

CASE REPORT

Pre-operative Non-Invasive Imaging for Neonatal Cholestasis in a Child with Extrahepatic Biliary Atresia

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ABSTRACT

Hepatobiliary iminodiacetic acid (HIDA) scintigraphy is a non-invasive, functional imaging of the hepatobiliary system that serves as an adjunct imaging modality for neonatal cholestasis work-up. In view of the urgency to diagnose biliary atresia and restore bile flow through surgery, HIDA scintigraphy could help to distinguish between neonatal cholestasis due to biliary atresia and neonatal hepatitis of various causes. We describe a full-term male infant with jaundice beyond the physiological period in which HIDA scintigraphy showed absent tracer excretion from the biliary system into the intestines up to 5 hours on follow-up imaging. The intraoperative diagnosis confirmed the diagnosis of biliary atresia. The prognosis of the patient with biliary atresia depends on early surgical planning and intervention. Therefore, non-invasive diagnostic tools play an important role in the evaluation of a child with neonatal cholestasis.

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INTRODUCTION

Hepatobiliary iminodiacetic acid (HIDA) scintigraphy is a non-invasive, functional imaging of the hepatobiliary system that serves as an adjunct imaging modality for neonatal cholestasis work-up. In view of the urgency to diagnose biliary atresia and restore bile flow through surgery, HIDA scintigraphy could help to distinguish between neonatal cholestasis due to biliary atresia and neonatal hepatitis of various causes (1). We describe a full-term male infant with jaundice beyond physiological period in which HIDA scintigraphy showed absent tracer excretion from the biliary system into the intestines up to 5 hours on follow-up imaging. The intraoperative diagnosis confirmed the diagnosis of biliary atresia.

CASE REPORT

A 36-day-of-life boy presented with yellowish discoloration of the sclera since day 7 of life and intermittent episodes of passing pale stools since day 22 of life. He was born term at 39 weeks and 3 days via spontaneous vaginal delivery with a birth weight of

2.9 kg. Postnatally, he was started on combine breast and formula milk feeding on demand. He was thriving well and there was no episode of fever, vomiting or tea-coloured urine. Initially, he had pigmented stools since birth. However, at first week of life, he was noted to be jaundiced during the primary care visit. Subsequently after 2 weeks of monitoring, he was referred to paediatric team for prolonged jaundice workout. His mother noticed that he intermittently passed acholic stool during the admission to the ward.

On examination, he was afebrile and active on handling. There was tinge of jaundice on the sclera with dark complexion of the skin. No dysmorphism features noted. His anterior fontanelle was not bulging with good hydration and stable vital signs. Other systemic examinations were unremarkable.

Laboratory investigation showed deranged liver function test with conjugated hyperbilirubinemia and transaminitis. Other investigations including full blood count, renal profile and viral serology were unremarkable.

His ultrasound abdomen at 26 days of life showed homogenous liver echo textures with smooth margin. Liver span was 5.7 cm with no liver lesion or biliary dilatation. Portal veins and hepatic artery were patent

with normal caliber (ratio of 0.28). Echogenic band visualised anterior to the right portal vein measures 1.1mm and at main portal vein 0.9mm. The gallbladder was not visualised despite adequate fasting for 6 hours. He was referred for diagnostic HIDA scintigraphy to rule out biliary atresia and imaging was performed at day 36 of life following 5-days oral phenobarbital ingestion (2.5mg/kg BD) as pre-imaging preparation to activate liver enzymes and improve liver function. He was kept nil-by-mouth for 4 hours prior HIDA scintigraphy. Following intravenous injection of 54 MBq technetium-99m mebrofenin, dynamic imaging was performed anteriorly using dual-head gamma camera for 60 minutes (2 min/frames) followed by static imaging at right lateral and left lateral views (500K counts). In view of no tracer excretion from the liver into the intestine at first hour despite good tracer uptake by the liver and early clearance of tracer from the blood pool, subsequent serial static imaging was performed at 3rd hour and 5th hour (Fig. 1 and 2). Unfortunately, there was persistent non-visualisation of the tracer excretion from the liver into the bowel even at the 5th hour. Thus, HIDA scintigraphy showed high suspicion of biliary atresia.

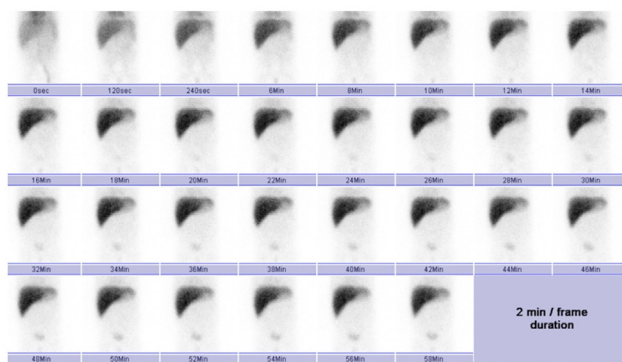


Figure 1: Dynamic images HIDA scintigraphy at 1 hour shows prompt uptake of tracer by the liver and adequate tracer clearance from the blood pool following intravenous injection.

