Acute Recurrent Pancreatitis (ARP)

Diagnosis:

- **Definition:** ≥2 distinct episodes of acute pancreatitis (AP), as defined by complete resolution of pain for ⩾1 month between AP diagnosis or normalization of amylase and lipase between episodes
- **Differential Diagnosis:** Peptic ulcer, acute cholecystitis, acute hepatitis, cholangitis, bowel obstruction, renal colic

Key history to review:

- Prior episodes of pancreatitis, gallstones, IBD, celiac disease, genetic predispositions, known pancreatic anatomical abnormalities
- Relevant medication, infection history, family history of pancreatitis

Key Risk Factors:

- **Genetic**: include but not limited to PRSS1, CFTR, SPINK1, and CTRC mutations
- **Obstructive:** choledocholithiasis, biliary sludge, sphincter of Oddi dysfunction, pancreas divisum, pancreatico-biliary duct outflow obstruction
- **Toxic/Metabolic:** hypertriglyceridemia, hypercalcemia, organophosphates, low serum levels of antioxidants, medications
- **Others**: infections, autoimmune, vascular injury, vasculitis
- **Idiopathic:** 30% cases are idiopathic

Key Physical Exam Components:

- Eyes/Skin: possible scleral icterus/jaundice
- Abdomen: epigastric tenderness in older children (could be any location in younger children) with possible radiation to the neck or back. Younger children may have fussiness, vomiting
  - May also have positive Murphy's sign (right upper quadrant pain on deep inhalation during palpation) which may suggest concomitant cholecystitis

Key Labs/Imaging:

- **Labs:**
  - **Blood:** CBC, CMP, lipase, amylase, TTG IgA, IgA, IgG4, fat-soluble vitamins, fasting lipid panel, HgA1c.
  - **Stool:** fecal elastase (best to test at least 4 weeks post AP attack/resolution of symptoms)
  - **Skin:** sweat chloride test if CFTR mutations suspected
  - **Genetic**: genetic panel which should include but not limited to PRSS1, CFTR, SPINK1, CTRC
- **Imaging:**
  - Abdominal ultrasound, MRCP (with secretin if available), CT scan to assess pancreas parenchyma and ductal anatomy (MRCP best at looking at ductal anatomy)
    - If possible, wait at least 4 weeks post AP episode
- **Endoscopy**:
  - EUS (evaluate pancreatic anatomy if cross sectional imaging is inconclusive)
  - ERCP (treat obstructive etiologies)

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Management Considerations:

- Inpatient supportive measures
  - IV hydration & pain management (acetaminophen, NSAIDs, and if needed opioids)
    - May trial enteral nutrition within 24-48 hrs
  - Identify underlying risk factors/treatable causes
  - Please see additional management recommendations at AP flash card
- Outpatient supportive measures
  - Wean off pain medications
  - Screen for EPI at least annually and treat with PERT if clinically indicated
  - Monitor for endocrine pancreatic insufficiency/DM at least annually and refer to a pediatric endocrinologist if needed
  - Avoid controllable risk factors
  - Consider referral to a multi-disciplinary pancreas center
- Consider evaluation for Total Pancreatectomy with Islet Auto transplantation (TPIAT) to optimize pain relief, quality of life, and diabetes outcomes (independence of narcotics and insulin)

Additional Notes/References:

https://doi.org/10.1053/j.gastro.2018.12.043
https://doi.org/10.1097/MPG.0000000000001715
https://doi.org/10.1097/mpa.0000000000002008

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