Pediatric Biliary Interventions in the Native Liver

Lisa H. Kang, MD¹ Colin N. Brown, MD²

¹ Department of Radiology, University of Texas Southwestern Children's Health, Dallas, Texas

² Department of Radiology, Texas Scottish Rite Hospital for Children, Dallas, Texas

Address for correspondence Lisa H. Kang, MD, Department of Radiology, Children's Health, 1935 Medical District Drive, Mail Code F1.02, Dallas, TX (e-mail: lisa.kang@utsouthwestern.edu).

Semin Intervent Radiol 2016;33:313–323

Abstract

Keywords

- pediatric biliary interventions
- bile ducts
- interventional radiology
- pediatric

Biliary disease in infants and children frequently presents diagnostic and therapeutic challenges. Pediatric interventional radiologists are often involved in the multidisciplinary teams who care for these patients. This article reviews several notable causes of biliary disease in children who have not undergone liver transplantation, describes the role of percutaneous interventional procedures in managing these conditions, and details applicable biliary interventional techniques.

Objectives: Upon completion of this article, the reader will be able to provide an overview of several important causes of biliary disease which affect pediatric patients, to explain the role of interventional radiology procedures in the management of these conditions, and to describe the interventional techniques used.

Accreditation: This activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint providership of Tufts University School of Medicine (TUSM) and Thieme Medical Publishers, New York. TUSM is accredited by the ACCME to provide continuing medical education for physicians.

Credit: Tufts University School of Medicine designates this journal-based CME activity for a maximum of **1 AMA PRA Category 1 Credit™**. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

The diagnosis and treatment of native liver biliary disease in infants and children are often challenging and best approached with input from multidisciplinary pediatric specialists including gastroenterologists, surgeons, and radiologists.¹

Over time, the role of interventional radiology (IR) procedures in the diagnosis and treatment of biliary disease has evolved. Magnetic resonance cholangiopancreatography (MRCP) allows noninvasive visualization of the biliary system

and has largely replaced percutaneous transhepatic cholangiography (PTC) for diagnostic purposes.² With increased experience of pediatric gastroenterologists and availability of specialized small endoscopes, endoscopic cholangiopancreatography (ERCP) has become the first-line procedure for most pediatric biliary interventions.^{3,4} Despite these shifts in practice, procedures performed by IRs such as percutaneous cholecystocholangiography (PCC), PTC, biliary drainage, and percutaneous cholecystostomy (PC) remain valuable techniques.

This article provides an overview of several causes of biliary disease which are of particular interest in the non-transplant pediatric population, describes the role of procedures performed by IRs in their diagnosis and treatment, and discusses technical considerations of common procedures.

Pediatric Biliary Diseases

Biliary Atresia

Neonatal jaundice is common and most often due to transient unconjugated hyperbilirubinemia. When jaundice persists beyond 2 weeks of age, evaluation for cholestatic causes is required, as cholestatic jaundice is often associated with serious hepatobiliary disease which requires prompt evaluation and treatment.⁵ In cholestatic jaundice, there is impaired excretion of conjugated bilirubin, resulting in conjugated hyperbilirubinemia.⁵ Etiologies of cholestatic jaundice are

numerous and can be broadly categorized as obstructive or hepatocellular. Obstructive causes include biliary atresia (BA), choledochal cysts, and bile plug syndrome. Hepatocellular causes, sometimes collectively referred to as neonatal hepatitis, include infection, genetic diseases such as α -1-antitrypsin deficiency and Alagille syndrome, and parenteral nutrition-associated cholestasis.⁶

BA is one of the most commonly identified causes of neonatal cholestasis, accounting for approximately 25% of cases. BA is an obliterative cholangiopathy which variably affects both the intrahepatic and extrahepatic bile ducts. The etiology of BA is poorly understood and it is likely the common endpoint of several underlying processes. Without treatment, infants with BA develop cirrhosis and typically die by 2 years of age.⁷

BA is classified according to the level of proximal obstruction (►Fig. 1). In type I disease, obstruction is located at the level of the common bile duct (CBD), which may be dilated, and a bile-filled gallbladder (GB) is present. In type II disease, obstruction is at the level of the common hepatic duct; the CBD, cystic duct, and GB remain patent in subtype IIa and are obliterated in subtype IIb. In type III BA, obstruction is above the level of the common hepatic duct, with only microscopic biliary ductules present at the porta hepatis and obliteration of the extrahepatic biliary tree. Types IIb and III account for approximately 90% of cases.⁸

Kasai portoenterostomy (KP), in which a jejunal loop is anastomosed to the porta hepatis to allow enteric drainage of bile through remaining microscopic biliary ductules, is the preferred treatment for most cases of BA. KP allows clearance

of jaundice and prolongation of native liver survival. However, it is chiefly a palliative procedure and approximately 80% of KP patients will require liver transplantation before reaching adulthood. Primary liver transplantation is used in a minority of patients.⁷

Liver injury in BA is progressive. Although the influence of patient age at surgery varies among studies, long-term surgical outcomes of KP may be worse when it is performed in patients older than 60 to 100 days.^{9–12} Thus, timely diagnosis of BA is important.

