Fetal Sonography and Neonatal Scintigraphy of a Choledochal Cyst

Martha A. Wiedman, Anton Tan, and Charles J. Martinez

Department of Diagnostic Radiology and Division of Nuclear Medicine, Mount Sinai Hospital Medical Center, Chicago, Illinois

A choledochal cyst, observed by ultrasound at 27 wk gestation and diagnosed at 36 wk gestation, was confirmed postpartum by [^{99m}Tc]DISIDA cholescintogram. Cystic dilatation of the common bile duct was proven by surgical excision and histological verification. Ultrasonography provides in utero diagnosis of choledochal cyst, often vital for successful management. Technetium-99m DISIDA imaging is a valuable companion for defining function of the cyst.

J Nucl Med 26:893–896, 1985

Choledochal cyst is a rare, localized dilatation of the common bile duct first reported by Todd in 1817 *(1)*. The following report describes a choledochal cyst observed by ultrasound at 27 wk gestation, one of the earliest detected *(2)*. A similar case was documented at 2 days of age *(2)*. Early suspicion of a cyst by ultrasound and functional definition by technetium-99m disofenin ([^{99m}Tc]DISIDA) cholescintigram enable prompt surgical excision and reconstruction of the biliary tree. These examinations permit safe and noninvasive diagnosis and immediate repair of a potentially fatal anomaly.

CASE REPORT

A 17-yr-old primagravida underwent ultrasonography for dating purposes at our institution. The study, performed at 27 wk (Fig. 1), revealed a cystic right upper quadrant mass, separate from the kidneys and bladder, which showed no change in size during the examination. At 33 wk, a persistent mass was confirmed in the right upper quadrant, anterior to the kidneys, measuring 3 cm × 3 cm. At 36 wk, repeat sonography identified the anechoic structure inferior to the umbilical vein and portal sinus and adjacent to the right adrenal and kidney (Fig. 2). The location and persistence of this mass were consistent with the diagnosis of choledochal cyst. Differential diagnoses included adrenal, ovarian and mesenteric cyst.

A female infant was delivered by normal spontaneous vaginal delivery at 38 wk by dates, with Apgars of 9 and a weight of 3,350 g. There was no palpable abdominal mass and the child was anicteric. An ultrasound performed immediately postpartum showed a 2.9 × 2.9 round, cystic structure in the right upper quadrant. This mass, which showed no postprandial change in size, was consistent with a choledochal cyst. At 18 hr of age, the patient underwent an injection of 1 mCi of [^{99m}Tc]DISIDA for an abdominal scintigram. This imaging study revealed a large defect in the gallbladder region at 5 min postinjection (Fig. 3). Increasing radionuclide activity was seen within the cyst at 45 min. Tracer increased in the general region of the gallbladder at 1½ hr. Neither common duct nor bowel activity was seen. There was no emptying of the dilated structure after feeding of the infant. Seventeen hours postinjection, a follow-up view showed bowel activity but could not identify a gallbladder. The images were compatible with a choledochal cyst.

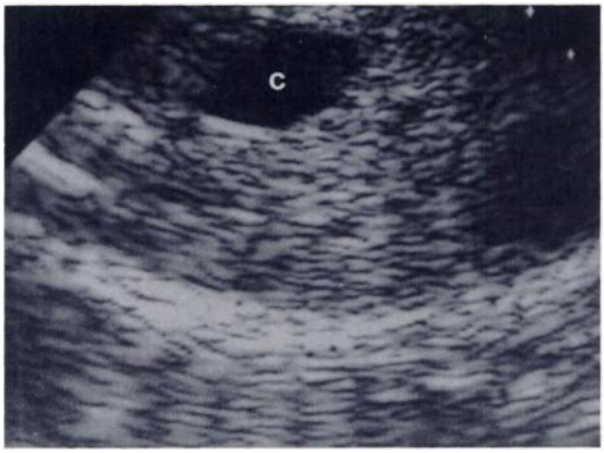


FIGURE 1
Longitudinal ultrasound of abdomen at 27 wk. Cystic area in center of fetal abdomen (c)

Received Oct. 16, 1984; revision accepted May 7, 1985.

For reprints contact: Martha A. Wiedman, MD, Dept. of Diagnostic Radiology, Mt. Sinai Hospital Medical Center, California and 15th Sts., Chicago, IL 60608.

