Pancreatic Disorders

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KEYWORDS

• Acute pancreatitis • Acute recurrent pancreatitis • Chronic pancreatitis
• Pancreatic insufficiency • Diabetes

KEY POINTS

- Once considered rare, pancreatic diseases, specifically acute, acute recurrent, and chronic pancreatitis, are increasingly recognized in children.
- Etiologies and risk factors of adult and pediatric pancreatitis are very different; therefore it is expected that their management, natural history, and response to therapy would also be different; however, studies on pediatric pancreatitis are limited.
- Genetic risk factors seem to play a role in the progression from acute recurrent to chronic pancreatitis; disease burden is high in chronic pancreatitis.
- Cystic fibrosis is the most common cause of exocrine pancreatic insufficiency in children; chronic pancreatitis and Shwachman Diamond syndrome are second most common.
- There is an urgent need for an exocrine pancreatic function that would be simple to perform, accurate, reliable, reproducible, and noninvasive.

Pediatric pancreatic diseases are increasingly recognized in childhood, possibly because of increased awareness among physicians.[1] Acute pancreatitis (AP) is estimated to occur at an incidence approaching that of adults. Although AP resolves without complications in most children, a subset continues to have recurrent attacks of pancreatitis (acute recurrent pancreatitis or ARP), and some progress to chronic pancreatitis (CP). In contrast to the adult population, most children with ARP or CP have genetic mutations; environmental risk factors are rare. Disease burden is significant in CP. Cystic fibrosis (CF) is the most common cause of exocrine pancreatic insufficiency (EPI) in childhood, followed by Shwachman-Diamond syndrome (SDS) and CP. Long-term effects of pancreatic diseases in children include possible nutritional deficiencies, pancreatogenic diabetes, and potentially pancreatic cancer later in life.

Disclosure Statement: None.
Funded by: NIH. Grant number(s): DK096327; DK097820; DK108334.
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Pediatr Clin N Am 64 (2017) 685–706
http://dx.doi.org/10.1016/j.pcl.2017.01.010 **pediatric.theclinics.com**

ACUTE PANCREATITIS

Risk Factors/Etiologies

Recent studies estimate the incidence of AP at between 3.6 and 13.2 cases per 100,000 children per year,[1] which is similar to incidences reported in adults.[2] **Box 1** lists etiologies of AP in children.

There are unique differences between risk factors of adult and pediatric AP.[3–17] In adults, alcohol use and gallstones account for the majority of cases, while etiologies in children are broad and variable. Biliary/obstructive factors, systemic illness, and medications are commonly identified in childhood AP; 15% to 30% cases are idiopathic. AP triggered by genetic mutations, metabolic factors, trauma, or alcohol is uncommon in children. In infants and toddlers, systemic illness is the leading cause.[3]

Pathophysiology

Pancreatitis may occur in the setting of an inciting factor (eg, medication, obstruction, genetic mutation) that triggers a cascade of events. There are several competing mechanisms of pancreatic inflammation including

- The traditional trypsin-dependent theory (activation of the enzymes leading to destruction of pancreas)[18]
- Inflammatory pathways (supported by animal models lacking trypsinogen and still developing inflammation)[19]
- Endoplasmic reticulum stress (independent of trypsin activation)[20]

Models that mimic human disease are needed to better dissect the mechanisms of pancreatic inflammation.

Clinical Manifestations

The most common symptoms of AP are abdominal pain and vomiting. Young children may present with vague symptoms and/or irritability; thus diagnosis in this age group requires a high degree of suspicion.[3] Signs and symptoms of cholangitis may be present in gallstone pancreatitis, but mild jaundice and liver enzyme elevations may occur in nonbiliary pancreatitis due to significant inflammatory changes in the distal bile duct as it traverses through the head of the pancreas.

Diagnosis

AP is a clinical diagnosis based on a combination of history, physical examination, laboratory testing, and imaging findings as listed in **Table 1**.[21]

Laboratory findings

Amylase and lipase are the most commonly used biochemical markers of pancreatic inflammation. Amylase and lipase elevations are not specific for AP, but lipase appears to be a more sensitive marker for pancreatitis. In the absence of a known etiology or family history, liver indices (aminotransferases, conjugated and unconjugated bilirubin and GGT), along with fasting glucose, triglycerides, and calcium are recommended laboratory studies for the first episode of AP.

Imaging findings

Imaging may be done to confirm AP and/or its complications, assessing pancreatic parenchyma and the surrounding organs and vasculature. Imaging may include transabdominal ultrasound (TUS), contrast-enhanced computed tomography (CECT), MRI of the abdomen including magnetic resonance cholangiopancreatography (MRCP), and endoscopic ultrasound (EUS).