In patients with possible BA, the goal of evaluation is to expeditiously and confidently confirm or exclude the diagnosis while minimizing morbidity related to diagnostic procedures. However, current noninvasive tests have limitations, and establishing a diagnosis of BA can be challenging. Laparotomy with intraoperative cholangiogram remains the gold standard for diagnosing or excluding BA. When BA is confirmed, the surgeon can proceed to perform KP.¹⁰

Fasting right upper quadrant sonography is the initial imaging modality used to evaluate infants with cholestatic jaundice. Several sonographic findings supportive of a diagnosis of BA have been described, including the “triangular cord” sign (visualization of an echogenic triangular ductal remnant in the porta hepatis),^{13,14} the “gallbladder ghost triad” (atretic GB measuring less than 1.9 cm in length, indistinct GB wall with thinned or irregular/incomplete echogenic mucosa, and irregular wall contour),¹³ absence of the GB,¹⁰ and absence of the CBD.¹⁴ Sonography is reported to have accuracy of up to 97 to 98% in distinguishing between infants who have BA and those who do not, when performed

Fig. 1 Classification of biliary atresia.

by experienced operators at specialized referral centers. However, less experienced operators may have difficulty achieving the same results.¹⁴

Hepatobiliary scintigraphy, performed using Tc99m-labeled iminodiacetic acid derivatives with early and delayed (24-hour) imaging, is commonly used when BA is suspected. In BA, there is no biliary-to-bowel transit of radiotracer on early or delayed images.¹⁵ Scintigraphy has nearly 100% sensitivity for BA—radiotracer activity in bowel essentially excludes the diagnosis. However, the specificity is lower (~70%) and false positives can occur with severe hepatocellular dysfunction. Thus, a lack of activity in the bowel supports, but does confirm a diagnosis of BA.¹⁶ Premedication with phenobarbital is reported to increase the accuracy of scintigraphic evaluation for BA.¹⁷

MRCP offers the potential to visualize the intra- and extrahepatic biliary system, although experience with MRCP for the diagnosis of BA is less than that of US and scintigraphy. Published series report high sensitivity, with visualization of a normal and complete biliary system providing strong evidence that BA is not present. However, specificity is limited and incomplete visualization of the biliary system on MRCP does not confirm a diagnosis of BA.^{18,19}

When the clinical picture is unclear and less invasive tests have failed to confirm or exclude a diagnosis of BA with adequate certainty, IRs are often consulted to perform additional diagnostic tests. Standard PTC is rarely attempted in infants with suspected BA.^{20,21} However, when a sonographically identifiable GB is present, PCC can be performed. In PCC, a needle is inserted percutaneously into the GB using sonographic guidance and iodinated contrast material is injected (→ Fig. 2).²² When contrast opacifies a complete biliary tree, BA is excluded. Failure to visualize a complete biliary tree is supportive of BA and an indication for operative cholangiogram.

In a series of 22 patients, 7 of whom were eventually diagnosed with BA, PCC was successfully performed in 18 (5 with BA) and was unsuccessful in 4 (2 with BA) due to an inability to identify or puncture the GB.²³ In a different series, the authors attempted PCC only in 9 patients who were deemed to have an “adequate” GB lumen, out of 22 evaluated with sonography. The procedure was able to exclude BA in all these patients.²⁴ In four studies, in which a combined total of 39 infants underwent PCC, no procedural complications occurred.^{21–24} In summary, PCC, although not feasible in all patients, appears safe and has the potential to either definitively exclude BA or provide strong evidence that operative cholangiogram is needed.¹⁰

Biliary Complications Following Kasai Portoenterostomy

Patients with BA who are treated with KP are at ongoing risk of biliary complications. Recurrent cholangitis is the most common complication, and may lead to deterioration of liver function.²⁵ Intrahepatic cyst formation and areas of cystic biliary ductal dilation occur as both early and late complications of KP and may be associated with infection and jaundice. PTC with aspiration or drain placement can aid in the management of these complications.^{26,27} ERCP is generally

Fig. 2 (a) Operative cholecystocholangiogram in an infant with biliary atresia. Contrast fills a small gallbladder (arrow), but the remainder of the biliary system is not visualized. (b) Cholecystocholangiogram in an infant with cholestasis and abnormal scintigraphy (not shown). Contrast fills normal biliary system, excluding biliary atresia.

not technically feasible in patients who have undergone KP due to surgical alteration of the upper gastrointestinal tract (GI) tract anatomy.²⁸ Obstruction of the roux loop, which provides enteric drainage of bile following KP, can occur secondary to kinking, adhesions, and luminal stenosis.²⁵ Percutaneous puncture of the obstructed roux loop can be used to determine the site of obstruction, to place a biliary drainage catheter, and potentially to perform balloon dilation of a stricture.²⁹

Choledochal Cysts

Choledochal cysts (CCs) are uncommon lesions characterized by cystic dilation affecting one or more segments of the intrahepatic and/or extrahepatic bile ducts.^{30,31} Anomalous junction of the pancreatic and biliary ducts, which allows

reflux of pancreatic secretions into the biliary system, appears to be an important factor in the pathogenesis of CCs.³⁰ Although CC may present at any age from the prenatal period to adulthood, many are diagnosed in the first decade of life. Clinical manifestations are variable and include the classic clinical triad of abdominal pain with jaundice and a right upper quadrant mass, bile peritonitis from cyst rupture, and neonatal cholestasis.^{30–33} Cholangiocarcinoma is a feared complication of CC, although this is much more common in adults.³⁴

CCs are categorized under the Todani classification (►Fig. 3). Type I CCs, which are confined to the extrahepatic biliary tree, are the most common (80–90%) and include three subtypes. Subtype IA is cystic dilation of the extrahepatic bile ducts which communicates with the cystic duct. Subtype IB is segmental extrahepatic dilation, usually of the distal CBD. Subtype IC is diffuse fusiform dilation of the CBD and common hepatic duct. A type II CC (2%) is an extrahepatic bile duct diverticulum. A type III CC (1–5%) is a choledochocoele of the intraduodenal segment of the CBD. Type IV CCs (15–20%) are multiple cystic dilations of the intrahepatic and extrahepatic bile ducts (subtype IVA) or of the extrahepatic ducts only (subtype IVB). Type V disease, also known as Caroli disease and often considered a distinct entity, is cystic dilation of the intrahepatic bile ducts.^{1,30,34}

Sonography is the initial imaging test of choice for diagnosing CC, and typically demonstrates an anechoic cystic or fusiform structure in the porta hepatis which is distinct from the GB.³⁰ Notably, in type I BA, cystic dilation of the CBD can be confused with CC.³⁴ Scintigraphic findings in CC are variable, although an area of photopenia in the porta hepatis which gradually accumulates radiotracer is suggestive. Activ-

ity in bowel may or may not be seen depending on whether or not obstruction is present.³⁰ MRCP has largely supplanted PTC and ERCP for detailed diagnostic evaluation of CCs due to its noninvasive nature and ability to accurately demonstrate the anatomy of the lesion.³⁴

