HYPERTROPHIC PYLORIC STENOSIS

Hypertrophic pyloric stenosis (HPS) is the **most common cause of gastric outlet obstruction** in infancy from **2-12 weeks CGA.** HPS is **rare before 2 weeks** of age in a term infant as **pyloric musculature hypertrophies** until gastric outlet obstruction occurs. This obstruction causes the classic symptoms and lab findings of HPS.

PRESENTATION

- Usually well before symptom onset (median 6 weeks)
- Non-bloody, non-bilious, **projectile** vomiting **after** feeds
- Appears **hungry** post-feed
- Dehydration and weight loss
- Distended abdomen
- Visible **peristaltic waves**

Although rare, if there is a palpable thickened mass in RUQ (the "olive"), this is pathognomonic of HPS.

RISK FACTORS

- 2-5 weeks at symptom onset
- Male gender
- First born
- Formula feeding
- In utero / neonatal macrolide exposure
- Parental history of HPS
- C section delivery
- Certain Genetic syndromes:
  Cornelia de Lanage Syndrome, Smith-Lemli-Opitz Syndrome, Apert Syndrome, Down Syndrome, and Trisomy 18 Syndrome

Differential Dx of Infantile Non-Bilious Emesis

- ❑ Hypertrophic pyloric stenosis
- ❑ GER/GERD
- ❑ Duodenal stenosis (proximal to Ampulla of Vater)
- ❑ Gastroenteritis
- ❑ Cow's milk protein intolerance
- ❑ Inborn errors of metabolism
- ❑ Liver disease
- ❑ Gastric, antral, or pyloric atresia
- ❑ Pyloric or antral membrane
- ❑ Gastric volvulus
- ❑ Overfeeding

INVESTIGATIONS

- ❑ Abdominal ultrasound: positive if muscle thickness >3mm and length ≥15mm
- ❑ Upper GI study if US unavailable → "string sign"
- ❑ Observed feeding trial if imaging is inconclusive
- ❑ Metabolic panel with electrolyte assessment
- ❑ Bilirubin if jaundiced

PATHOPHYSIOLOGY

- ❑ Impaired neuronal nitric oxide synthase synthesis may be impaired in HPS → disrupted smooth muscle relaxation in myenteric plexus → pyloric hypertrophy
- ❑ Gastric hyperacidity may also play a role in causing HPS, although the exact etiology is unknown.

ELECTROLYTE ABNORMALITIES IN HPS
⬇ Chloride
⬇ Potassium
⬆ pH, ⬆ HCO_3^-

SIGNS OF HYPOVOLEMIC SHOCK:
Severe dehydration:
- ❑ Altered mental status
- ❑ Impaired end-organ perfusion
- ❑ Decreased blood pressure

ALTHOUGH UNCOMMON IN HPS, RAPID FLUID RESUSITATION IS REQUIRED IF THESE SIGNS ARE PRESENT.

MANAGEMENT

Consult Gen Surg

1. Assess hydration status and correct fluid deficit
   - ❑ Electrolyte imbalance and dehydration must be corrected prior to anesthetic
2. Correct any metabolic or electrolyte abnormalities
3. Laparoscopic pyloromyotomy is the standard approach: surgery is only definitive treatment
4. Can resume oral feeds within a few hours of surgery, often with some regurgitation but an excellent long-term prognosis when identified early.

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