Medical Management of Chronic Pancreatitis in Children

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TUMS

CP requires imaging findings characteristic of and consistent with CP (specifically, radiographically evident calcifications, and pancreatic duct irregularities, such as strictures and dilations) along with 1 of the following 3: abdominal pain consistent with pancreatic origin, evidence of exocrine pancreatic insufficiency (EPI), or evidence of endocrine pancreatic insufficiency, or a pancreatic biopsy specimen demonstrating histological evidence of CP

In recent years, studies have shown a relatively high rate of associated genetic mutations, with positive genetic testing in 64% to 70% of pediatric chronic pancreatitis cases.

Chronic ductal obstruction from anatomic abnormalities, medication induced pancreatitis, autoimmune pancreatitis, and inborn errors of metabolism should also be considered.

Currently, the known common mutations include the cationic trypsinogen gene, *PRSS1*, along with the cystic fibrosis transmembrane conductance regulator *(CFTR)* and serine peptidase inhibitor Kazal type 1 *(SPINK1)* mutations.[87] Other associated mutations include chymotrypsin C (caldecrin) gene *(CTRC)* and the calcium- sensing receptor gene *(CASR)*, among other associated genetic mutations.

Etiology of Chronic Pancreatitis

- Toxic-metabolic
  - Medications (see Box 82.1)
  - Hypercalcemia
  - Hyperlipidemia
  - Post–graft-versus-host disease
  - Toxins
  - Organic compounds
- Genetic
  - Autosomal dominant
  - *PRSS1* mutations
  - Autosomal recessive/modifiers
  - *CFTR* mutations
  - *SPINK1* mutations
  - *CPA1* mutations
  - *CTRC* mutations
- Autoimmune
  - Isolated autoimmune pancreatitis
  - Syndromic autoimmune pancreatitis
  - Inflammatory bowel disease associated
- Anatomic
  - See Box 82.1
- Severe acute pancreatitis
  - Postnecrotic
  - Vascular disease/ischemic
  - Postirradiation
- Indeterminate

Imaging

Dietary Considerations for Children With Chronic Pancreatitis

Use of Pancreatic Enzyme Replacement Therapy for Exocrine Pancreatic Insufficiency in Children With Chronic Pancreatitis

Pancreatogenic Diabetes Mellitus in Children With Chronic Pancreatitis

Medical Management of Pain in Chronic Pancreatitis

Sequelae of Chronic Pancreatitis

Imaging

In general terms, the roles of imaging in CP are to:
Contribute to/establish the initial diagnosis of CP;
Stage and monitor disease, including complications;
Assess for superimposed AP;
Identify potential etiologies of CP;
Identify findings that might herald endocrine or exocrine dysfunction;
Characterize secretory (exocrine) function;
Plan for intervention.

Although findings of CP may be identified on ultrasound or CT, MRI/magnetic resonance cholangiopancreatography (MRI/MRCP) is favored for the diagnosis and characterization of CP given its superiority in visualizing parenchymal and duct changes

Ultrasound examination

Ultrasound examinations are ideally performed fasting (4 hours) to reduce bowel gas that can obscure the pancreas and to distend the gallbladder. When the patient is able, drinking water immediately prior to the examination to distend the stomach with fluid and displace gastric air may provide an improved acoustic window for imaging the pancreas.

A complete ultrasound examination should assess pancreatic size, contour, echogenicity, pancreatic duct diameter (and for duct filling defects), and should assess for peripancreatic edema and pancreatic or peripancreatic fluid collections. In addition, the gallbladder should be assessed for calculi (an etiology of pancreatitis), and the biliary tree should be assessed for dilation and calculi.
Color Doppler along with gray-scale imaging can evaluate the peripancreatic vascular structures for complications, such as splenic vein or portal vein thrombosis and can assess vascularity of the pancreas.

Computed Tomography

Use of IV contrast material is recommended when performing CT for CP.
IV contrast material allows optimal assessment of the pancreatic parenchyma and allows evaluation of the peripancreatic vessels for patency.
For adults, multiphase protocols that include an unenhanced phase, parenchymal/arterial phase, and portal venous phase have been recommended.
No such recommendations exist for pediatrics but given the relative infrequency of calcifications in pediatric pancreatitis, an unenhanced phase is likely unnecessary.
As with AP, a parenchymal/arterial phase can be useful if clinical questions relate to the arteries but a single portal venous phase examination is generally sufficient to characterize CP in children.
CT with IV contrast material provides excellent assessment of the pancreatic parenchyma, allowing identification of features of ARP and CP, particularly calcifications and pancreatic atrophy.
CT can also identify congenital anomalies, such as annular pancreas and can assess for superimposed acute pancreatitis and complications of pancreatitis, including established vascular collaterals because of chronic/established thrombosis.
CT (and MRI) outperform ultrasound to define vascular anatomy relevant to surgical planning. CT is, however, limited by suboptimal visualization of the pancreatic and biliary ducts.

Magnetic Resonance Imaging/Magnetic Resonance Cholangiopancreatography

MRI and MRCP have the benefits of providing information on both parenchymal and duct changes of CP but are limited in their ability to visualize calcifications.

Adult and pediatric data suggest that the sensitivity of MRCP to detect pancreatic duct abnormalities may be improved by the administration of secretin.

Theoretically secretin distends the pancreatic duct and may allow for earlier detection of side branch-ectasias and provide information on exocrine function by quantifying duodenal filling

Chronic Pancreatitis Summary Statements and Recommendations

Initial Diagnosis

MRI is the recommended modality for imaging of suspected CP.

a. This recommendation reflects inadequate characterization of findings (particularly duct findings) of CP by transabdominal ultrasound. This also reflects the superior soft tissue contrast of MRI, which allows characterization of both parenchyma and duct findings of CP.

