Pyloric Stenosis

Description/Etiology

Pyloric stenosis (PS; also called infantile hypertrophic pyloric stenosis) is the most common cause of intestinal obstruction in infants and children and the most common condition requiring surgical intervention in the first year of life. In PS, abnormal thickening of the circular muscle of the pylorus causes progressive stenosis (i.e., narrowing) of the gastric outlet to the duodenum. PS is the result of both pyloric muscle hypertrophy (i.e., increased size) and hyperplasia (i.e., increased mass). Over time, pyloric thickening causes inflammation, edema and eventual occlusion of the pyloric canal. When the stomach can no longer empty into the small intestine, vomiting of all ingested material occurs. PS presents most often as an isolated anatomic abnormality, but it can be associated with intestinal malrotation, esophageal and duodenal atresia, and anorectal abnormalities. Prompt recognition and treatment are crucial to restoring nutritional health. Untreated, PS causes dehydration, metabolic acidosis, and failure to thrive. The exact pathogenesis is unknown but might involve neurotransmitter dysfunction.

Treatment consists of careful monitoring, maintenance of hydration and electrolyte balance, and surgical correction with pyloromyotomy. Open pyloromyotomy (OP; the traditional surgical procedure) uses an abdominal incision through which the pyloric muscle is incised and spread. Laparoscopic pyloromyotomy (LP; a less-invasive surgical alternative) is performed through two stab incisions, leaves minimal scarring, and is associated with successful short-term outcomes (e.g., emesis alleviation, reduced pyloric muscle thickness). IV atropine is alternative medical treatment that is reserved for cases in which anesthesia or surgery is not possible.

Facts and Figures

The global incidence is ~ 1–5:1,000 live births, but incidence varies widely with geographic region and ethnic origin. PS is more common in firstborn children and full-term infants, with a male-to-female ratio of occurrence of 2–5:1. Incidence is highest in White infants and lowest in Asian and Black infants. With early recognition and prompt treatment, death from PS is extremely rare.

Risk Factors

Risk factors include male gender, White race, maternal smoking, family history of PS, and oral erythromycin during the first 2 weeks of life. Maternal history of PS increases risk tenfold in infants. PS might be more common in bottle-fed than breastfed infants.

Signs and Symptoms/Clinical Presentation

Signs and symptoms most often occur within 2–8 weeks after birth; infants initially experience mild vomiting 30–60 minutes after feedings and projectile vomiting usually develops 1–2 weeks later. Visible peristalsis (i.e., gastric waves) from left to right across the upper abdomen soon after eating often appears before vomiting. Other signs include abdominal distension, dehydration, and weight loss. Bowel movements often decrease in number and stools can be loose, green, and contain mucus. The infant appears hungry and will continue to feed despite vomiting.
Assessment

› Patient History
• Obtain detailed history: Pattern/type of vomiting, feeding schedule, symptom duration, recent illness/medications, and behavioral changes
• Measure infant’s length and weight, comparing growth patterns to developmental norms

› Physical Findings of Particular Interest
• After 1–2 weeks of mild vomiting after feeding, projectile nonbilious (i.e., absence of bile) vomiting typically occurs 30–60 minutes after each feeding
  – Less frequent/less forceful vomiting can indicate gastroesophageal reflux or formula intolerance
• The enlarged pyloric muscle can be palpated as a small, hard, mobile, olive-shaped lump
• Depressed fontanelles, dry mucous membranes, poor skin turgor, reduced tearing, and lethargy can be present, indicating dehydration

› Laboratory Tests That Can Be Ordered
• Serum chemistry (electrolytes, pH, BUN, and creatinine) can show metabolic alkalosis in early-stage PS with ↓ chloride and ↑ bicarbonate levels, and acidosis in later-stage PS with ↓ potassium and ↓ bicarbonate levels
• Unconjugated bilirubinemia with jaundice can be present in later stages