Owing to an association with progressive hepatic fibrosis and the risk of malignancy, definitive treatment of CC is surgical excision and biliary reconstruction. Surgical technique is dictated by the anatomy of the lesion.^{34,35} However, both PTC and ERCP have an adjunctive role in the management of complicated CCs. Indications for cyst drainage using these techniques include cholangitis which fails to respond adequately to antibiotics and decompression of large cysts to facilitate surgery.³⁶ Cyst rupture is generally treated with a two-stage surgical approach (T-tube drainage followed by delayed resection). However, there are reports of successful temporization of ruptured CCs with PTC and catheter drainage (►Fig. 4).^{33,36} When cyst drainage is required as a temporizing measure in young children, ERCP is preferred over PTC, as an internal catheter may be more stable and better tolerated.³⁷

Bile Plug Syndrome and Inspissated Bile Syndrome

Bile plug syndrome and inspissated bile syndrome are closely related conditions in which biliary obstruction is caused by bile plugs or inspissated bile in the absence of abnormal biliary anatomy. These conditions are uncommon causes of cholestasis in infants.^{38,39} Predisposing factors include cystic fibrosis, parenteral nutrition, hemolysis, and conditions producing hepatocellular injury such as CMV infection.³⁸

On US, typical findings include biliary ductal dilation with echogenic material within the bile ducts and sometimes in

Fig. 3 Todani classification of choledochal cysts.

Fig. 4 (a) Coronal contrast-enhanced CT image in a 3-year-old girl presenting with peritonitis from ruptured type I choledochal cyst. Note the small defect in medial wall of the cyst (arrow) and ascites (asterisk). (b) Fluoroscopic image with percutaneous transhepatic cholangiography via direct cyst puncture prior to temporary cyst drainage. Peritoneal drain is present.

the GB lumen.¹ Depending on the degree of obstruction, hepatobiliary scintigraphy may show absent or reduced radiotracer activity within the bowel, which can lead to confusion with BA.⁴⁰ If PTC is performed, it will reveal biliary ductal dilation with ductal filling defects and/or obstruction.^{40,41}

Although in some cases obstruction may resolve spontaneously or with oral ursodeoxycholic acid, surgery has been the mainstay of treatment.^{38,40–42} There is a report of successful percutaneous management of cholestatic jaundice believed to be caused by bile plug syndrome in a 5-month-old. In this case, a cholecystostomy catheter was placed for biliary drainage with subsequent balloon dilation of the distal CBD.³⁸

Acute Acalculous Cholecystitis

Acute acalculous cholecystitis (AAC) is defined as acute inflammation of the GB in the absence of gallstones.⁴³ AAC in children can be grouped into two major categories. The first is AAC which occurs in the setting of severe illness, such as leukemia or major trauma. In this group, GB inflammation is thought to be related to bile stasis and ischemia and there is a significant risk of GB perforation and mortality.⁴³ The second group is AAC which occurs in otherwise healthy children with viral infection (e.g., hepatitis A and Epstein-Barr virus), bacterial infection (e.g., *Salmonella typhi* and *Leptospira* species), or noninfectious systemic disease (e.g., Kawasaki disease). The prognosis in the second group is much better.^{44–46}

Sonography is the imaging modality most commonly used to diagnose AAC; typical findings are GB wall thickening (> 3 mm), GB distension, GB sludge, pericholecystic fluid, and an absence of stones (►Fig. 5). Observation of sonographic Murphy sign increases specificity, although this often cannot be assessed in patients who are intubated and sedated.^{47,48} CT and MRI demonstrate findings similar to those seen on sonography.⁴⁹ AAC usually produces nonvisualization of the GB when scintigraphy is performed, although adult studies report a false-negative rate of approximately 30%.^{49,50}

In patients with serious systemic illness, AAC has traditionally been treated with cholecystectomy. However, based on more recent experience in adults and children, PC appears to be the treatment of choice in patients who are at high risk of surgical morbidity and mortality (►Fig. 6).^{47,51} In otherwise healthy children who develop AAC due to conditions such as

Fig. 5 Right upper quadrant abdominal sonographic image in a 9-year-old girl with acute acalculous cholecystitis. Note the gallbladder distention and wall thickening (arrow).

Fig. 6 Fluoroscopic image obtained following percutaneous cholecystostomy in a 1-year-old child with acute acalculous cholecystitis. Note the cystic duct patency (arrow), which is present in some cases of acalculous cholecystitis.

viral infection, medical therapy is adequate in most patients, although cholecystostomy or cholecystectomy may be required in some.^{44–46}

In some patients with AAC treated with PC, cholecystectomy is performed at a later date when the patient's overall condition has improved. In others, GB function recovers and cholecystectomy is not necessary.⁴⁷ Complications of PC include catheter malpositioning and dislodgement, bleeding, bile leak and bile peritonitis, and inadvertent puncture of adjacent structures including the colon.⁵² In a series of 10 children with AAC and immune compromise, PC was technically successful in all patients and there were no procedural complications, although 4 patients later died due to underlying illness. Of the six surviving patients, three did not undergo interval cholecystectomy.⁴⁷

Malignant Biliary Obstruction

Obstructive jaundice can be a presenting feature or complication of abdominal malignancies in children including biliary rhabdomyosarcoma (BR), neuroblastoma, and non-Hodgkin lymphoma (NHL).^{53–55} BR is the most common cause of malignant biliary obstruction (MBO) in pediatric patients.⁵³ MBO can lead to pruritus, cholangitis, impaired liver function, and exacerbated toxicity of certain chemotherapy drugs.^{53,56}

In patients with MBO, sonography reveals biliary ductal dilation upstream from the site of obstruction and is frequently able to identify the obstructing mass.⁵⁷ BR typically manifests as an intraluminal mass within the large extrahepatic ducts. On CT, it has variable morphology, attenuation,

and enhancement. On MRI, BR is usually T1-hypointense and prominently T2-hyperintense. It may have a botryoid (i.e., bunch of grapes) appearance. Notably, tumors with a large cystic component can be mistaken for CC on cross-sectional imaging.⁵⁴ Cholangiography, whether accomplished by MRCP, PTC, or ERCP, usually demonstrates an intraluminal mass in cases of BR and extrinsic compression at the site of obstruction in other tumors.

