Imaging makes advances in pancreatic diseases

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Ultrasound, CT, and MRI are less effective that believe when reviewing pediatric pancreatic disorders.

Many disorders of the pancreas in pediatric patients have a characteristic appearance on imaging. Reviewing these conditions on ultrasound, CT, and MRI should aid understanding of pathologic processes affecting the pancreas in this patient population.

This analysis is based on imaging findings from our own database of pediatric pancreatic pathology. We performed ultrasound in all cases and CT and/or MRI when ultrasound was inconclusive. MR cholangiopancreatography (MRCP) was used to study the pancreatic ducts. The pancreas is a nonencapsulated, multilobal gland that extends from the second portion of the duodenum to the splenic hilum.\(^1\) It forms embryologically from a ventral anlage that becomes the inferior pancreatic head and uncinate process, and a dorsal anlage that becomes the superior pancreatic head, body, and tail. The two anlagen fuse at seven weeks' gestation, and, in over 90% of cases, the ducts also fuse.\(^2\)

The size of the pancreas varies according to the child's age. The duct of Wirsung ends at the second portion of the duodenum at the major papilla, together with the common bile duct. An accessory pancreatic duct draining through the minor ampulla is present in 44% of individuals.\(^3\)

Ultrasound is the modality of choice for initial evaluation of suspected pancreatic disease in children,\(^2\) due to their lack of fat, the prominence of their left hepatic lobe, and the modality's lack of ionizing radiation or need for sedation. No special preparation is necessary other than six to eight hours of fasting (three hours for neonates).

The investigation is generally performed with a 5- or 7.5-MHz sector transducer, although a 3.5-MHz sector transducer can be used in older children. Transverse and longitudinal scanning of the entire pancreas is performed. Each portion of the organ should be measured and its echogenicity compared with that of the liver. A systematic scan of the entire superior abdomen should also be performed (Figure 1A).

Pancreatic MRI is indicated in cases of suboptimal or equivocal ultrasound findings with a high clinical suspicion of pathology.\(^3\)\(^4\) Specific indications include evaluation of acute and chronic pancreatitis and characterization of complex peripancreatic fluid collections. Complete evaluation of pancreatic disease generally requires fat-suppressed T1-weighted sequences before and after contrast, as well as T2-weighted sequences, but the choice of sequences will depend on the scanner. MRCP is a noninvasive technique for imaging the pancreaticobiliary tract.\(^2\) The pancreas has a higher T1 signal intensity on fat-suppressed sequences than any other intra-abdominal organ. Signal intensity is higher than that of the liver on the arterial-capillary phase of gadolinium-enhanced images and is similar to liver on delayed images. T2 signal intensity of the normal pancreas varies; it can be isointense to liver or as high as abdominal fat (Figure 1B).

CT is not recommended as the primary modality for pediatric pancreatic imaging, but it is useful when ultrasound findings are nondiagnostic and MRI is not available.\(^5\) CT may be used for diagnostic and therapeutic procedures such as aspiration biopsy or drainage. Patients undergoing CT should receive adequate intravenous contrast and oral contrast when possible. We vary the slice thickness, pitch, and reconstruction interval according to patient age, keeping dose to a minimum. CT attenuation of the pancreas is normally similar to that of the liver.

CONGENITAL ANOMALIES

True epithelial cysts of the pancreas are uncommon and are caused by anomalous development of the pancreatic ducts.\(^1\) The cysts may appear alone or in association with systemic diseases such as Von Hippel-Lindau syndrome or polycystic kidney disease.\(^6\)\(^7\) When a large number of epithelial cysts are present, distinguishing their origin can be difficult. The differential diagnosis includes mesenteric cysts, choledochal cysts, and enteric duplication cysts.\(^6\)\(^8\)

Ultrasound shows a well-defined, thick-walled anechoic mass with posterior acoustic enhancement.\(^1\) CT reveals a mass with well-defined walls and central low attenuation.\(^2\) MRI shows a mass with
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smooth walls of waterlike signal intensity on T1- and T2-weighted images. MRCP is useful to rule out communication with the pancreatic duct or biliary tree. Definitive diagnosis can be obtained following pathologic study of the specimen.