› Other Diagnostic Tests/Studies
• Abdominal ultrasound can show abnormal pyloric thickness (> 4 mm), a narrow and elongated pyloric channel (> 14 mm), and redundant mucosa
• Upper GI series with barium swallow is usually performed; findings confirming PS include elongated pyloric channel, bulging of the pyloric muscle into the antrum, and the “double track sign” of two parallel barium streaks in a narrowed channel where barium should normally readily empty
• Upper GI endoscopy is used only when other studies are nondiagnostic

Treatment Goals

› Promote Optimal Gastrointestinal Function and Monitor for Complications
• Assess for signs of pain by observing for muscle tension, facial grimacing, and inconsolable crying; administer analgesics, as ordered
• Assess for signs of dehydration (e.g., poor skin turgor, sunken fontanelles, dry mucous membranes, urine output < 1 mL/kg/hr, and impaired consciousness [e.g., failure to rouse upon tactile stimulation])
  – Administer IV fluids to restore hydration and electrolyte balance, as ordered
• Monitor vital signs frequently, measure I & O, and review blood chemistry results, reporting abnormalities to treating clinician
• Follow facility pre- and postsurgical protocols if patient becomes a surgical candidate (e.g., for OP/LP); reinforce pre- and postsurgical education to the parent(s) and verify completion of facility informed consent documents
  – Monitor postsurgically for complications of swelling and esophagitis, which can cause vomiting; administer prescribed medications for symptomatic relief
  – Assist with prescribed feedings, which are usually initiated by introducing smaller quantities 12–24 hours postoperatively with the goal of full oral feedings within 36–48 hours

› Educate Parents About Treatment Options and Provide Emotional Support
• Assess parental coping ability and emotional status; reassure parents that PS is the result of a structural abnormality and not a consequence of their care
• Encourage parent(s) to participate in pre- and postsurgical care and encourage rooming-in per facility protocols.
  – Demonstrate how to be physically close to the infant and how to assist with supportive care and postsurgical feeding, as appropriate
• Educate that the patient has an excellent prognosis with treatment and if growth has been delayed, the infant will usually soon achieve normal developmental milestones

Food for Thought

› Although uncommon, bilious vomiting can occur in patients with incomplete pyloric obstruction, and thus does not rule out PS. Researchers who analyzed the medical records of 354 infants with PS found that bilious vomiting occurred in 1.4% (Piroutek et al., 2012)
The authors of a meta-analysis of randomized controlled trials comparing OP and LP found that the procedures are equally safe and effective, with a trend favoring LP for shorter surgery time, shorter length of stay, and shorter time to full feedings (Jia et al., 2011).

The concurrent incidence decline of sudden infant death syndrome (SIDS) and PS in Sweden from 1970–1997 following the “Back to Sleep” campaign promoting supine sleeping position for SIDS prevention suggests that the prone sleeping position might contribute to PS development or exacerbation.

Investigators in a study including 2933 participants reported that in infants diagnosed with hypertrophic PS who underwent pyloromyotomy, a history of premature birth is associated with increased mortality rate (Costanzo et al., 2017).

Red Flags

Careful physical examination is important when evaluating for PS; on palpation, the pyloric mass can be obscured by the liver, distended stomach, or tense abdominal muscles, or mistaken for the liver margin, contracted muscles, or upper kidney. According to researchers of a retrospective cohort study, ingestion of oral azithromycin and erythromycin places infants at increased risk for infantile hypertonic PS, particularly if the exposure occurred in the first 2 weeks of life (Eberly, 2015).

What Do I Need to Tell the Patient’s Family?

Most infants recover fully and quickly following either OP or LP.

Postoperative vomiting secondary to swelling or esophagitis is common for 24–48 hours.

Feedings are gradually advanced, as tolerated. Notify the treating clinician or seek immediate medical attention for new or worsening signs and symptoms, and keep scheduled follow-up appointments.

References