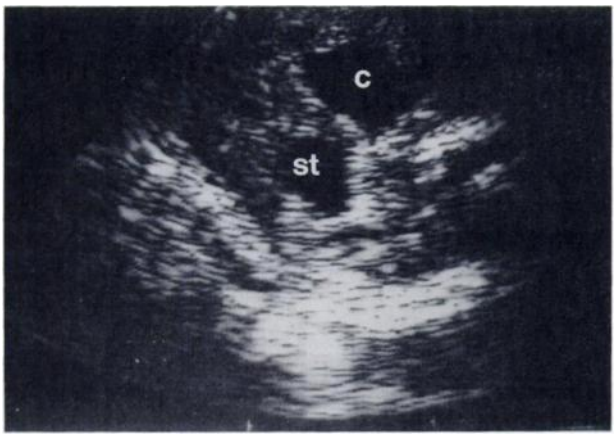


FIGURE 2
Longitudinal abdominal ultrasound at 36 wk. Choledochal cyst (c), 2.6 cm × 2.2 cm, anterior to stomach (st) and inferior to umbilical vein and portal sinus

The patient was transferred to another institution for surgical resection of the cyst. A preoperative ultrasound (Fig. 4) revealed a 4 cm × 3 cm × 3 cm anechoic cystic mass in the periportal and subhepatic region. No intra- or extrahepatic ductal dilatation was noted. A small gallbladder was identified anterior to the mass. Laparatomy disclosed a cystic dilatation of the common bile duct, extending from the confluence of the hepatic ducts to the intrapancreatic portion of the bile ducts. The dilated portion measured 6 cm in diam. Choledochocystectomy and choledochoduodenostomy were performed. The patient had a transient postoperative wound infection with an otherwise uneventful recovery.

DISCUSSION

Alonso-Lej classified four types of cyst, of which the most common, cystic dilatation of the bile duct, was found in this study (*3*). It is most often seen in children or in young adults, but may be diagnosed at any age, with a female predominance of 4:1 (*2*). The classical clinical triad of jaundice, abdominal pain and abdominal mass is found in less than one-half of patients (*3*). The currently accepted etiology of choledochal cyst is the anomalous insertion of the common bile duct into the pancreatic duct which precludes normal sphincter development at their juncture (*1,4*). The resulting reflux of pancreatic juices into the biliary tree may predispose to cholangitis, fibrosis of the common duct, pancreatitis, common duct stone, portal hypertension (*1*) and biliary cirrhosis. The incidence of adenocarcinoma in patients with choledochal cysts has been estimated as twenty times that of the general population (*5*). Choledochal cysts are also associated with intrahepatic cystic dilatation of the biliary tract (Caroli's disease) in ~50% of these patients (*6*). Patients with Caroli's disease are also prone to develop cholangitis, due to intrahepatic cholestasis and to choledochojejunostomy (*6*).

Radionuclide scintigrams have gained increasing acceptance for their accuracy and practicality in biliary imaging. One study describes accurate detection of choledochal cyst in ten of 12 patients using ^{99m}Tc-labeled agents (*7*). The ^{99m}Tc-labeled compounds give improved diagnostic information with decreased ab-

