Exocrine Pancreatic Insufficiency (EPI)

Diagnosis:

- **Definition:** Insufficient pancreatic enzyme and/or bicarbonate secretion to match nutrient digestion/absorption needs. Clinical diagnosis not dictated by fecal elastase level.
- **Differential Diagnosis:** Other conditions that may impair digestion or absorption of nutrients, such as liver diseases, intestinal diseases (celiac disease, carbohydrate malabsorption, inflammatory bowel disease, diarrheal diseases), abetalipoproteinemia

Key History to Review:

- Pancreatitis, celiac disease, any other GI disease, fat soluble vitamin deficiencies (A, D, E, K); symptoms including steatorrhea, abdominal pain, weight loss/poor weight gain, bloating, rickets, easy bruising/bleeding, night blindness
- History of pancreatitis, pancreas-related surgery, Schwachman-Diamond syndrome
- Family history of pancreatitis, cystic fibrosis, celiac disease, autoimmune disease

Key Physical Exam Components:

- Features of malnutrition: wasting, stunting, underweight, micronutrient deficiencies
- Abdomen: abdominal distension, +/- abdominal tenderness (may be diffuse/generalized)
- Signs of fat-soluble vitamin deficiencies, i.e. bruising, jaundice, abnormal skeletal exam, abnormal neurologic exam
- Genetic syndromes affecting pancreas may present with a variety of congenital abnormalities (such as nasal wing aplasia or hypoplasia, absence of permanent teeth, imperforate anus, congenital deafness, short stature, scalp defects)

Key Labs/Imaging:

- **Labs:**
  - CBC, electrolytes, liver enzymes, vitamin A/D-25/E, retinol-binding protein, INR, total cholesterol, albumin, HbA1C, magnesium, zinc, selenium, carotene, apolipoproteins
  - Fecal elastase-1: limited sensitivity in detecting mild to moderate EPI
  - Genetic tests to consider but not limited to: PRSS1, SPINK1, CFTR, CTRCSBDS
- **Imaging:**
  - MRCP with secretin: can provide anatomic and functional information of the pancreas. Can give qualitative assessment of intestinal fluid from pancreas stimulation.
- **Endoscopy:**
  - Endoscopic pancreatic function test (ePFT) may have increased sensitivity and specificity to detect isolated and generalized enzyme deficiencies, mild to moderate EPI

Management Considerations:

- Monitor growth and pubertal development along with nutrition status
- Serial monitoring of nutritional markers: vitamin A/D-25/E, INR, apolipoproteins, total cholesterol, magnesium, retinol-binding protein, calcium, zinc, selenium, and carotene
- Pancreas enzyme replacement therapy: General dosage: Infant: 2000-5000 lipase units/120ml of formula/breast milk. 1-4yr old: 1000-2000 lipase units/kg per meal. >4yr old: 500-2500 lipase units/kg per meal. Snack: half of the meal dose. Max 10,000 lipase units/kg/day.

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November 2024

- Evaluate for other associated abnormalities according to the underlying diagnosis

Additional Notes/References:
https://pubmed.ncbi.nlm.nih.gov/25915425/

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