In contrast to MBO in adults, which is usually caused by tumors which are poorly responsive to chemotherapy and have a poor prognosis, tumors causing MBO in children are often responsive to therapy. Thus, the need for drainage is more often temporary.⁵⁸ Additionally, in some cases of pediatric MBO, biliary drainage procedures are neither necessary nor advisable. This is particularly true for NHL, which usually responds rapidly to medical therapy (►Fig. 7).^{59–61}

However, interventional techniques remain valuable temporizing and palliating measures for MBO. Transhepatic

Fig. 7 (a) Axial T1-weighted MRI image in a 10-year-old boy with a non-Hodgkin lymphoma pancreatic head mass (arrow) presenting with jaundice. (b) Coronal maximum intensity projection magnetic resonance cholangiopancreatography with diffuse biliary ductal dilation to the level of the pancreatic head (arrow). Patient was treated with chemotherapy and jaundice resolved without drainage.

percutaneous biliary drainage can be accomplished with an external drain (ED) or an external–internal drain (EID) which extends through the CBD into the duodenum. The latter is preferred due to better catheter stability and restoration of physiologic biliary flow to the bowel. ED can be used if the obstruction cannot be crossed with a wire and can be later converted to EID in many cases.⁵⁸ More than one drainage catheter may be required if portions of the intrahepatic bile ducts are isolated by tumor.⁵⁸ PC is an alternate route of drainage, but can only be used if the intrahepatic ducts, common hepatic duct, and cystic duct remain patent.⁵⁸

Complications of BD include catheter dislodgement, bacteremia, bleeding into the biliary system or peritoneal cavity, pancreatitis, inadvertent puncture of adjacent structures including the colon and diaphragm, and duodenal perforation.^{53,58,62} Additionally, chemotherapy may impede formation of a well-defined tract at the site of ED, leading to bile leak and peritonitis following catheter removal.⁵⁹ Two small series (six and eight patients) reported high technical success rates of percutaneous biliary drainage (100 and 87.5%, respectively) in children with MBO. Of the 14 patients in these series, 2 experienced nonfatal procedural complications (hemoperitoneum managed with transfusion and conservatively managed duodenal perforation).^{53,58}

Periprocedural Considerations

ERCP versus Percutaneous Procedures

ERCP, usually performed by pediatric gastroenterologists, is an invasive test in which the ampulla of Vater is cannulated via an endoscope, allowing injection of contrast material as well as insertion of various instruments for diagnostic and therapeutic purposes. ERCP is generally considered the first-line approach for pediatric biliary interventions.^{3,4}

However, there are several notable limitations of ERCP. In up to 10% of patients, cannulation of the ampulla is unsuccessful.^{63–65} Additionally, ERCP may be difficult or impossible in patients who have surgically altered upper GI tract anatomy, such as patients who have undergone KP or certain bariatric procedures.²⁸ Finally, ERCP in infants requires a specialized small side-viewing endoscope, which is not available at all institutions.²²

Thus, the decision of whether to perform biliary interventions via ERCP or percutaneously is dependent both on patient factors and on the availability of qualified practitioners and appropriate equipment. In some complex cases, percutaneous and endoscopic approaches are combined, with an IR and a gastroenterologist collaborating to perform a “rendezvous procedure.”³⁷

Periprocedural Considerations for Percutaneous Biliary Interventions in Children

Indications for percutaneous biliary procedures in children are described in detail in the preceding sections. In general, PCC is performed for diagnostic purposes in infants with suspected BA, PTC is performed for diagnostic purposes and

as a route of access for other interventions, biliary drainage procedures are performed for decompression of an obstructed biliary system, and PC is performed in some cases of AAC.

Contraindications include uncorrected coagulopathy and lack of a safe access route.¹ Biliary interventions have a significant bleeding risk and can lead to bleeding which is challenging to detect and control. The Society of Interventional Radiology Standards of Practice Committee has published recommendations for the management of coagulopathy for these procedures. INR should be corrected if greater than 1.5, usually with transfusion of fresh frozen plasma. Platelets should be transfused for thrombocytopenia with counts less than 50,000/ μ L. Anticoagulation and antiplatelet agents should be withheld prior to the planned procedure.⁶⁶

Although data for the use of prophylactic antibiotics for hepatobiliary procedures in pediatric patients are sparse, prophylaxis recommendations can be made based on surgical data as well as adult data.^{67–69} In infants, intravenous ampicillin (50 mg/kg) and gentamicin (2 mg/kg) can be used for prophylaxis.^{37,70,71} In older children, adult guidelines can be followed, albeit with adjustment of antibiotic dose based on the patient's weight as needed.

In accordance with the principle of ALARA (“as low as reasonably achievable”), radiation dose in pediatric interventional procedures should be kept to the minimum required. When possible, sonography should be considered as an alternative to fluoroscopy. In many cases, adequate visualization of needle, wire, and catheter position can be obtained using sonography. When fluoroscopy is used, dose reduction measures should be employed. These include using intermittent pulsed fluoroscopy and collimation, using magnification only when necessary, and documenting with the last image hold feature.⁷²

At most children's hospitals, sedation and anesthesia are administered by a dedicated pediatric sedation or pediatric anesthesiology team. Maintaining patient warmth is often managed by these personnel. However, IRs should be mindful that keeping the patient dry is a significant factor in preventing heat loss, particularly in neonates who are less able to regulate their body temperature.⁷³

Tracking the patient's fluid balance during interventional procedures is especially important in neonates and small infants. Intravascular volume in neonates is approximately 85 to 96 mL/kg; shifts of 10% can cause fluid overload or hypovolemia. Contrast used should be iso-osmolar and diluted 1:1 with sterile normal saline whenever possible. In neonates and small children, an attempt to limit intravascular contrast dose to less than 3 mL/kg should be made, with a maximum of 10 mL/kg.⁷³ Although contrast administered for biliary procedures ideally remains within the biliary system, some contrast may be intravascularly injected or absorbed. Therefore, adhering to volume limits for intravascular injection is prudent.

