QUESTIONS

PEDiatric ACUTE RECURRENT AND CHRONIC PANCREATITIS

1. All of the following may be used as part of criteria to make a diagnosis of Acute Pancreatitis in a child **EXCEPT:**
   - a. Epigastric pain x 2 days
   - b. Amylase level 3 times upper limits of normal
   - c. Lipase level 2 times upper limits of normal
   - d. Ultrasound demonstrating pancreatic and peri-pancreatic edema
   - e. Computer tomography scan demonstrating diffuse pancreatic swelling

2. A 5 year old child with seizure disorder presents with vomiting and abdominal tenderness. Lipase values are 6x upper limits of normal. Medications include valproic acid and melatonin. There is no history of trauma nor fever. Family history is unremarkable. The child is stable. Laboratory tests reveal normal CBC, liver enzymes, triglycerides, calcium values. Abdominal ultrasound reveals a mildly prominent pancreas without any signs of chronicity; the liver, gallbladder and biliary tree are all within normal limits. What would be your **next step?**
   - a. Discontinue the valproic acid
   - b. ERCP
   - c. Cholecystectomy
   - d. Sweat chloride
   - e. CT scan

3. All of the following genes have been implicated in ARP and/or CP **EXCEPT:**
   - a. CFTR
   - b. SPINK1
   - c. PRSS1
   - d. SBDS
   - e. CTRC

4. Which of the following would **NOT** generally be considered “baseline” or “first line” investigations in an 8 year old child presenting with first attack of acute pancreatitis, mild severity, without obvious etiology nor family history?
   - a. Liver enzyme panel
   - b. CFTR gene panel
   - c. Triglyceride level
   - d. Calcium 2+ level
   - e. Abdominal ultrasound
ANSWER KEY

1. (c) Lipase level 2 times upper limits of normal

The diagnosis of pediatric acute pancreatitis relies on fulfilling at least 2 of the following 3 criteria:

i. Abdominal pain compatible with pancreatic origin
ii. Amylase and/ or lipase at least 3 times upper limits of normal
iii. Pancreatic imaging suggestive of acute inflammation of pancreas.

A lipase level 2 times upper limits of normal would not fulfill the above criteria. All other options listed would.

2. (a) Discontinue the valproic acid

With a presentation of acute pancreatitis, one must review the presentation and particulars of a patient and compare them against common etiologies (including trauma, anatomic, biliary/ stones, medications/ toxic, infectious, metabolic, genetic, “idiopathic”). In the scenario provided, there is no indication of anatomic abnormalities nor biliary causes (ultrasound; liver enzymes), first-line metabolic workup is unremarkable (calcium 2+, triglycerides), there is no history of trauma nor any suggestion of infection (including lack of fever), and family history is unremarkable. The most likely culprit is the valproic acid, a medication that has been strongly associated / linked with acute pancreatitis (which can occur at any time, likely as an idiosyncratic complication not related to duration of therapy nor dose of medication). In this case, the recommendation would be to discontinue the valproic acid.

3. (d) SBDS gene

Cystic fibrosis transmembrane conductance regulator (CFTR), serine protease inhibitor Kazal type 1 (SPINK1), serum cationic trypsinogen (PRSS1), and chymotrypsin C (CTRC) have all been implicated in ARP and CP.

In contrast, the Shwachman-Bodian-Diamond Syndrome (SBDS) gene is implicated in Shwachman-Diamond syndrome, the second most frequent cause of pediatric pancreatic exocrine insufficiency. This gene has not been implicated in ARP / CP.

4. (b) CFTR gene panel

From the list of more “frequent” etiologies of AP and many for which a therapy can be instituted to prevent further attacks, anatomic (abdominal ultrasound), biliary/ stones (abdominal ultrasound, liver enzymes), metabolic (hypercalcemia, hypertriglyceridemia) baseline testing may be performed. Without high clinical suspicion/ family history, undertaking genetic testing would NOT be considered an “initial” investigation (it tends to be reserved for children who have demonstrated either ARP or CP, and even in these cases, genetic testing must be undertaken with the understanding that results may not necessarily
change clinical management, and ideally are undertaken with the involvement of a genetics team / genetic counselor).
1. A 12-year old girl presents to the ED with acute pancreatitis. After assessing her as having a moderately severe case of acute pancreatitis, you decide to admit her to GI service. The first 24 hours focus on fluid resuscitation, and she does well. Remembering a recent presentation you heard on medical and nutritional management of pancreatitis, you decide the recommended NEXT STEP in management is:
   a. Calling the PICC line team to place long-term venous access
   b. Starting the patient on peripheral parenteral nutrition
   c. Order the patient NPO status
   d. Place nasogastric tube in anticipation of enteral feeds

