due to current rapid methods of diagnosis, these classical symptoms are no longer common.

Diagnostic Work-up for Pyloric Stenosis

Due to repeated vomiting, patients with IHPS can become dehydrated. Serum electrolytes, blood-urea-nitrogen, creatinine, and blood pH should be monitored.

Patients with repeated vomiting episodes due to IHPS lose large amounts of acid. This eventually leads to metabolic alkalosis. Loss of fluid also leads to contraction of the extracellular fluid; thus, the kidneys compensate by activating the renin-angiotensin system. Increased angiotensin II and aldosterone levels lead to increased potassium secretion (hypokalemia), further hydrogen secretion, and HCO₃⁻ reabsorption (more alkalosis). This metabolic process is called contraction alkalosis.

When dehydration is more severe, patients can develop hypernatremia, which can have deleterious effects on the brain. Even though these laboratory findings are helpful in the diagnosis of IHPS, they are becoming less common today due to the advances in imaging studies.
Infants between three and twelve weeks of age, who present with repeated vomiting after each meal and reveal an olive-like mass upon abdominal examination can be clinically diagnosed with IHPS without being indicated additional studies. When the clinical presentation is less typical, i.e. repeated vomiting without an olive-like mass, ultrasonography (USG) is the best imaging modality to aid in the diagnosis.

On ultrasonography, an increase in the thickness of the pyloric muscles is usually the main finding of pyloric stenosis. Pyloric muscle thickness of > 4 mm is indicative of pyloric stenosis.

Upper gastrointestinal barium studies are not routinely used in the evaluation of IHPS and should be reserved for infants with non-conclusive USG. A ‘double track’ sign is defined as the appearance of two thin barium lines between the hypertrophic layers of the pylorus and is found in both pyloric stenosis and pylorospasm.

Upper gastrointestinal endoscopy is the last resort in the diagnostic work-up of pyloric stenosis. It can help exclude possible complications such as Mallory-Weiss tears due to repeated vomiting and confirm the diagnosis of pyloric stenosis by direct visualization of the gastric antrum and pylorus.

When USG and upper gastrointestinal studies are unhelpful in diagnosing clinical signs, endoscopy is considered as the last option.

Treatment of Pyloric Stenosis

It is important to evaluate infants for signs of dehydration and assess the degree of fluid loss. Infants who are in shock due to severe fluid loss should undergo fluid replacement therapy according to the pediatric advanced life support guidelines.

Accordingly, correction of the electrolyte imbalance, restoration of the normal acid-base balance, and replacement of fluids should be prioritized.

The first step in the emergency management of an infant with IHPS is to infuse a bolus of 20 mL/kg of a crystalloid solution to correct dehydration.

Infants who are not dehydrated should receive maintenance fluid replacement twice to treat metabolic alkalosis. The maintenance fluid is either a 5% dextrose or 0.33% sodium chloride solution, depending on whether the patient is hyponatremic or
hypernatremic at presentation. Infants with pyloric stenosis also develop hypokalemia and should receive up to 4 mEq KCl/100 mL fluid.

Once the infant is hemodynamically stable, the treatment for pyloric stenosis is corrective surgery. The currently recommended procedure is Ramstedt pyloromyotomy, in which the pylorus and antrum are excised while the muscle layer is left intact. This procedure can also be done laparoscopically.

Laparoscopic pyloromyotomy has been found to be safe and as effective as the open approach, but with lower morbidity, a smaller skin incision, and faster recovery.

Infants who are poor surgical candidates might benefit from medical treatment, which consists of nasogastric administration of atropine. Atropine causes relaxation of the pylorus muscles and eventually relieves obstruction after 21 days of administration. Atropine administration has only been evaluated in a single study that included 12 patients; therefore, generalization or recommendations cannot be made until larger, well-controlled clinical trials have been conducted.

Prognosis of Pyloric Stenosis

In patients with delayed or missed diagnoses, there exists a risk of dehydration leading to shock. The prognosis for such cases with early diagnosis is good after laparoscopic pyloromyotomy.

Frequently asked question:

Is pyloric stenosis in adults possible?

Pyloric stenosis mostly occurs in adults as a result of other conditions such as ulcers, cancer, or abdominal surgery. Although rare, it can be idiopathic and occur in middle-aged men.

References


Pediatric Pyloric Stenosis via medscape.com


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