Chlorhexidine gluconate/isopropyl alcohol is the preferred skin antiseptic for neonates, as iodine from povidone iodine preparations can be absorbed through the skin, affecting thyroid function.⁷³

Common Percutaneous Biliary Procedures

Percutaneous Cholecystocholangiography

The GB is identified with survey sonography using a curved, 6-MHz probe and a suitable route for puncture is planned. In most patients, a transhepatic route is preferred over a transperitoneal route to decrease the risk of peritoneal bile leak. Puncturing the GB with the needle parallel to the long axis of the GB has been suggested.²³ Under continuous sonographic visualization with either a curved or linear probe, a 22- to 25-gauge needle is advanced percutaneously into the GB lumen.

Under fluoroscopy, a small amount of dilute contrast is injected through the needle to confirm placement within the GB. Once correct positioning is confirmed, more dilute contrast is gently injected, producing opacification of the biliary system. To facilitate contrast filling of intrahepatic bile ducts, the patient can be placed in Trendelenburg position or intravenous morphine can be given to cause sphincter of Oddi spasm.^{24,73}

Once diagnostic images are obtained, residual contrast material is aspirated through the needle prior to needle removal. To aid hemostasis, 5 to 10 minutes of manual pressure is applied to the puncture site. Post-PCC, sonography is then performed to evaluate for hematoma or fluid collection suggestive of bile leak. If indicated, ultrasound-guided percutaneous liver biopsy can be performed during the same session.^{24,73}

Percutaneous Transhepatic Cholangiography and Drain Placement

After survey sonography of the liver, a 21- or 22-gauge needle is used to access the biliary system, preferably via a peripheral duct, under continuous US guidance. If spontaneous return of bile is not seen, an attempt at aspiration is made prior to contrast injection. If the needle tip is suspected to be within a bile duct, a small amount of dilute contrast is injected through the needle under fluoroscopy for confirmation. When difficulty accessing a bile duct is encountered because of non-dilation, PCC can be performed to delineate the intrahepatic biliary system.¹ However, PCC will be unsuccessful if there is blockage between the GB and intrahepatic bile ducts.

Once desired needle tip position within the biliary system is confirmed, more dilute contrast can be injected to obtain a diagnostic cholangiogram. However, if drain placement is planned, cholangiogram may be deferred until after drain placement to avoid inducing bacteremia.

For EID placement, a 0.018-in. guidewire is placed through the needle and a coaxial introducer system is then used to place a 0.035-in. guidewire. An angled catheter can then be used to guide the wire through the biliary system and into the duodenum. Positioning is confirmed as needed with injection of a small amount of dilute contrast through the angled catheter. The 0.035-in. working wire can then be used for EID placement. Drain placement is facilitated by dilation of the bile duct entry site.

If access to the duodenum is not possible due to obstruction or other factors, ED can be achieved by placing a small 1-cm

diameter pigtail, such as a Dawson-Mueller catheter (Cook, Bloomington, IN), at the most distal accessible position within the biliary system. Additional attempts at accessing the duodenum and conversion to an EID can be made at a later time, typically after 1 week.⁷⁴

In infants and small children, standard biliary drains with pre-formed side holes may be too long. In such cases, a locking pigtail catheter such as a Dawson-Mueller or Mac-Loc multipurpose drainage catheter (Cook) can be used with extra side holes fashioned manually.^{1,37} A 6- to 8.5F catheter is usually placed. After 48 hours to gravity drainage via a bag, an EID catheter may be capped if the patient is afebrile and liver function tests have normalized.⁷⁴

Initial reassessment of biliary drains is performed 4 weeks after placement. Bile duct patency is evaluated by removing the existing catheter over a guidewire, advancing a sheath sized 1F smaller than the drain catheter over the wire until the tip is at the site of bile duct entry, and injecting dilute contrast. Duct patency is confirmed if the contrast drains into the small bowel within 3 to 5 minutes.^{75,76} To evaluate tract maturation, the sheath is retracted to the skin entry site and contrast is injected gently. If there is leakage of contrast into the peritoneal cavity, the tract is not yet mature.⁷⁷

If occlusion remains or the tract is not mature, the drain is replaced. The drain is exchanged and reevaluated every 1 to 3 months until both duct patency and the tract maturity have been demonstrated. Before placing the new biliary drain, cholangioplasty and/or drain upsizing can be considered if there is persistent obstruction.⁷⁴ Metallic stenting is rarely used in children but may be considered for palliative purposes when MBO is not expected to improve with treatment.⁵³

Percutaneous Cholecystostomy

After sonographic survey, a transhepatic or transperitoneal approach is chosen. Although an individual patient's anatomy may dictate the safest route, a transhepatic route is preferred with ascites while a transperitoneal route is preferred with coagulopathy.⁷⁷ There are two techniques which can be used to access the GB- the Seldinger and trocar techniques.

Using the Seldinger technique, a 21- or 22-gauge needle is advanced into the GB under continuous sonographic guidance, avoiding puncture through the back wall of the GB. After the GB is accessed, needle tip position is confirmed with injection of a small amount of dilute contrast. A 0.018-in. guidewire is placed through the needle and a coaxial introducer system is used to place a 0.035-in. guidewire. Dilation over the 0.035-in. guidewire then facilitates drain placement. An 8.5F locking pigtail catheter such as a Dawson-Mueller or Mac-Loc multipurpose drainage catheter (Cook) is placed.

