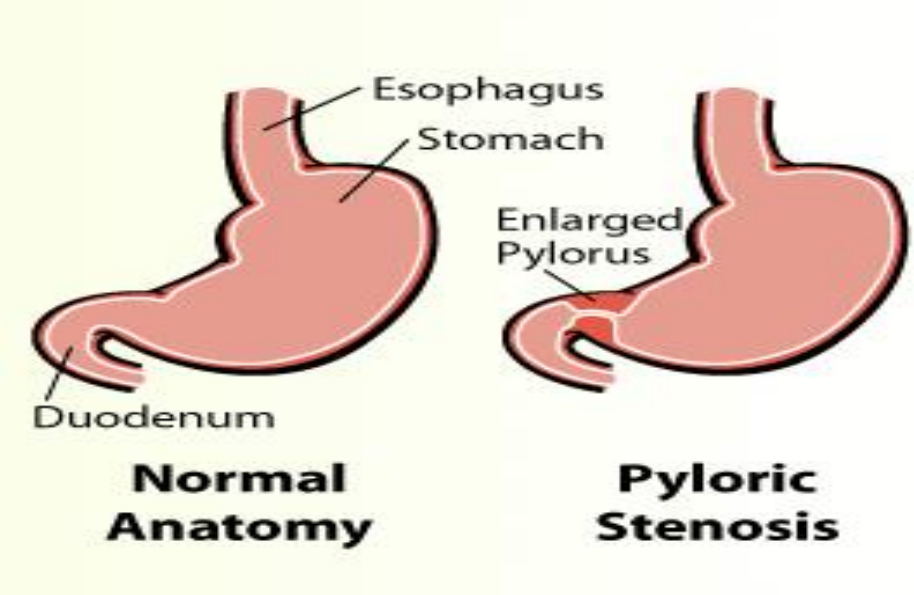
Infantile Hypertrophic Pyloric Stenosis



Infantile Hypertrophic Pyloric Stenosis

Disorder of young infants caused by hypertrophy of the pylorus, which can progress to near-complete obstruction of the gastric outlet, leading to forceful vomiting

- 2 to 3.5 per 1000 live births
- More common in males than females
- More common in infants born preterm as compared with those born at term
- 30 to 40 percent of cases occur in first-born children
- Less common in infants of older mothers

Etiology

Multifactorial

- Genetic predisposition
- Environmental factors

- Neonatal hypergastrinemia
- Gastric hyperacidity
- Prematurity

Environmental factors

- Maternal smoking during pregnancy
- bottle feeding rather than breastfeeding
- bottle feeding during the first four months of life

Genetic factors

- higher rate among monozygotic twins
- 20-fold increase among dizygotic twins or siblings

Macrolide antibiotics

- erythromycin
- azithromycin

particularly when administered to infants younger than two weeks of age

Pathophysiology

Marked hypertrophy and hyperplasia of the 2 (circular and longitudinal) muscular layers of the pylorus

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Narrowing of the gastric antrum

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Pyloric canal becomes lengthened, and the whole pylorus becomes thickened

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Stomach becomes markedly dilated