Pancreas divisum, the most common congenital anomaly of the pancreas, represents an incomplete fusion of the dorsal and ventral ductal structures, in which a longer dorsal pancreatic duct drains through the smaller minor papilla. The condition may be diagnosed on CT when the pancreatic head is enlarged in the absence of a visible pancreatic mass or when a fatty cleft is seen within the pancreas. When MRCP shows the unconnected ventral and dorsal pancreatic ducts entering the duodenum, it confirms the altered ductal anatomy.

Annular pancreas, the second most common congenital anomaly of the pancreas, is characterized by pancreatic tissue completely or incompletely surrounding the descending duodenum. Patients with complete annular pancreas present during the neonatal stage with symptomatic bowel obstruction and a specific "double-bubble" sign on plain-film radiography. Those with incomplete or partial annular pancreas may not present until adulthood. CT or MRI may show thickening of the anterior, lateral, and posterior aspect of the descending duodenum caused by tissue signal intensity and enhancement characteristics that are identical to those of pancreatic parenchyma.

Von Hippel Lindau syndrome is an autosomal dominant disease that results in retinal angiomas, cerebellar hemangioblastoma, and cysts to various organs. Cysts are typically small and multiple. Clustering of multiple cysts makes it difficult to distinguish them from microcystic adenomas. The presence of pancreatic cysts in a patient with a familial history of Von Hippel-Lindau syndrome is considered diagnostic. Ultrasound, CT, and MRI observations are similar to those for congenital pancreatic cysts, pancreatic cysts in patients with polycystic kidney disease, and pancreatic cystosis. Definitive diagnosis is made from clinical history and other imaging findings.

Shwachman-Diamond syndrome is a rare autosomal recessive disorder that usually manifests in infancy. It is characterized by exocrine pancreatic insufficiency that leads to malabsorption (with normal results on the sweat test), short stature, and bone marrow dysfunction. The characteristic pathologic finding is fatty infiltration of the pancreas, acini reduction, and conservation of the islets. Ultrasound depicts a hypechoic pancreas that is unchanged in size, aiding its differentiation from cystic fibrosis, in which the pancreas is smaller. CT shows total fatty replacement of the pancreas, ductal ectasia, and calcifications. The fatty infiltrated pancreas has a similar or higher signal intensity than the remainder of the pancreas on T1-weighted MRI.

Nesidioblastosis represents the persistence of the normal fetal state of the pancreas, characterized by diffuse proliferation and persistence of nesidioblasts. Patients may become hypoglycemic due to the fetal pancreatic cells' abnormal insulin secretion; nesidioblastosis is the most frequent and severe cause of hypoglycemia in newborns and infants.

Ultrasound, CT, and MRI do not demonstrate pathologic changes in most cases, although imaging may show a nonspecific increase in the size of the pancreatic head, body, or tail (Figure 2). Near-total (95%) pancreatectomy is recommended to avoid repeat operations. A remnant of pancreatic tissue (mainly the head) is left to protect the common bile duct. Pancreatic regeneration without recurrent hypoglycemia after near-total pancreatectomy can occur, and ultrasound demonstrates a normal pancreas in such cases. Partial regeneration of pancreatic tissue may also occur.

Cystic fibrosis is the most significant autosomal recessive pancreatic disorder among whites. It is characterized by dysfunctional chloride ion transport across epithelial surfaces. Although pulmonary disease is the predominant cause of morbidity and death, diagnosis in children is usually made following gastrointestinal symptoms. Cysts, which are typically small, most likely occur secondary to duct obstruction by inspissated secretions. Ultrasound shows an increase in echogenicity and a decrease in pancreatic size. A fine lobular (cobblestone-like) echo pattern is typically seen in the pancreas and small or clustered calcifications can be observed in any part of the pancreatic gland.

CT demonstrates complete replacement of the pancreas by fat as an increase in pancreatic size with low attenuation values. MR reveals an enlarged pancreas with high signal intensity on T1-weighted images. Complete pancreatic atrophy without fatty replacement may also be visible, in which case the pancreas appears decreased in size with soft-tissue attenuation but without fat attenuation or high signal intensity. Areas of fibrosis exhibit low attenuation values with no postcontrast enhancement on CT and low signal intensity on both T1- and T2-weighted MR images. Complete pancreatic fibrosis and calcifications within the pancreas may also be seen.

