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Techniques in Vascular and Interventional Radiology

Pediatric Biliary Interventions

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An interventional radiologist is frequently called to evaluate and treat biliary diseases in children; a tailored approach specific to this population is required. Imaging with an emphasis on minimizing ionizing radiation is used not only in the initial workup but also to guide interventions. The most common form of intervention generally consists of transhepatic biliary drainage to treat either biliary obstruction or bile leakage, a scenario frequently encountered after pediatric liver transplantation. Other pathologies referred for evaluation and management include biliary atresia and, rarely, symptomatic choledochal cysts. Biliary complications caused by an underlying malignancy are not a frequently encountered problem in the pediatric population. The initial evaluation, role of preprocedural imaging, and interventional management with an emphasis on technique are discussed regarding these common biliary pathologies in children.
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Introduction

Clinical Evaluation of Pediatric Patients

As with adult patients with symptoms of cholestasis or obstruction, initial evaluation of pediatric patients begins with obtaining a comprehensive clinical history, physical examination, and review of laboratory results including liver function tests and coagulation factors. If no prior imaging is available for review, ultrasound (US) should always be the first-line imaging modality in infants and children. US is advantageous because it is readily available at most institutions and does not require sedation to obtain an adequate evaluation. Additionally, one of the clear benefits of US is that it does not expose the child to ionizing radiation, which is a very important factor when treating pediatric patients. Children are more radiosensitive than adults are, and both the "Image Gently" and "Step Lightly" campaigns have made great strides in raising awareness and educating physicians regarding this (www.ImageGently.org). It follows that there is an extremely limited indication for computed tomography (CT) scanning in a child with suspected biliary disease, especially as secondary imaging in an infant or child can and should be performed with magnetic resonance (MR) imaging with the addition of MR cholangiopancreatography (MRCP) sequences for the evaluation of the biliary tree. If CT is absolutely necessary, child-sized protocols that minimize radiation dose (such as low kVp, low mAs, use of radiation exposure–limiting filters, and new reconstruction algorithms) should be used.

Indications for Percutaneous Biliary Procedures in Children

Indications for biliary procedures in children vary based on the age of the child. In neonates, hyperbilirubinemia is very common but is only rarely associated with severe disease. Hyperbilirubinemia can be broadly categorized as decreased bilirubin conjugation, bilirubin overproduction, or problems with bilirubin excretion. Problems with biliary excretion may cause elevation in the levels of conjugated bile salts, termed "direct" hyperbilirubinemia. In a premature infant, this direct cholestasis often is because of prolonged total parenteral nutrition. In full-term infants, neonatal hepatitis and biliary atresia are the diagnoses that must be considered. This is often when the radiologist becomes involved in the patient's evaluation, with US, MR imaging, and percutaneous cholecystocholangiography performed for exclusion of biliary atresia. Many patients with biliary atresia progress over time to severe liver disease and require liver transplantation. These transplants are at risk for biliary complications including bile

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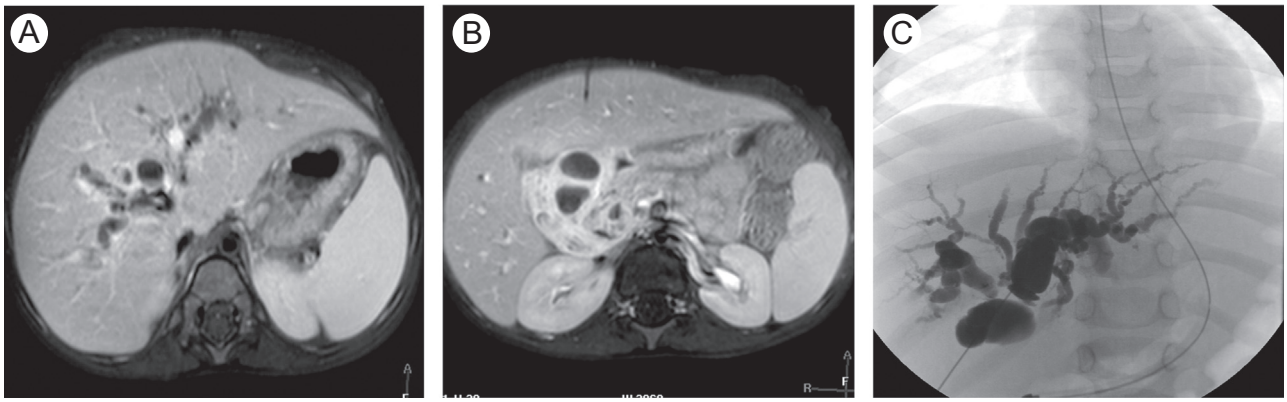