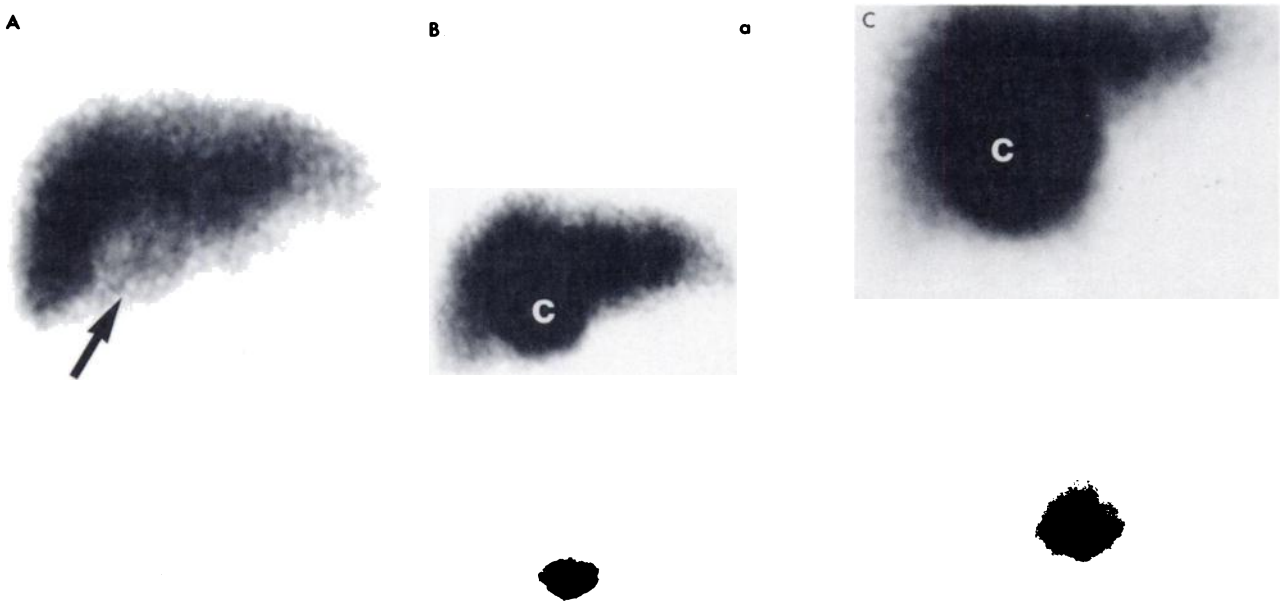
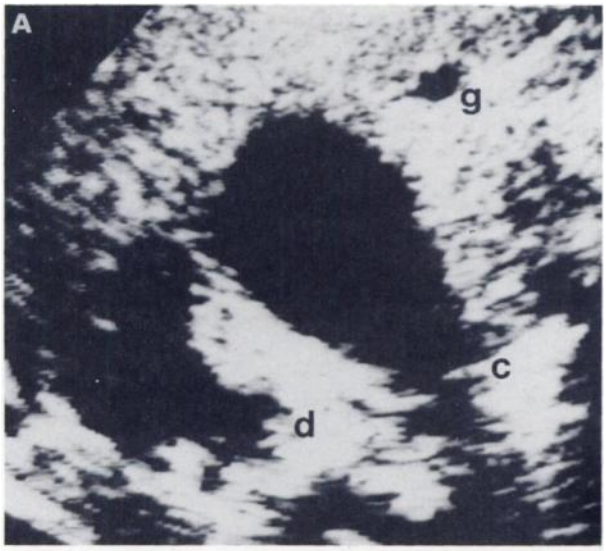
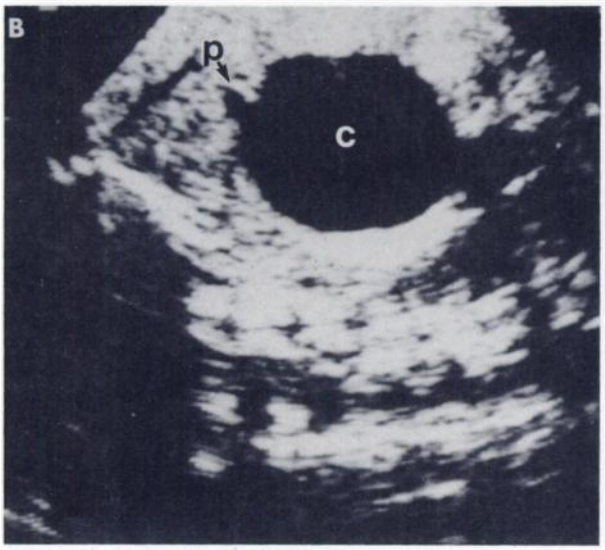


FIGURE 3
[^{99m}Tc]DISIDA imaging study at 18 hr of age. A: Anterior image of liver at 5 min shows photon-deficient area in gallbladder region (arrow). B: Increased radionuclide activity occurs within choledochal cyst (c) at 45 min. C: Accumulation of tracer is seen within cyst (c) at 1½ hr





