Ultrasound-guided Percutaneous Cholecysto-Cholangiography: A Potpourri of Infantile Cholestasis Conditions

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Learning objectives

1. To explain how to perform ultrasound (US)-guided percutaneous cholecysto-cholangiography (PCC) in infants.
2. To describe the PCC characterizations in various conditions of cholestatic jaundice with correlation to other imaging modalities based on illustrated clinical cases that are unique or more common in infants.
Background

- Infantile cholestatic jaundice defined as persistent direct hyperbilirubinemia if it continues beyond two weeks after birth.
- It has a broad differential diagnosis including congenital, acquired and inherited etiologies. The major diagnostic challenge is to distinguish medical causes from the surgical conditions.
- The accurate diagnosis is based on the information obtained from the different imaging modalities and other investigative examinations at early stage as any delay in the diagnosis of certain conditions can lead to poorer prognosis.
- Although several imaging modalities are available for assessment of infants with cholestatic jaundice, all imaging modalities lack complete specificity.
- US-guided PCC is considered an additional valuable definitive diagnostic modality because it provides definitive cholangiogram with precise visualization of the biliary tree.
Imaging findings OR Procedure details

PCC Contraindications

- Uncorrectable or refractory coagulopathy
- Coagulopathy can be corrected to have International normalized ratio (INR) $\leq 1.5$ and Platelets count $\geq 50,000$
- Allergy history to iodinated contrast agents
- Large volume ascites: Pre- / peri- procedural paracentesis can be performed.
- No safe percutaneous access route to the gallbladder.

PCC Technique

- The patient is usually fasting for more than 4 hours prior to the procedure. The patient receives intravenous antibiotic
- The procedure is performed in the Interventional Radiology Suite under general anesthesia or moderate sedation provided by the anesthesiologist or pediatrician.
- The procedure is performed with aseptic technique with full precautions taken.
- Local anesthesia with 1% of lidocaine is usually used for skin access.
- US is used to identify the gallbladder and plan the pathway through the intervening liver.
- With real-time US guidance, a small 21- to 23-gauge needle is advanced percutaneously into the gallbladder at a parallel or minimal acute angle to the long axis of the gallbladder.
- Following the puncturing of the gallbladder, normal saline was slowly injected into the lumen with real-time US guidance to ascertain an appropriate puncture by observing the dilation of the gallbladder.
- With fluoroscopic guidance, nonionic contrast material is slowly injected and multiple images are then obtained.
- At the end of the procedure, the needle was removed and pressure applied to the puncture site for 10 min.
- Percutaneous liver biopsy is frequently required at the same session of PCC to complete the study. It is usually obtained under ultrasound guidance with an 18-G core needle using coaxial technique after performing PCC procedure.
- Follow-up US is performed to evaluate for a fluid collection or bleeding.
- Careful monitoring for 4 - 6 hours after the procedure.
- Complete blood count is usually obtained after 4 hours.

Biliary Disorders
Biliary Atresia

- Biliary atresia is the most common cause of neonatal cholestasis and the primary indication for pediatric liver transplantation.
- It is a cholangio-destructive disease of the bile ducts of unknown etiology. Its clinical presentation is a hard liver, white stools and direct hyperbilirubinemia.
- It is the most common surgically treatable cause of cholestasis encountered during the newborn period.
- The most reliable information is obtained by hepatic histopathology and direct visualization of obliterated extrahepatic bile ducts during cholangiography (Fig 2).
- Biliary atresia types is classified based on anatomic findings as proposed by Kasai et al (1976):
  - Type I: common bile duct is obliterated.
  - Type II: common hepatic duct atresia.
  - Type III: extrahepatic ducts atresia to the level of porta hepatis (>90%).

  - Hepatobiliary scintigraphy with 99mTc-DISIDA shows lack of excretion into the intestines, on 24-hour delayed images suggesting biliary atresia or other extrahepatic occlusion. Phenobarbital can be used to differentiate between biliary atresia and neonatal hepatitis in the absence of small bowel activity.
  - The appearance of biliary atresia on PCC varies according to the type and whether the choledochal remnant and biliary ducts are opacified,

Choledochal cyst

- Choledochal cyst is a rare cause of neonatal cholestatic jaundice and it is more common in females.
- It is the result of an abnormally long pancreaticobiliary duct (common channel) allowing reflux of pancreatic enzymes into the biliary ducts.
- Although its clinical manifestation is not specific, its association with biliary atresia should always be ruled out.
- Its differential diagnoses include biliaryolithiasis, pancreatic pseudocyst, hepatic cyst, primary sclerosing cholangitis, enteric duplication, biliary hamartomas, microabscess and biliary papillomatosis.
- Five types of congenital biliary cysts have been described

Type 1:

- It is the most common (~90%) and is confined to the extrahepatic bile ducts.
- Three subtypes of type 1 are described, depending on the extent of disease.

