

Imaging Sciences Interesting Cases

CASE 549

Vikas Datta, MD

CLINICAL PRESENTATION: Patient is an 8-day-old female born at 34 weeks gestational age, presenting with direct hyperbilirubinemia.

IMAGING FINDINGS: See figure legends.

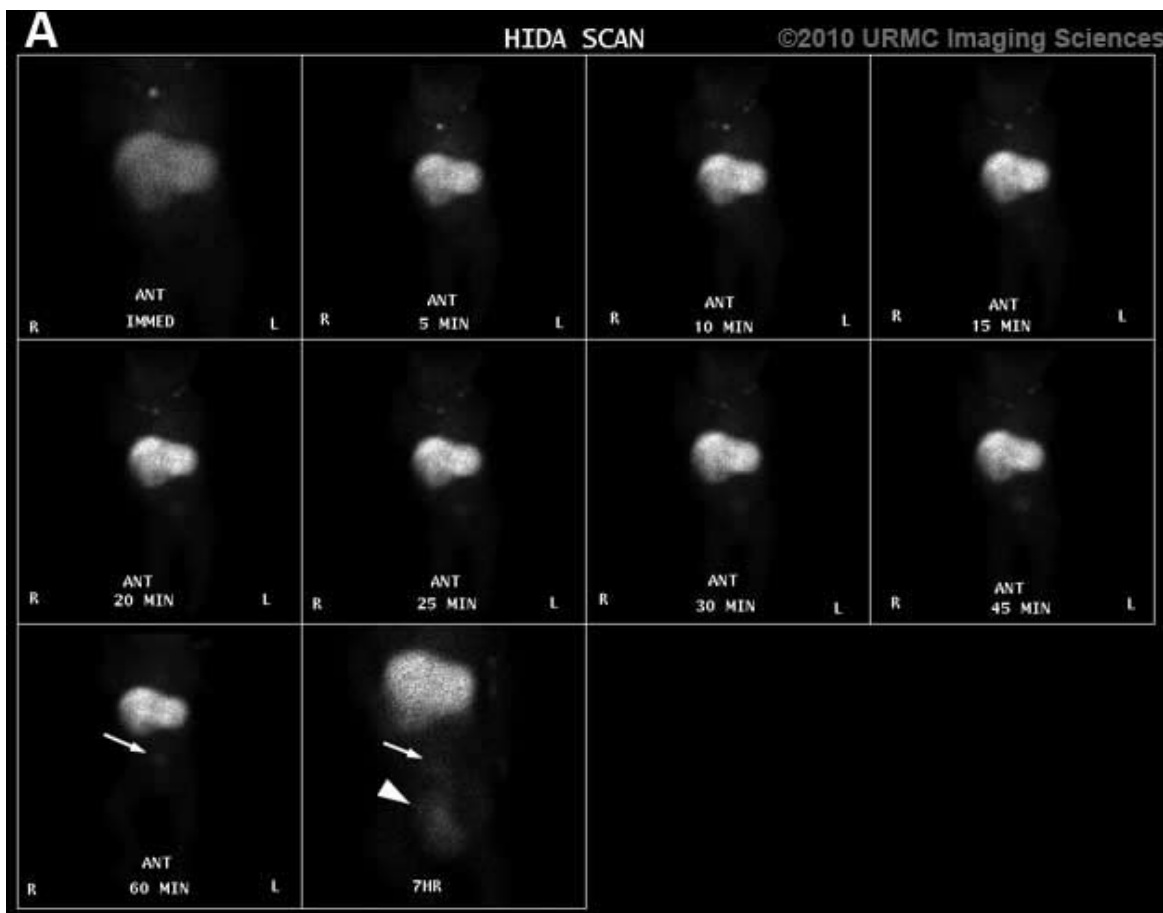


Figure 1A: Hepatobiliary scan demonstrates prompt uptake of radiotracer by the liver with increasing hepatocellular concentration on successive images. The extrahepatic biliary tree and gallbladder are not visualized. Bowel activity is absent. Radiotracer is visualized within the bladder (arrow) and diaper (arrowhead), secondary to alternate excretion.