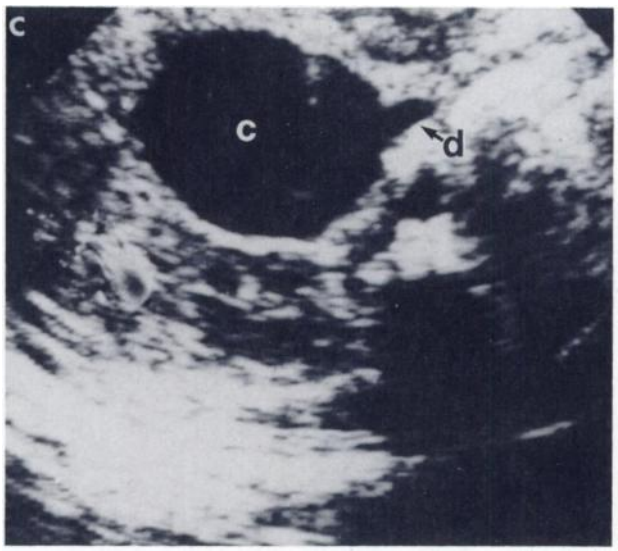


FIGURE 4
Transverse abdominal ultrasound at 10 days of age. A: Choledochal cyst (c), small anterior gallbladder (g) and duodenum (d). Letters are adjacent and to right of structures. B: Proximal common bile duct (p) and cyst (c). C: Distal common bile duct (d) and cyst (c)

sorbed radiation dose compared to iodine labeled complexes, such as iodine-131 rose bengal. They enable recognition of the hepatobiliary abnormalities by showing the transport of the radiopharmaceutical from the hepatocyte to the biliary tree and its excretion into the gut (*8*). A normal [^{99m}Tc]DISIDA study demonstrates the liver, gallbladder, intra- and extrahepatic ducts and gut within 1 hr of injection (*8*). In the current case, the gallbladder is never visualized and gut activity is delayed until seventeen hr postinjection, consistent with the diagnosis of choledochal cyst. The [^{99m}Tc]DISIDA study of this case resembles "pattern A" discussed by Huang (*7*). His report shows a photon-deficient area around the gallbladder in early images which progressively accumulates radioactive tracer in 24-hr postinjection scintigrams. This is the most common presentation of choledochal cyst.

This case provides an unusual opportunity to observe an asymptomatic patient with choledochal cyst in prenatal and postnatal stages. This patient lacks the jaundice, palpable mass and abdominal pain found in classical histories. Although there is no definite change in the size of the cyst from fetal to neonatal studies, the markedly abnormal [^{99m}Tc]DISIDA imaging at 10 days of age predicts inevitable biliary malfunction. In the absence of preliminary studies, delay in diagnosis and corrective surgery can increase the risk of morbidity. The value of sonography in early detection of choledochal cyst lies in the preparation for surgical intervention and prevention of serious biliary obstruction. While ultrasound depicts the size, contour and location of the choledochal cyst, nuclear medicine imaging shows technical superiority by demonstrating both anatomy and function of the cyst. In this case, despite the patient's lack of symptoms, radionuclide images reveal a distinct delay in biliary transport with the potential for obstruction and medical compromise. Ultrasound is quick, well tolerated by children and in-

volves no known radiation risk. Although routine use of ultrasound is inadvisable (*9*), it is the only available method for prenatal diagnosis of choledochal cyst. Complementary use of ultrasound and postnatal scintigraphy is essential for optimal patient management.

ACKNOWLEDGMENT

The author thanks Orestes Sanchez, MD for his aid in research and editorial assistance.

REFERENCES

1. Babbitt DP, Starshak RJ, Sty JR: Choledochal cyst: Pathogenesis, diagnosis and surgical implications. *Appl Radiol* 10:125–137, 1981
2. Frank JL, Hill MC, Chirathivat S, et al: Antenatal observation of a choledochal cyst by sonography. *Am J Roentgenol* 137:166–168, 1981
3. Alonso-Lej F, Rever WB, Pessagno DJ: Congenital choledochal cyst, with a report of 2, and an analysis of 94 cases. *Int Abstr Surg* 108:1–30, 1959
4. Babbitt DP, Starshak RJ, Clemett AR: Choledochal cyst: A concept of etiology. *AJR* 119:57–62, 1973
5. Kagawa Y, Kashihara S, Kuramoto S, et al: Carcinoma arising in a congenitally dilated biliary tract. *Gastroenterology* 74:1286–1294, 1978
6. Kuni CC, Klingensmith WC III: Congenital malformations. In *Atlas of Radionuclide Hepatobiliary Imaging,* Boston, GK Hall, 1983, pp 189–213
7. Huang M-J, Liaw Y-F: Intravenous cholescintigraphy using Tc-99m-labeled agents in the diagnosis of choledochal cyst. *J Nucl Med* 23:113–116, 1982
8. Sty JR, Starshak RJ, Miller JH: Gastrointestinal nuclear medicine. In *Pediatric Nuclear Medicine,* Norwalk, CT, Appleton-Century-Crofts, 1983, pp 66–74
9. Diagnostic ultrasound in pregnancy (Editorial). *Lancet* ii:201–202, 1984