The trocar technique is an alternative to the Seldinger technique which can be performed in the IR suite or at the bedside. This method is performed by placing a sharp stylet into a locking pigtail catheter and advancing this coaxial unit into the GB under continuous sonographic guidance. A sharp jab may be required to penetrate the anterior GB wall. Once

the device tip is in the GB lumen, the catheter is advanced and the stylet is removed. The catheter pigtail is then formed in the GB and locked. Positioning is confirmed with sonography and/or dilute contrast injection.⁷⁷

Evaluation of cystic duct and CBD patency and of drain tract maturity is performed at 2 to 4 weeks, in a similar fashion to that used for other biliary drains. Allowing at least 3 weeks for tract maturation is suggested when a transperitoneal route is used.⁷⁸ If the ducts are not patent and/or the tract is immature, the drain is exchanged and reevaluated in 4 to 6 weeks. Alternatively, interval cholecystectomy may be performed.

Conclusion

Procedures performed by IRs remain important in the frequently challenging management of pediatric biliary disease. Pediatric IRs should be aware of the conditions which cause biliary disease in infants and children and should be familiar with the various techniques used in their diagnosis and treatment.

References

- Rozel C, Garel L, Rypens F, et al. Imaging of biliary disorders in children. *Pediatr Radiol* 2011;41(2):208–220
- Fitoz S, Erden A, Boruban S. Magnetic resonance cholangiopancreatography of biliary system abnormalities in children. *Clin Imaging* 2007;31(2):93–101
- Racadio JM, Kukreja K. Pediatric biliary interventions. *Tech Vasc Interv Radiol* 2010;13(4):244–249
- Rocca R, Castellino F, Daperno M, et al. Therapeutic ERCP in paediatric patients. *Dig Liver Dis* 2005;37(5):357–362
- Moyer V, Freese DK, Whittington PF, et al; North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. Guideline for the evaluation of cholestatic jaundice in infants: recommendations of the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. *J Pediatr Gastroenterol Nutr* 2004;39(2):115–128
- Gottesman LE, Del Vecchio MT, Aronoff SC. Etiologies of conjugated hyperbilirubinemia in infancy: a systematic review of 1692 subjects. *BMC Pediatr* 2015;15:192
- Hartley JL, Davenport M, Kelly DA. Biliary atresia. *Lancet* 2009;374(9702):1704–1713
- Wildhaber BE. Biliary atresia: 50 years after the first Kasai. *ISRN Surg* 2012;2012:132089
- Davenport M, Caponcelli E, Livesey E, Hadzic N, Howard E. Surgical outcome in biliary atresia: etiology affects the influence of age at surgery. *Ann Surg* 2008;247(4):694–698
- Jancelewicz T, Barmherzig R, Chung CT, et al. A screening algorithm for the efficient exclusion of biliary atresia in infants with cholestatic jaundice. *J Pediatr Surg* 2015;50(3):363–370
- Chardot C, Carton M, Spire-Bendelac N, Le Pommelet C, Golmard JL, Auvert B. Prognosis of biliary atresia in the era of liver transplantation: French national study from 1986 to 1996. *Hepatology* 1999;30(3):606–611
- Schoen BT, Lee H, Sullivan K, Ricketts RR. The Kasai portoenterostomy: when is it too late? *J Pediatr Surg* 2001;36(1):97–99
- Tan Kendrick AP, Phua KB, Ooi BC, Tan CE. Biliary atresia: making the diagnosis by the gallbladder ghost triad. *Pediatr Radiol* 2003;33(5):311–315
- Humphrey TM, Stringer MD. Biliary atresia: US diagnosis. *Radiology* 2007;244(3):845–851
- Ziessman HA. Hepatobiliary scintigraphy in 2014. *J Nucl Med Technol* 2014;42(4):249–259
- Kianifar HR, Tehranian S, Shojaei P, et al. Accuracy of hepatobiliary scintigraphy for differentiation of neonatal hepatitis from biliary atresia: systematic review and meta-analysis of the literature. *Pediatr Radiol* 2013;43(8):905–919
- Majd M, Reba RC, Altman RP. Effect of phenobarbital on 99mTc-IDA scintigraphy in the evaluation of neonatal jaundice. *Semin Nucl Med* 1981;11(3):194–204
- Norton KI, Glass RB, Kogan D, Lee JS, Emre S, Shneider BL. MR cholangiography in the evaluation of neonatal cholestasis: initial results. *Radiology* 2002;222(3):687–691
- Liu B, Cai J, Xu Y, et al. Three-dimensional magnetic resonance cholangiopancreatography for the diagnosis of biliary atresia in infants and neonates. *PLoS ONE* 2014;9(2):e88268
- Hashimoto T, Yura J. Percutaneous transhepatic cholangiography (PTC) in biliary atresia with special reference to the structure of the intrahepatic bile ducts. *J Pediatr Surg* 1981;16(1):22–25
- Treem WR, Grant EE, Barth KH, Kremers PW. Ultrasound guided percutaneous cholecystocholangiography for early differentiation of cholestatic liver disease in infants. *J Pediatr Gastroenterol Nutr* 1988;7(3):347–352
- Meyers RL, Book LS, O'Gorman MA, et al. Percutaneous cholecystocholangiography in the diagnosis of obstructive jaundice in infants. *J Pediatr Surg* 2004;39(1):16–18
- Lee SY, Kim GC, Choe BH, et al. Efficacy of US-guided percutaneous cholecystocholangiography for the early exclusion and type determination of biliary atresia. *Radiology* 2011;261(3):916–922
- Nwomeh BC, Caniano DA, Hogan M. Definitive exclusion of biliary atresia in infants with cholestatic jaundice: the role of percutaneous cholecysto-cholangiography. *Pediatr Surg Int* 2007;23(9):845–849
- Houben C, Phelan S, Davenport M. Late-presenting cholangitis and Roux loop obstruction after Kasai portoenterostomy for biliary atresia. *J Pediatr Surg* 2006;41(6):1159–1164
- Takahashi A, Tsuchida Y, Suzuki N, et al. Incidence of intrahepatic biliary cysts in biliary atresia after hepatic portoenterostomy and associated histopathologic findings in the liver and porta hepatis at diagnosis. *J Pediatr Surg* 1999;34(9):1364–1368
- Tsuchida Y, Honna T, Kawarasaki H. Cystic dilatation of the intrahepatic biliary system in biliary atresia after hepatic portoenterostomy. *J Pediatr Surg* 1994;29(5):630–634
- Orman ES, Miller CB, Grimm IS, Barritt AS IV. Single-balloon enteroscopy-assisted endoscopic retrograde cholangiopancreatography for treatment of cholangitis in a patient with a Kasai portoenterostomy. *J Pediatr Surg* 2012;47(5):E1–E5
- Levy J, Martin EC, DeFelice A, Abramson S, Altman RP, Laffey KJ. Obstruction of the Roux limb after portoenterostomy for biliary atresia: a delayed complication. *J Pediatr Surg* 1990;25(12):1264–1265
- Kim OH, Chung HJ, Choi BG. Imaging of the choledochal cyst. *Radiographics* 1995;15(1):69–88
- Soares KC, Kim Y, Spolverato G, et al. Presentation and clinical outcomes of choledochal cysts in children and adults: a multi-institutional analysis. *JAMA Surg* 2015;150(6):577–584
- De Bruyne R, Van Biervliet S, Vande Velde S, Van Winckel M. Clinical practice: neonatal cholestasis. *Eur J Pediatr* 2011;170(3):279–284
- Kang CM, Lee KH, Kim DH, Lee WJ. Percutaneous transhepatic cyst drainage as a “bridge procedure” to definitive treatment of perforated choledochal cysts: a case report. *Surg Laparosc Endosc Percutan Tech* 2008;18(6):598–600
- Soares KC, Arnaoutakis DJ, Kamel I, et al. Choledochal cysts: presentation, clinical differentiation, and management. *J Am Coll Surg* 2014;219(6):1167–1180