2. The best available evidence to support early enteral feedings in the setting of acute pancreatitis include all the following outcomes EXCEPT:
   a. Reduced incidence of infections
   b. Reduced multi-organ failure
   c. Reduced mortality
   d. All of the above have been shown

3. Your patient follows-up in the clinic after her discharge and wants to discuss how to prevent further episodes of pancreatitis. You recommend all the following as options EXCEPT:
   a. Antioxidants/micronutrients
   b. Low-fat diet
   c. Pancreatic enzymes
   d. Antibiotics

4. Potential complications of chronic, recurrent pancreatitis includes chronic pain syndrome. Patients presenting with chronic, recurrent pancreatitis and chronic pain need to be particularly screened for:
   a. Eating disorders
   b. Depression
   c. Obesity
   d. Learning disability

QUESTIONS
ENDOSCOPIC AND SURGICAL THERAPIES OF RECURRENT AND CHRONIC PANCREATITIS

1. A 14 year old female has had her second episode of acute pancreatitis. Her evaluation to date has shown a normal abdominal ultrasound, normal triglyceride and calcium, a negative sweat chloride test and no evidence for infectious, metabolic or drug induced pancreatitis. You have decided to perform further imaging as part of the evaluation. The best option in this setting would be:
   a. Repeat abdominal Ultrasound
   b. MRCP
   c. ERCP
   d. Endoscopic Ultrasound

2. The MRCP in the above mentioned patient is normal with a normal biliary tree and pancreatic duct. She has had another episode and is looking for further options. Repeat laboratory evaluation demonstrates no causes. Genetic testing has not shown a mutation in CFTR, SPINK1 or PRSS. In considering the further endoscopic options for her in this scenario, which procedure has the highest likelihood of finding a cause for her recurrent pancreatitis?
   a. ERCP without sphincter of Oddi manometer
   b. ERCP with sphincter of Oddi manometry
   c. Endoscopic US alone

3. ERCP with SOD manometry is performed and pancreatic ductal hypertension is found. Rank the endoscopic options from lowest success rate to the highest success rate in reducing episodes of pancreatitis
   a. Bile duct sphincterotomy alone
   b. Pancreatic duct stent alone
   c. Pancreatic duct sphincterotomy alone
   d. Dual duct sphincterotomy
4. A 12 year old boy with chronic pancreatitis due to PRSS mutation (hereditary pancreatitis) has had multiple episodes of pancreatitis and has been hospitalized 5 times in the last year. He has failed endoscopic treatments and is on chronic pain medications and has missed 25% of the school days in the last year. In considering surgical options of this boy, which statement is true.

   a. The Frey procedure (lateral pancreaticojejunostomy with extension to the head of the pancreas) leads to 50% of children having significant improvement in pain and eliminates the risk of pancreatic cancer

   b. Total pancreatectomy and islet cell autotransplantation will lead to pancreatic sufficiency and no need for insulin

   c. The Puestow procedure (lateral pancreaticojejunostomy) has a better success rate for pain relief than the Frey procedure

   d. All three surgical procedures offer a 75%-85% improvement rate. The choice of surgery will depend on the expertise of the surgeon and the long term goals of the family.
ANSWER KEY

1. B is the correct answer. MRCP is the best option in this scenario of the possible choices. MRCP guides the next steps and planning for the next procedures. The yield on repeat abdominal US would be low as this is generally used for ascertainment of biliary tract disease. ERCP can yield imaging, but the risk/benefit favors MRI for diagnosis. Endoscopic US could yield findings similar to MRCP, but requires procedural expertise and in more invasive.

2. B is the correct answer. ERCP alone in the setting of a normal MRCP is of minimal yield. Similarly Endoscopic US is of low yield. ERCP with SOD manometry appears to be the best option in this scenario. About 50% of children will have pancreatic ductal hypertension.

3. B, A, C, D

4. D is the correct answer. There is no significant difference in pain relief. For A: 85% improve, but the risk of cancer remains, B: Patients will be pancreatic insufficient for life C. If anything the Frey has a better pain relief outcome, but there is likely no difference.