Pyloric Stenosis (Gastric Outlet Obstruction) — Causes and Symptoms

Pediatric pyloric stenosis, also known as infantile hypertrophic pyloric stenosis, 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 associated with an abdominal olive-like palpable mass. The clinical picture is usually enough to make the diagnosis, but ultrasonography can help confirm the diagnosis of pyloric stenosis in most of the patients. Surgical correction with a pyloromyotomy is the treatment of choice.
Definition of Pediatric Pyloric Stenosis

Pediatric pyloric stenosis is defined as the hypertrophy and hyperplasia of the muscularis propria of the pylorus in infants. This hypertrophy/hyperplasia results eventually in gastric outlet obstruction.

Epidemiology of Pediatric 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, the condition is not
associated with significant morbidity or mortality.

For poorly understood reasons, pyloric stenosis seems to be slightly more common among whites compared to other ethnic groups. Additionally, pyloric stenosis is 4 times more common in males, especially first-born males.

Most of the patients are diagnosed at the age of 3 weeks. By 18 weeks, almost all patients with pyloric stenosis would have already presented to the health care system.

Etiology of Pediatric Pyloric Stenosis

The etiology of pediatric pyloric stenosis seems to be both genetic and environmental. For instance, exposure to bottle-feeding and macrolide antibiotics has been linked to an increased risk of infantile hypertrophic pyloric stenosis, another name for pediatric pyloric stenosis.

Additionally, infants who have abnormal myenteric plexus innervation or deficiency of nitric oxide synthase are at risk of developing pyloric stenosis.

Pathophysiology of Pediatric Pyloric Stenosis

For pediatric pyloric stenosis to become symptomatic, hypertrophy associated with hyperplasia of the pylorus should happen. This eventually leads to obstruction of the gastric outlet causing abdominal distension and if left untreated gastric dilatation.

Several mechanisms and hypotheses have been put to explain why hypertrophy/hyperplasia of the pylorus occur in the first place. For instance, patients with family history of the disease are at risk of having neurons that are deficient of the enzyme nitric oxide synthase.
Nitric oxide plays a vital role as a neurotransmitter in the gastrointestinal tract where it inhibits sphincters and causes smooth muscle relaxation. Patients with 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 which could lead to gastroparesis.

There is no single gene that can be directly linked to pediatric pyloric stenosis but familial aggregation is usually evident in most of the cases.

Clinical Presentation of Pediatric Pyloric Stenosis

Because the level of the obstruction is at the gastric outlet, patients typically show non-bilious vomiting which can be projectile. After vomiting, infants are hungry again. Despite increased feeding, the infant will show poor weight gain due to repeated vomiting almost after each feeding.

This presentation is usually evident at 3 weeks of age. If adequate fluid replacement is not attempted, the infant can develop dehydration and become lethargic or have a decreased urinary output.

Physical examination can reveal an olive like non-tender mass in the upper right abdomen. Additionally, when gastric obstruction is almost-complete, gastric peristaltic waves can be visible before vomiting. Fortunately, the diagnosis nowadays is earlier and these classical symptoms are no longer common.

Diagnostic Work-up for Pediatric Pyloric Stenosis

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

Patients with repeated vomiting due to pyloric stenosis lose large amounts of acid. This eventually leads to metabolic alkalosis. The kidneys try to compensate for that by retaining hydrogen and secreting potassium, hence patients become hypokalemic.

When dehydration is more severe, patients can become hypernatremic which could have deleterious effects on the brain. Even though these laboratory findings are helpful in the diagnosis of pediatric pyloric stenosis, they are becoming less common today due to the advances in imaging studies.

Patients with repeated vomiting after each meal, who are between 3 to 12 weeks of age and who have an olive-like mass in abdominal examination do not need any further studies and the diagnosis of infantile hypertrophic pyloric stenosis can be made. When the image is less typical, i.e. repeated vomiting without an olive-like mass, ultrasonography is the best imaging modality to diagnose pyloric stenosis.

On ultrasonography, increased pyloric muscles’ thickness is usually the main finding in pyloric stenosis. A pyloric muscle thickness > 4 mm is diagnostic of pyloric stenosis.
Upper gastrointestinal barium studies should be reserved for patients with non-conclusive ultrasonography and not routinely used in the evaluation of pyloric stenosis. The double track sign is defined as the appearance of two thin barium lines between the hypertrophic layers of the pylorus and can be found in both pyloric stenosis and pylorospasm.

Upper gastrointestinal endoscopy is a last resort in the diagnostic work-up of pediatric 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 the pylorus.

**Treatment of Pediatric Pyloric Stenosis**

It is important to evaluate infants for signs of dehydration and to 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 guideline.

Accordingly, correction of electrolytes imbalances, restoration of the normal acid-base balance and replacement of fluid loss is essential at the emergency level.

The first step in the emergency management of the infant with pyloric stenosis is to **infuse a bolus of 20 mL/kg of a crystalloid** to correct dehydration.

Infants who are not dehydrated should receive 2 times their maintenance fluid replacement, which should contain either a 5% dextrose or a 0.33% sodium chloride solution depending on whether the patient is hyponatremic or hypernatremic at presentation. Patients with pyloric stenosis also develop hypokalemia and they should receive up to 4 mEq KCL per 100 ml fluid.

Once the infant is hemodynamically stable, corrective surgery for pyloric stenosis should be attempted. The currently recommended procedure is a 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 traditional procedure but with lower morbidity, smaller skin incisions and faster recovery.

Infants who are poor surgical candidates might benefit from **medical treatment**. Medical treatment consists of nasogastric administration of atropine which is
believed to result in relaxation of the pylorus muscles and eventually relieve the obstruction after 21 days of daily administration. Atropine administration was evaluated so far in a single study that included only 12 patients and generalization or recommendations cannot be made yet until larger, well-controlled clinical trials can be made.

References


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