- 35 Lee SC, Kim HY, Jung SE, Park KW, Kim WK. Is excision of a choledochal cyst in the neonatal period necessary? *J Pediatr Surg* 2006;41(12):1984–1986
- 36 Lal R, Agarwal S, Shivhare R, et al. Management of complicated choledochal cysts. *Dig Surg* 2007;24(6):456–462
- 37 Atchie B, Kalva S, Josephs S. Pediatric biliary interventions. *Tech Vasc Interv Radiol* 2015;18(4):276–284
- 38 Duman L, Büyükyavuz BI, Akcam M, Koroglu M, Tepeli H. Percutaneous management of bile-plug syndrome: a case report. *J Pediatr Surg* 2011;46(12):e37–e41
- 39 Bernstein J, Braylan R, Brough AJ. Bile-plug syndrome: a correctable cause of obstructive jaundice in infants. *Pediatrics* 1969;43(2):273–276
- 40 Heaton ND, Davenport M, Howard ER. Intraluminal biliary obstruction. *Arch Dis Child* 1991;66(12):1395–1398
- 41 Lang EV, Pinckney LE. Spontaneous resolution of bile-plug syndrome. *AJR Am J Roentgenol* 1991;156(6):1225–1226
- 42 Lieber J, Piersma FE, Sturm E, Schäfer JF, Fuchs J, Szavay PO. Surgical treatment of inspissated bile syndrome using a 2-stage pure laparoscopic approach: a case report. *J Pediatr Surg* 2012;47(12):e47–e50
- 43 Poddighe D, Tresoldi M, Licari A, Marseglia GL. Acalculous acute cholecystitis in previously healthy children: general overview and analysis of pediatric infectious cases. *Int J Hepatol* 2015;2015:459608
- 44 Kim A, Yang HR, Moon JS, Chang JY, Ko JS. Epstein-Barr virus infection with acute acalculous cholecystitis. *Pediatr Gastroenterol Hepatol Nutr* 2014;17(1):57–60
- 45 Chen CJ, Huang FC, Tiao MM, et al. Sonographic gallbladder abnormality is associated with intravenous immunoglobulin resistance in Kawasaki disease. *ScientificWorldJournal* 2012;2012:485758
- 46 Verma M, Chhatwal J, Deodhar MC, Aneja R, Singh T. Enteric cholecystitis 15 years experience. *Indian Pediatr* 1995;32(7):808–810
- 47 Schaefer CM, Towbin RB, Aria DJ, Kaye RD. Safety and effectiveness of percutaneous cholecystostomy in critically ill children who are immune compromised. *Pediatr Radiol* 2016;46(7):1040–1045
- 48 Khoury NJ, Kanj V, Abboud M, Muwakkit S, Birjawi GA, Haddad MC. Abdominal complications of chemotherapy in pediatric malignancies: imaging findings. *Clin Imaging* 2009;33(4):253–260
- 49 Tulchinsky M, Colletti PM, Allen TW. Hepatobiliary scintigraphy in acute cholecystitis. *Semin Nucl Med* 2012;42(2):84–100
- 50 Mariat G, Mahul P, Prév t N, et al. Contribution of ultrasonography and cholescintigraphy to the diagnosis of acute acalculous cholecystitis in intensive care unit patients. *Intensive Care Med* 2000;26(11):1658–1663
- 51 Simorov A, Ranade A, Parcels J, et al. Emergent cholecystostomy is superior to open cholecystectomy in extremely ill patients with acalculous cholecystitis: a large multicenter outcome study. *Am J Surg* 2013;206(6):935–940, discussion 940–941
- 52 Garber SJ, Mathieson JR, Cooperberg PL, MacFarlane JK. Percutaneous cholecystostomy: safety of the transperitoneal route. *J Vasc Interv Radiol* 1994;5(2):295–298
- 53 Akinci D, Gumus B, Ozkan OS, et al. Percutaneous management of tumoral biliary obstruction in children. *Pediatr Radiol* 2007;37(10):975–980
- 54 Chung EM, Lattin GE Jr, Cube R, et al. From the archives of the AFIP: pediatric liver masses: radiologic-pathologic correlation. Part 2. Malignant tumors. *Radiographics* 2011;31(2):483–507
- 55 Gow KW, Blair GK, Phillips R, et al. Obstructive jaundice caused by neuroblastoma managed with temporary cholecystostomy tube. *J Pediatr Surg* 1995;30(6):878–882
- 56 Spunt SL, Lobe TE, Pappo AS, et al. Aggressive surgery is unwarranted for biliary tract rhabdomyosarcoma. *J Pediatr Surg* 2000;35(2):309–316
- 57 Koenigsberg M, Wiener SN, Walzer A. The accuracy of sonography in the differential diagnosis of obstructive jaundice: a comparison with cholangiography. *Radiology* 1979;133(1):157–165
- 58 Roebuck DJ, Stanley P. External and internal-external biliary drainage in children with malignant obstructive jaundice. *Pediatr Radiol* 2000;30(10):659–664
