CF Pancreatic Disease: Pancreatitis

Diagnosis:

- **Definition:** Inflammation, auto-digestion of the pancreas, and ductal obstruction, which results from impaired HCO_3^- secretion leading to viscous/acidic pancreatic secretion
  - **Associated with:**
    - **Less severe CFTR genotype -** less severe mutations lead to milder CF phenotype or increased CFTR function → increased acinar reserve/partial ductal obstruction
    - **Lower sweat chloride value**
- **Differential Diagnosis:** peptic ulcer disease, constipation, hepatobiliary disease/cholelithiasis

Key History to Review:

- *CFTR* genetics, sweat chloride, history of chronic abdominal pain (distinguishing between other causes of abdominal pain that may occur in CF), # known pancreatitis episodes, oily/greasy stools, autoimmune diseases, pancreatic ductal anatomy, meds (especially CFTR modulator therapy presence and timing)
- Family history of pancreatic diseases and cancer, cholecystectomy

Key Physical Exam Components:

- Abdomen: epigastric tenderness in older children (could be any location in younger children)
- Growth: weight loss/poor weight gain, growth failure
- Signs of fat-soluble vitamin deficiencies (bruising, abnormal skeletal or neuro exam)

Key Labs/Imaging:

- **Blood testing:** CBC, amylase, lipase, CMP, direct bilirubin, GGT, vitamin A/E/D-25, INR, IgG subclasses, OGTT (from age 10)
- **Genetic tests:** *CFTR* full sequencing (with deletions/duplications) if not already completed, testing for other pancreatitis risk variants (including but not limited to PRSS1, SPINK1, CTRC)
- **Stool:** fecal elastase for EPI assessment
- **Imaging:** MRCP with secretin - to look for any structural pancreatic changes and assess pancreatic function

Management Considerations:

- Monitor growth and symptoms/signs of malabsorption
- Annual diabetes mellitus screening with fasting glucose, HbA1c, +/- OGTT (if not already performed per standard CF care)
- Screen for exocrine pancreatic insufficiency (EPI) at time of CF diagnosis and if history of pancreatitis, especially >1 episode and/or symptoms of EPI and monitor annually. Treat with PERT as indicated
  - Check for fat soluble vitamin deficiencies if EPI is diagnosed
- Analgesia with initial use of non-opioids and weaker to stronger opioids if needed

Additional Notes/References:

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