Collections of cysts up to several centimeters in diameter may, rarely, replace the pancreas altogether, causing a mass effect within the abdomen. This condition is known as pancreatic cystosis.
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Pancreatic tumors, including lymphoma and rhabdomyosarcoma, can be primary or metastatic. Nonepithelial tumors, which are relatively rare, arise from cells in the pancreatic islets of Langerhans.

Microcystic adenoma (serous cystoadenoma) is the second most common cystic neoplasm of the pancreas. These benign tumors are seen with increased frequency in patients with Von Hippel-Lindau disease.

In chronic pancreatitis, the pancreas generally becomes atrophic in chronic pancreatitis. If focal or diffuse enlargement is seen in such cases, the differential diagnosis should include malignancy.

Chronic pancreatitis is seen as diminished signal intensity on T1-weighted MRI. Immediate postcontrast and delayed enhancement images show decreased heterogeneous enhancement and gland atrophy. MRCP reveals dilation and irregular contours in the duct.

Pseudocysts are the most common local complication of acute pancreatitis and the most common type of pancreatic cyst. Ultrasound, CT, and MRI can visualize this complete replacement of the pancreas by uni- or multiloculated cystic masses.

Ultrasound shows multiple sonolucent, rounded, well-defined, fluid-filled lesions extending from the porta hepatitis to the splenic hilum and hyperechoic pancreatic tissue scattered among cysts. Most of the lesions demonstrate a homogeneous water-density content on CT, and less frequently, a slightly higher attenuation value. Small calcifications may be seen, and identifiable pancreatic tissue disappears when macrocysts completely replace the pancreas. T1-weighted MRI reveals homogeneous low signal intensity, which is significantly lower than that of pancreatic tissue. The cysts have very high signal intensity on T2-weighted MRI. Cyst walls appear as thin, regular lines of low signal intensity.

**PANCREATITIS**

Acute pancreatitis is a mainly diffuse, inflammatory process. The extent of involvement of the pancreatic gland can vary widely. Pediatric cases of acute pancreatitis are uncommon; those that do occur are usually caused by trauma. The most common ultrasound finding is diffuse or focal glandular enlargement and decreased echogenicity, with poorly defined borders. In some cases, the pancreas appears normal. CT shows diffuse pancreatic enlargement, heterogeneous attenuation, a poorly defined pancreatic contour, and peripancreatic fluid in the anterior pararenal space and lesser sac (Figure 4). The attenuation pattern is caused by a striking decrease (or total lack) of enhancement caused by ischemic and necrotic zones. T1-weighted MRI of acute pancreatitis reveals focal or diffuse enlargement and peripancreatic inflammatory changes, while T2-weighted MRI demonstrates peripancreatic edema and fluid collections. The main pancreatic duct retains a smooth contour on MRCP but may be compressed by surrounding edematous parenchyma.

The lesions are usually homogeneous with signal intensity comparable to that of water on T1- and T2-weighted MRI. MRCP shows communication with the pancreatic duct or with other adjacent organs. Although pancreatic abscesses can appear similar to pseudocysts, they may be distinguished by clinical history or gas within the collection.

Chronic pancreatitis is an inflammatory process of the pancreas with irreversible exocrine and endocrine dysfunction, most often caused by hereditary pancreatitis in children. Ultrasound shows parenchymal atrophy with focal pancreatic enlargement, increased echogenicity, calcifications, ductal dilation, and irregular pancreatic outline. Calcium deposits, visualized as an echogenic focus that casts an acoustic shadow, and pseudocysts are sometimes seen as well. CT demonstrates calcifications (clustered and/or scattered), focal or diffuse parenchymal atrophy, and dilation of the main pancreatic and biliary ducts. The pancreas generally becomes atrophic in chronic pancreatitis. If focal or diffuse enlargement is seen in such cases, the differential diagnosis should include malignancy.

Chronic pancreatitis is seen as diminished signal intensity on T1-weighted MRI. Immediate postcontrast and delayed enhancement images show decreased heterogeneous enhancement and gland atrophy. MRCP reveals dilation and irregular contours in the duct.