- 59 Watanabe Y, Ito T, Horibe K, et al. Obstructive jaundice, an unusual initial manifestation of intraabdominal non-Hodgkin's lymphoma in children: complications of percutaneous transhepatic cholangial drainage. *J Pediatr Surg* 1997;32(4):650–653
- 60 Ugur H, Tacyildiz N, Yavuz G, et al. Obstructive jaundice: an unusual initial manifestation of intra-abdominal non-Hodgkin lymphoma in a child. *Pediatr Hematol Oncol* 2006;23(1):87–90
- 61 Pietsch JB, Shankar S, Ford C, Johnson JE. Obstructive jaundice secondary to lymphoma in childhood. *J Pediatr Surg* 2001;36(12):1792–1795
- 62 Kim SH, Zangan SM. Percutaneous transhepatic biliary drainage complicated by bilothorax. *Semin Intervent Radiol* 2015;32(1):54–56
- 63 Keil R, Snajdauf J, Rygl M, et al. Diagnostic efficacy of ERCP in cholestatic infants and neonates—a retrospective study on a large series. *Endoscopy* 2010;42(2):121–126
- 64 Petersen C, Meier PN, Schneider A, et al. Endoscopic retrograde cholangiopancreatography prior to explorative laparotomy avoids unnecessary surgery in patients suspected for biliary atresia. *J Hepatol* 2009;51(6):1055–1060
- 65 Ohnuma N, Takahashi T, Tanabe M, Yoshida H, Iwai J. The role of ERCP in biliary atresia. *Gastrointest Endosc* 1997;45(5):365–370
- 66 Malloy PC, Grassi CJ, Kundu S, et al; Standards of Practice Committee with Cardiovascular and Interventional Radiological Society of Europe (CIRSE) Endorsement. Consensus guidelines for periprocedural management of coagulation status and hemostasis risk in percutaneous image-guided interventions. *J Vasc Interv Radiol* 2009;20(7, Suppl):S240–S249
- 67 Saad WE, Wallace MJ, Wojak JC, Kundu S, Cardella JF. Quality improvement guidelines for percutaneous transhepatic cholangiography, biliary drainage, and percutaneous cholecystostomy. *J Vasc Interv Radiol* 2010;21(6):789–795
- 68 Ryan JM, Ryan BM, Smith TP. Antibiotic prophylaxis in interventional radiology. *J Vasc Interv Radiol* 2004;15(6):547–556
- 69 Moon E, Tam MD, Kikano RN, Karuppasamy K. Prophylactic antibiotic guidelines in modern interventional radiology practice. *Semin Intervent Radiol* 2010;27(4):327–337
- 70 Venkatesan AM, Kundu S, Sacks D, et al; Society of Interventional Radiology Standards of Practice Committee. Practice guidelines for adult antibiotic prophylaxis during vascular and interventional radiology procedures. Written by the Standards of Practice Committee for the Society of Interventional Radiology and Endorsed by the Cardiovascular Interventional Radiological Society of Europe and Canadian Interventional Radiology Association [corrected]. *J Vasc Interv Radiol* 2010;21(11):1611–1630, quiz 1631
- 71 Bratzler DW, Dellinger EP, Olsen KM, et al; American Society of Health-System Pharmacists (ASHP); Infectious Diseases Society of America (IDSA); Surgical Infection Society (SIS); Society for Healthcare Epidemiology of America (SHEA). Clinical practice guidelines for antimicrobial prophylaxis in surgery. *Surg Infect (Larchmt)* 2013;14(1):73–156
- 72 Johnson C, Martin-Carreras T, Rabinowitz D. Pediatric interventional radiology and dose-reduction techniques. *Semin Ultrasound CT MR* 2014;35(4):409–414
- 73 Aria D, Vatsky S, Towbin R, Schaefer CM, Kaye R. Interventional radiology in the neonate and young infant. *Semin Ultrasound CT MR* 2014;35(6):588–607
- 74 Franchi-Abella S, Cahill AM, Barnacle AM, Pariente D, Roebuck DJ. Hepatobiliary intervention in children. *Cardiovasc Intervent Radiol* 2014;37(1):37–54

- 75 Lorenz JM, Denison G, Funaki B, Leef JA, Van Ha T, Rosenblum JD. Balloon dilatation of biliary-enteric strictures in children. *AJR Am J Roentgenol* 2005;184(1):151–155
- 76 Miraglia R, Maruzzelli L, Caruso S, et al. Percutaneous management of biliary strictures after pediatric liver transplantation. *Cardiovasc Intervent Radiol* 2008;31(5):993–998
- 77 Ginat D, Saad WE. Cholecystostomy and transcholecystic biliary access. *Tech Vasc Interv Radiol* 2008;11(1):2–13
- 78 Hatjidakis AA, Karampekios S, Prassopoulos P, et al. Maturation of the tract after percutaneous cholecystostomy with regard to the access route. *Cardiovasc Intervent Radiol* 1998;21(1):36–40