Assessment for Superimposed Acute Pancreatitis

When imaging is needed to assess a suspected or known episode of AP in a child with CP, transabdominal ultrasound is the preferred first-line imaging modality.

a. This recommendation reflects the availability and portability of ultrasound and the role of ultrasound in identifying biliary causes of AP.

b. Note: A negative ultrasound does not exclude AP (low to moderate sensitivity).

If ultrasound is negative for AP in a child with CP and an imaging diagnosis of AP is needed, either CT or MRI are recommended.

a. This recommendation reflects the only moderate sensitivity of ultrasound and the greater sensitivity of CT and MRI.

Intervention Planning

CT or MRI are recommended for planning of endoscopic or surgical interventions in a patient with known CP.

a. This recommendation is based on the large field of view afforded by CT and MRI, optimal characterization of the degree of organization of fluid collections, and optimal characterization of the peripancreatic vasculature.

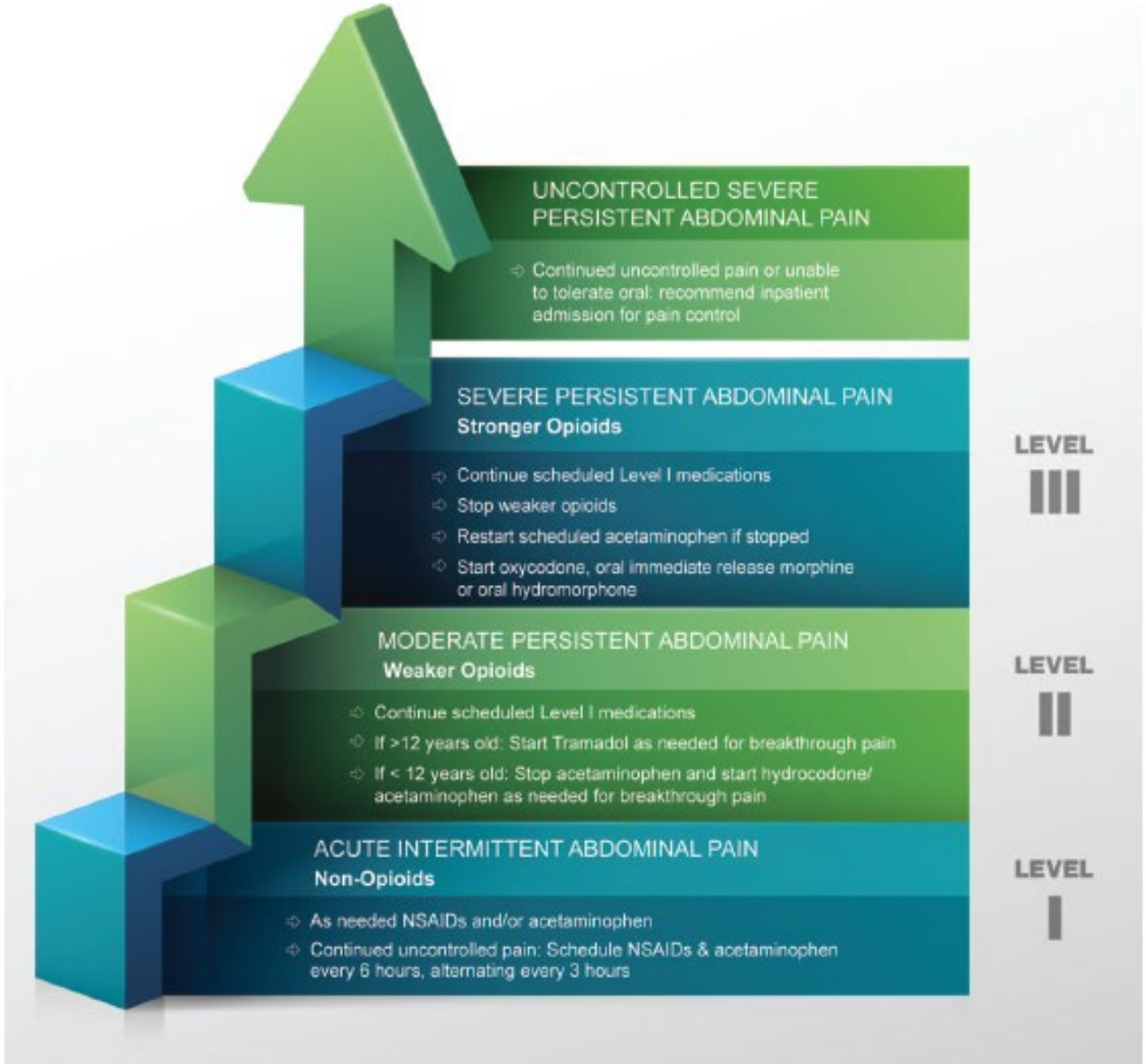
b. Note: When the intervention will target the duct, MRI is favored over CT.

Serial Monitoring

MRI is recommended for clinically indicated serial imaging of CP.

a. This recommendation reflects the optimal soft tissue contrast of MRI, no need for intravenous contrast material (in most cases), and the lack of associated ionizing radiation.

b. Note: In the child who requires sedation for MRI, risks of sedation must be balanced with the need for serial imaging.



Proposed analgesic ladder for the treatment of pain in children with chronic pancreatitis

EVALUATING EXOCRINE PANCREATIC INSUFFICIENCY (EPI) AND INITIATING PANCREATIC ENZYME REPLACEMENT THERAPY (PERT)