**NEOPLASMS, METASTASES**

Malignant and benign pancreatic neoplasms are extremely rare in pediatric patients. Pancreatic tumors are classified as epithelial or nonepithelial in origin. Epithelial pancreatic tumors may originate from endocrine or nonendocrine tissue and can be malignant or benign. Nonepithelial pancreatic tumors, including lymphoma and rhabdomyosarcoma, can be primary or metastatic. Mucinous cystic neoplasm, the most common pancreatic cystic neoplasm, occurs most often in middle-aged women and rarely in children. This tumor usually consists of a large multilocular cyst containing mucinous material or hemorrhagic fluid and should be distinguished from a pseudocyst. Microcystic adenoma (serous cystoadenoma) is the second most common cystic neoplasm of the pancreas. These benign tumors are seen with increased frequency in patients with Von Hippel-Lindau disease and are usually located in the pancreatic head. Microcystic adenomas are generally composed of numerous small cysts separated by fibrous septae that radiate from the center. Neuroendocrine tumors, which are relatively rare, arise from cells in the pancreatic islets of Langerhans. The five types are insulomas (the most common), gastrinomas, glucagonomas, somatostatinomas, and tumors that secrete vasoactive-intestinal peptide. All tend to occur in
middle-aged adults rather than children. Approximately 50% of neuroendocrine tumors have no hormonal function. Both functioning and nonfunctioning neuroendocrine tumors are characterized by their hypervascularity. Nonfunctioning islet cell tumors are usually large and sometimes contain calcifications.\textsuperscript{10} They may develop a cystic appearance secondary to degeneration and necrosis. Functioning islet cell tumors tend to be small, well-defined, and either round or oval.

Pancreatoblastoma predominantly affects young children. It is a rare tumor in general but is the most common childhood pancreatic neoplasm.\textsuperscript{7,10} These tumors tend to be large and solitary and can occur in any region of the pancreas. Ultrasound shows solid masses of mixed echogenicity with regions of cystic change or calcification. CT reveals heterogeneous enhancement with or without calcified foci. Tumors have a low to intermediate signal on T1-weighted MRI and high signal intensity on T2-weighted MRI.\textsuperscript{7,10}

Pancreatic metastases are more common than primary neoplasms. Non-Hodgkin's lymphoma, the most frequently seen tumoral pancreatic disease in children, involves the pancreas secondarily in about 30% of patients with widespread disease.\textsuperscript{1,20} Malignancy usually spreads to the pancreas by direct extension from peripancreatic lymphadenopathy. Primary involvement of the pancreas is uncommon, affecting 2% to 5% of patients with extranodal lymphomas. These metastases usually develop late in the course of disease and are often accompanied by concurrent extrapancreatic metastases.\textsuperscript{20}

Metastases appear on ultrasound as solitary or multiple lesions. They are visualized as solid, hypoechoic, well-defined masses, or as diffuse glandular infiltration. The pancreas itself appears larger and has decreased echogenicity.\textsuperscript{1} CT shows three patterns: a large, well-defined ovoid or round localized mass that is either isodense or hypodense (50% to 73% of cases), diffuse pancreatic enlargement with a smooth or lobulated contour (Figure 5), and multiple pancreatic nodules.\textsuperscript{20}

**ADDITIONAL FINDINGS**

A number of factors can cause fatty infiltration of the pancreas, including long-term treatment with corticosteroids or cytostatics, parenteral nutrition, Cushing's disease, and obesity.\textsuperscript{1,5,21} Pancreatic lipid in fatty infiltration is confined to the interstitial stroma, does not involve the exocrine or endocrine parenchymal cells, and usually has few clinical consequences.\textsuperscript{21}

Distribution of fatty infiltration is variable, but the pancreatic body and tail are the dominant areas of fatty replacement. Ultrasound shows a hyperechoic pancreas with no change in size.\textsuperscript{1} CT shows low-attenuation tissue interspersed between normal pancreatic parenchyma.\textsuperscript{5} MRI shows higher signal intensity in the fatty infiltrated pancreas than in the remainder of the pancreas, but this signal intensity is lost on opposed-phase T1-weighted gradient-echo MRI.\textsuperscript{3,21}

**REFERENCES**


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