Figure 1 Biliary rhabdomyosarcoma. (A and B) MR postgadolinium T1 axial images demonstrating ductal dilatation and large solid and cystic enhancing mass at porta hepatis. (C) PTC demonstrating the obstructing mass lesion.

duct strictures or leaks necessitating percutaneous transhepatic cholangiography (PTC) for diagnosis and treatment. Other conditions such as choledochal cysts, sclerosing cholangitis, autoimmune pancreatitis, and rare malignant causes of obstruction such as rhabdomyosarcoma may also be indications for biliary procedures in older children (Fig. 1).

Biliary Atresia

Biliary atresia is a progressive obliterative cholangiopathy, which, if left untreated, leads to biliary cirrhosis. As such, biliary atresia must be identified or excluded as early as possible when imaging an infant with jaundice. Prompt treatment with surgical diversion of bile flow with hepatoportoenterostomy (Kasai procedure) leads to a better long-term outcome. As stated previously, the initial imaging evaluation of an infant with suspected biliary atresia is with a US study of the abdomen. US is useful for assessing not only the hepatobiliary tree but also other anomalies that may be present, including heterotaxy syndromes. Features seen on US that suggest a diagnosis of biliary atresia include an absent or irregular gallbladder and the triangular cord sign of increased echogenicity just above the porta hepatis. A normal gallbladder may be seen in up to 10% of patients with biliary atresia; however, a small gallbladder with a length typically <1.9 cm is more common.[1] Other imaging options include nuclear scintigraphy, MR cholangiography (MRC), and endoscopic retrograde cholangiopancreatography (ERCP), all of which have limitations. MRC requires high-quality imaging and usually also requires general anesthesia and controlled breath-holds because of the small size of the normal ductal system in infants. Visualization of the extrahepatic bile ducts can exclude the diagnosis of biliary atresia with an accuracy rate of 82% and a sensitivity rate of 90%.[2] ERCP may be used for the diagnosis of biliary atresia; however, this technique also requires anesthesia and is often difficult to perform in small infants. Biopsy and direct cholecystocholangiography are often needed to make the final diagnosis. These procedures can be performed intraoperatively by a surgeon, with plans for immediate surgical intervention based on the findings, or percutaneously in the interventional suite under US and fluoroscopic guidance.

Percutaneous Cholecystocholangiography Technique

This procedure can only be performed in infants with an identifiable gallbladder lumen on US. The patient's clinical status and laboratory studies must be reviewed before the procedure, especially concerning platelet count and coagulation profile. Intravenous antibiotic pretreatment is recommended, with coverage for gram-negative organisms (usually ampicillin at a dose of 50 mg/kg and gentamicin at a dose of 2 mg/kg). Most procedures in infants are performed under general anesthesia or sedation provided by a qualified anesthesiologist. Under sterile conditions, a Chiba needle measuring 22-25 gauge is directed under real-time US guidance into the gallbladder lumen. With a very small needle, the bile may not readily flow or be aspirated into the needle hub after removal of the stylet; therefore, real-time guidance and observation of the needle within the lumen are important. In the absence of conjugation of the bile salts, the fluid may be clear rather than golden. Contrast material is then slowly injected into the lumen under fluoroscopic visualization. To reduce radiation exposure, optimal collimation, low pulse rate fluoroscopy (usually 3 frame/s), and image electronic magnification (instead of true image intensifier magnification, which significantly increases the radiation dose) are recommended (Fig. 2). If there is free flow of contrast agent to the duodenum, the patient may need to be repositioned to opacify the intrahepatic ducts, with Trendelenburg positioning usually sufficient to allow intrahepatic duct opacification. Last image hold is usually sufficient for documentation of the procedure, especially if the bile ducts fully opacify and are normal. In cases of abnormal examinations or poor visualization, a single spot film may suffice. Once images have been reviewed, the residual contrast material in the gallbladder is aspirated to decompress the gallbladder, and the needle is removed. Pressure is held at the puncture site for 5-10 minutes, preferably with US guidance to direct pressure to the appropriate site.