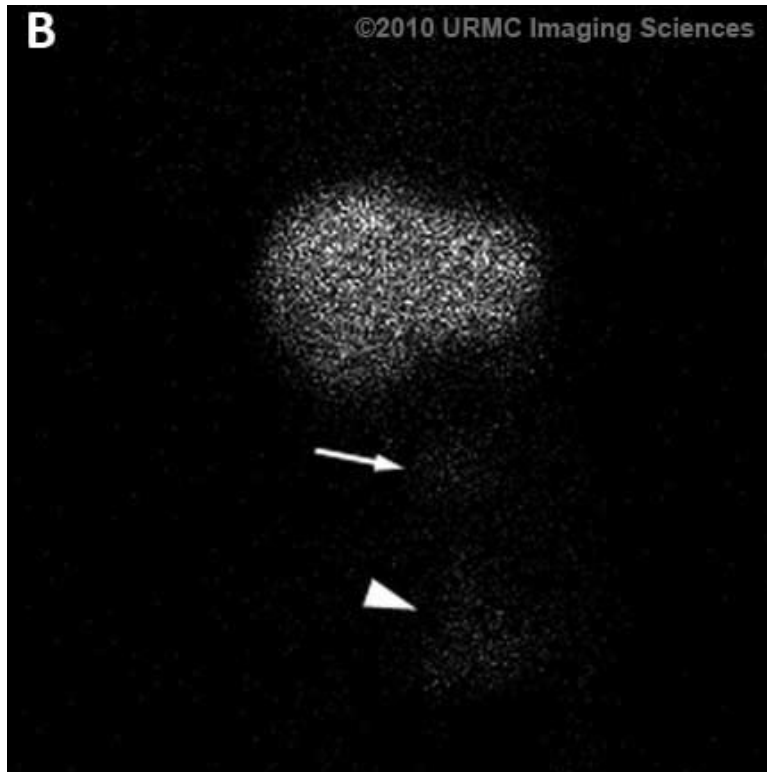


Figure 1B: A 24-hour delayed image demonstrates no evidence of radiotracer transit within the small bowel. Residual radiotracer is visualized in the bladder and diaper.

DIAGNOSIS: Biliary atresia

DISCUSSION: Biliary atresia is a rare abnormality seen in neonates characterized by the absence or severe deficiency of the extrahepatic biliary system. It is suspected to originate from prenatal biliary duct inflammation of unknown etiology. The incidence of biliary atresia is approximately 1 per 10,000-13,000 live births. Classification of biliary atresia is based on the predominant site of atresia: type I involves obliteration of the common duct; type II is characterized by atresia of the common hepatic duct; and type III (>90% of patients) involves atresia of the right and left hepatic ducts to the level of the porta hepatis. Biliary atresia is suggested clinically with progressive direct hyperbilirubinemia in the neonate. If not surgically corrected, secondary biliary cirrhosis invariably results. Approximately 12% have patent proximal ducts and can have simple re-anastomosis surgery; however, 88% require the Kasai procedure to anastomose a loop of small bowel to the dissected porta hepatis. In the evaluation of biliary atresia, hepatobiliary scans and sonography are of most utility.

Hepatobiliary scans are most commonly performed with Tc-99m disofenin (DISIDA) or mebrofenin (BRIDA). Patients are pretreated with either phenobarbital (most common) or ursodeoxycholic acid, which enhances biliary excretion and has been shown to improve scintigraphic accuracy. Initial images should demonstrate clearance of radiotracer from the blood pool with uptake by the liver. Hepatocyte uptake of radiotracer is usually preserved in the first 2-3 months and later deteriorates, at which point it can be confused with neonatal hepatitis. Lack of excretion of radionuclide into the intestines on 24-hour delayed images is highly suggestive of biliary atresia or other extrahepatic occlusion. Visualization of the gallbladder does not exclude biliary atresia, as it is seen in 25% of patients. Hepatobiliary scans have a sensitivity approaching 100% and specificity of approximately 87%.

Sonography is often performed to evaluate for alternate causes of jaundice. In biliary atresia, ultrasound may demonstrate hepatomegaly, although echotexture will commonly be normal. The gallbladder may be present in 25% of patients, with an irregular shape or wall thickness suggestive of biliary atresia. Extrahepatic bile ducts are replaced by an echogenic triangular cord sign which is thought to be a fibrotic rem-

nant of the common duct. Color Doppler is useful to demonstrate the main portal vein and hepatic artery when searching for the triangular cord sign.

REFERENCES:

1. Gerhold JP, Klingensmith WC 3rd, Kuni CC, et al: Diagnosis of biliary atresia with radionuclide hepatobiliary imaging. *Radiology*. 1983 Feb;146(2):499-504. PMID: 6681570 [PubMed]
2. Mettler FA, Guiberteau MJ. *Essentials of Nuclear Medicine Imaging*. 5th ed. Philadelphia: Saunders Elsevier, 2006.