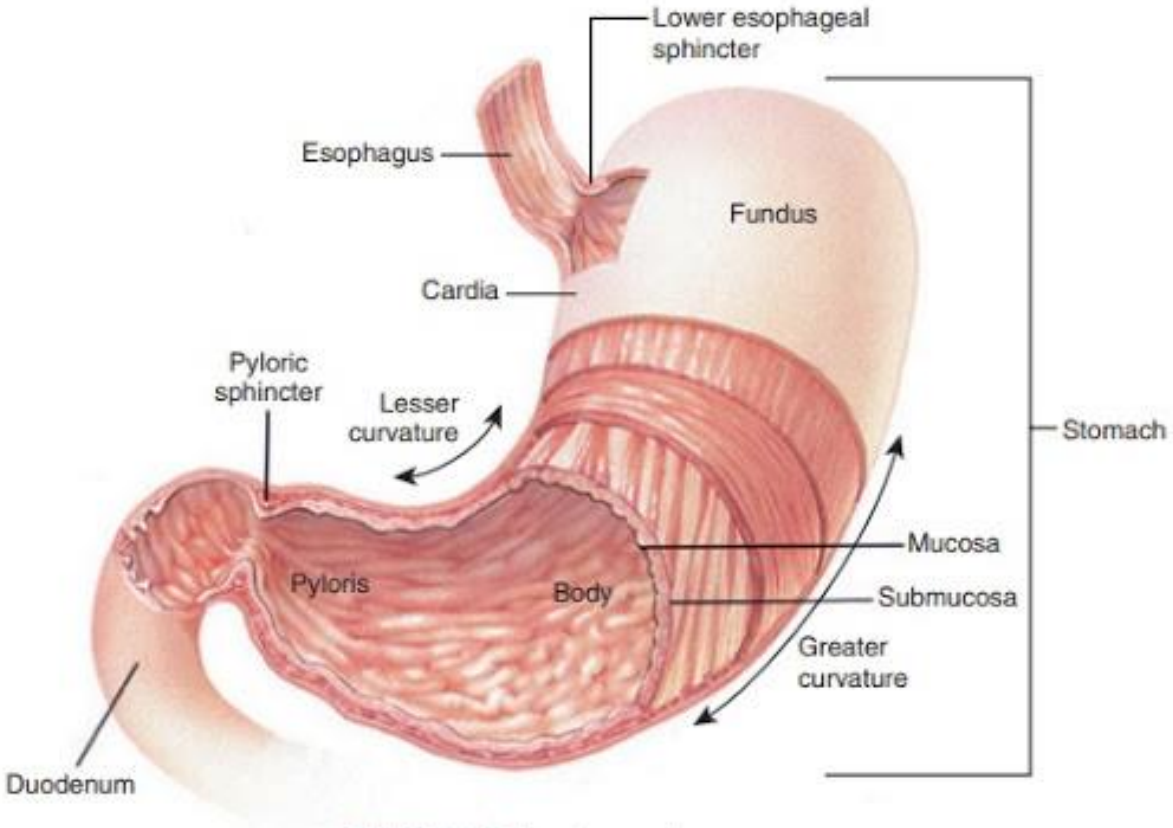

FIGURE 24-8 The stomach.

Clinical features

- Three- to six-week-old infant – usual age of presentation
- Immediate postprandial vomiting that is nonbilious and forceful
- Demands to be refed soon afterwards
- Rarely occurs after 12 weeks of age

Physical examination

- Emaciated
- Dehydrated
- Palpable "olive-like" mass at the lateral edge of the rectus abdominis muscle in the right upper quadrant of the abdomen

Atypical presentation

- Young infants with repeated nonbilious vomiting
- Hypochloremic alkalosis
- Rapid clinical improvement after rehydration
- Infants with congenital anomalies that affect swallowing - vomiting may not be projectile
- In premature infants –
  - Vomiting may be less forceful
  - Voracious appetite
  - Exaggerated gastric peristalsis may be lacking

Clinical associations

- Hyperbilirubinemia – most common clinical association
- Midgut malrotation
- Absent or hypoplastic mandibular frenulum
- Joint hypermobility
- Eosinophilic gastroenteritis
- Gastric ulcer
- Hiatal hernia

- Diaphragmatic hernia
- Congenital heart disease
- Esophageal atresia/tracheoesophageal fistula
- Cleft lip and palate
- Obstructive defects of the urinary tract
- Propionic acidemia
- Congenital nephrotic syndrome
- Congenital hypothyroidism

Investigations

- Serum electrolytes

1. Serum chloride and potassium - low
2. Bicarbonate - elevated (a hypochloremic alkalosis)
3. Hypernatremia or hyponatremia

- Blood urea nitrogen (BUN) and creatinine

Assess for dehydration and renal insufficiency

Imaging

- Can be confirmed by imaging
- Ultrasonography is the procedure of choice

1. Ultrasonography - "target" sign on transverse view
2. Fluoroscopic upper gastrointestinal series
3. Upper endoscopy

Treatment

Pyloromyotomy

- Definitive management
- Timing of surgery depends upon the clinical status of the infant
- Surgery should be delayed in the setting of dehydration and/or electrolyte derangements

Balloon dilation

- Endoscopically guided balloon dilation
- Does not reliably disrupt the seromuscular ring of the pylorus
- Best reserved for patients in whom general anesthesia would pose a significant risk or in whom a surgical approach to the pylorus is not possible

Conservative management

- Only for infants in whom a surgical approach is either not advisable or not feasible
- Anticholinergics - Oral and intravenous atropine sulfate
- Continuous nasoduodenal feedings

Until the obstructive process becomes less significant as the infant gains weight

Prognosis

- Surgery is curative with minimal mortality
- The prognosis is very good
- Complete recovery
- Catch-up growth