- **Subtype 1A - diffuse type:**

The entire extrahepatic bile duct is involved and the gallbladder communicates with the cyst.

-Cholangiogram shows marked dilatation of the extrahepatic biliary tree. The gallbladder arises from the choledochal cyst and the intrahepatic biliary tree is normal.

- **Subtype 1B - focal type:**

Only a focal segment of the duct is involved.

- Cholangiogram shows focal dilatation of the most distal aspect of the common bile duct with normal segment of it the cyst and the cystic duct.

- **Subtype 1C:**

a fusiform enlargement of the common bile duct with a cylindrical dilatation of the common hepatic duct.

- Cholangiogram shows smooth fusiform dilatation of the common hepatic duct and common bile duct with normal intrahepatic ducts. The gallbladder arises directly from the dilated common bile duct.

**Type 2: (2%)**

- An extrahepatic bile duct diverticulum.

- Cholangiogram shows true diverticulum of the common bile duct.

**Type 3: (1.4-5%)**

- A choledochocele emerging from the intraduodenal segment of the choledochal duct.

- Cholangiogram shows a focal dilated segment of the intraduodenal portion of the common bile duct with normal proximal duct and intrahepatic ducts.

**Type 4:**

- Multiple cystic dilatations along both intra- and extrahepatic ducts (type 4A) or limited to extrahepatic ducts (type 4B).

- Cholangiogram shows multiple common bile duct cysts.

**Type 5: Caroli disease.**
- A Mendelian recessive disorder characterized by a nonobstructive cystic beaded or irregular dilatation of the intrahepatic ducts without hepatic fibrosis or portal hypertension.

- The shape of the dilatation might be saccular or fusiform, and the extent of dilatation varies.

- True Caroli disease is exceptionally rare in infancy and the associated jaundice may be due to acute cholangitis.

- It can be as a distinct entity from the biliary ectasia seen in patients with renal polycystic kidney and congenital hepatic failure.

- Although US of the liver is often adequate for diagnosing; cholangiography is confirmatory.

- Cholangiogram shows multiple saccular or cystic dilatations of the intrahepatic biliary tree with normal extrahepatic biliary tree (Fig. 3).

- The percutaneous cholangiogram permits a better evaluation of the intrahepatic biliary tree as compared to endoscopic retrograde cholangiopancreatography.

### Infantile Sclerosing Cholangitis

- It is a rare, heterogeneous condition resulting from an inflammatory obliteratorive fibrosis that infiltrates the intra- and extrahepatic biliary ducts.
- It can evolve into hepatic insufficiency and cirrhosis, with a risk of cholangiocarcinoma.
- Its main clinical finding in the early form is cholestatic jaundice
- US shows non-specific biliary dilatation.
- PCC reveals intrahepatic biliary duct involvement in 100% of cases and extrahepatic involvement in 60% of cases. Additionally there is diffuse stricturing of the bile ducts with dilatation of the areas in between, giving a beaded appearance. Liver biopsy is diagnostic when cholangiography is normal.
- MR cholangiography shows irregular biliary ductal dilatation and beading that are segmental and intersect multiple hepatic territories.
- Different segmental hepatic areas are involved.
(a) PCC is obtained via direct puncture of the gallbladder with 21G needle.
(b-c) Contrast material fills intra- and extra-hepatic ducts which appear patent (arrow). Contrast material passes into the duodenum.

**Fig. 1:** Normal Cholangiogram

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Fig. 2: Biliary Atresia

(a) Contrast material is injected directly via the gallbladder.
(b) PCC shows no filling of extra-hepatic ducts and the contrast material passes into the duodenum.
(a-b) PCC shows irregular, dilated saccular ductal branches.

(c) Contrast-enhanced CT scan shows saccular dilatation of intrahepatic bile ducts.

Fig. 3: Caroli Disease

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Conclusion

• Well understanding of the infantile cholestatic jaundice and their imaging are important for the diagnosis and management.
• Although it is relatively invasive, ultrasound-guided PCC is safe, reliable, and diagnostically accurate imaging modality for evaluation of cholestatic jaundice in infants as it provides assessment of the whole biliary tree.
References


