This obstruction results from hypertrophy of the circular and longitudinal muscularis of the pylorus and the distal antrum of the stomach. It occurs in approximately 3 of every 1000 live births and is 4 times more common in boys. Pyloric stenosis (PS) is relatively uncommon in African American and Asian infants. The observation that it occurs primarily in first-born infants has been disputed.

PS is seen more frequently in infants with affected siblings or parents than in children with no family history. Approximately 20% of the male and 10% of the female descendants of mothers with congenital PS have this condition. In contrast, PS occurs in only 5% of males and 2.5% of females whose fathers were affected. A multifactorial mode of inheritance has been postulated. The hallmark of PS is progressive, nonbilious projectile vomiting that typically begins when the infant is about 3 weeks old. The child is characteristically eager to feed immediately after vomiting. Occasionally, there may be blood in the vomitus as a result of gastritis or esophagitis. The infant fails to gain—or actually loses—weight. Approximately 2% to 5% of infants have unconjugated hyperbilirubinemia as a result of glucuronyl transferase deficiency secondary to caloric deprivation.

Physical examination is best accomplished when the infant's stomach is empty and he or she is resting quietly. Peristaltic waves that progress across the upper abdomen from left to right during feedings may be visible. A pyloric, olive-shaped mass that may be palpable in the right epigastrium is pathognomonic. As many as 7% of infants with PS have associated malformations, such as esophageal atresia, intestinal malrotation, inguinal hernia, cryptorchidism, or obstructive uropathy. The differential diagnosis for nonbilious emesis includes overfeeding, gastroesophageal reflux, milk allergy, pylorospasm, salt-wasting adrenogenital syndrome, increased intracranial pressure, congenital metabolic dysfunction, prepyloric antral web, and gastric duplication.

In the absence of a palpable pyloric mass, abdominal ultrasonography is the diagnostic test of choice. The most commonly used criteria for a positive ultrasonographic study include pyloric muscle thickness greater than 4 mm and pyloric channel length greater than 14 mm. Because these measurements are age-dependent, pyloric muscle thickness greater than 3 mm is diagnostic for PS in infants younger than 30 days. If ultrasonography proves nondiagnostic, an upper GI tract barium study is recommended. The classic radiographic contrast findings include a single string sign or double railroad track sign (produced by the narrowed pyloric channel), a shoulder sign (caused by the bulging of the pyloric mass into the antrum of the stomach), and delayed gastric emptying (A).

PS may be complicated by dehydration and hypochloremic metabolic alkalosis. The serum pH is high. The urinary pH is high initially, but eventually drops as the severe potassium depletion leaves only hydrogen ions to exchange with sodium ions in the distal renal tubules; this results in paradoxical aciduria.

Fluid and electrolyte disturbances must be corrected with intravenous solutions before surgical intervention. The surgical procedure of choice—pyloromyotomy—can be performed through a short transverse or periumbilical incision or laparoscopically. On gross examination, the pylorus appears
Pyloric stenosis is a narrowing of the pyloric channel, typically causing gastric outlet obstruction. The anatomy is characterized by an elongated and thickened pyloric channel, often simulating an olive (B). The surgical approach involves splitting the underlying pyloric mass without cutting the mucosa and then closing the incision. Postoperative complications are rare but may include persistent vomiting more than 48 hours after surgery, which suggests incomplete myotomy or underlying gastroesophageal reflux.