He underwent Kasai hepatoportoenterostomy at 45 days of life to restore bile flow. Intraoperative findings showed atrophic gallbladder with a cyst near the Hartmann’s pouch. The extrahepatic biliary tree appears fibrotic. The liver appears soft with gross neovascularisation and the spleen was normal. There was minimal bile seen after the excision of the portal plate. Following the surgery, he recovered and tolerated feeding well. He was planned for routine follow-up to monitor his liver function.

DISCUSSION

Biliary atresia is an obliterative inflammatory cholangiopathy affecting variable length of both extrahepatic and/or intrahepatic biliary duct of neonates. This condition is rare with estimates prevalence of 1 in 5000 to 19000 live births and appears higher in Asian countries compared to Europe and America. Despite

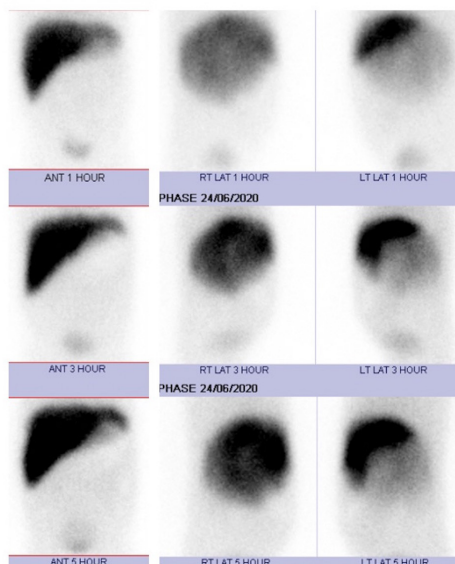


Figure 2: Serial static images taken at 1, 3, and 5 hour demonstrates homogenous liver uptake of tracer with minimal activity seen in the urinary bladder. The gallbladder is not visualised. No tracer excretion from the liver into the bowel noted following tracer injection.

its rarity, it is important to diagnose biliary atresia because early surgical intervention affects prognosis and morbidity of the patient.

Biliary atresia can be classified based on anatomical defect from distal to the most proximal part of biliary system whereby type 1 has obliterated common bile duct (CBD) and the rest of proximal biliary drainage has normal luminal patency (~5% of cases), type 2a has obliterated common hepatic duct (CHD) and type 2b has biliary obstruction involving CBD, CHD and cystic duct (total about 2% of cases). The majority (>90% of cases) is type 3 whereby the extrahepatic biliary system is entirely obliterated till within the porta hepatis (Fig. 3) (2).

Patient with biliary atresia usually present with prolonged jaundice beyond day 14 of life or pale stool as a result of cholestasis. The initial assessment of a child with neonatal cholestasis includes extensive history taking, clinical examination, laboratory investigation and special imaging techniques to exclude or confirm biliary atresia. Hodgson et al assessed bilirubin level among 420 neonates who were on follow-up for prolonged jaundice and found out that conjugated (direct) bilirubin of more than 25 µmol/L up to day 42 of life, as compared to ratio of direct/total bilirubin was a more useful parameter (3). The gold standard to differentiate between the different causes of neonatal cholestasis is by percutaneous liver biopsy with sensitivity of 90-100% and specificity of 80-90%. The presence of bile duct proliferation, bile plugs, periportal fibrosis, giant cells transformation, canalicular and cellular cholestasis on histopathological examination are diagnostic of biliary atresia (1). However, liver biopsy is not the most appropriate

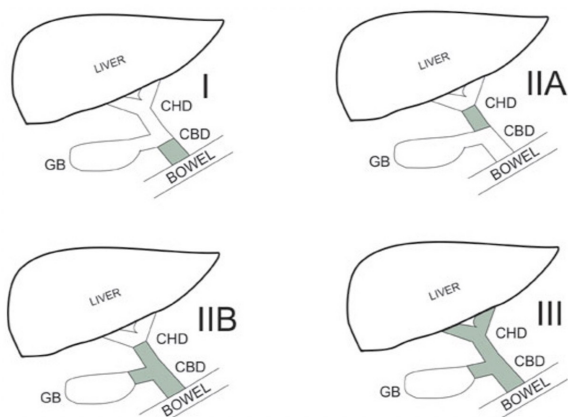


Figure 3: Ohi classification of biliary atresia. CBD - common bile duct, CHD - common hepatic duct, GB - gallbladder. Adapted from (2).

initial investigation as it is invasive and could be hazardous in patient with abnormal clotting factors due to compromised liver function. Different imaging modalities may be performed including ultrasonography (USG), HIDA scintigraphy and magnetic resonance cholangiopancreatography (MRCP). The first imaging modality of choice is abdominal USG as it is widely accessible, non-invasive and non-ionising radiation. However, although its specificity in detecting biliary atresia is high, it is highly operator dependent and has low sensitivity (49-73%) to detect the hyperechogenic liver hilum or triangular cord sign which is an indicator of extrahepatic biliary atresia (2). A study on 50 patients with various causes of cholestasis demonstrated that all patients with intrahepatic biliary atresia had negative triangular cord sign as compared with extrahepatic biliary atresia (4). Thus, HIDA scintigraphy is recommended as the subsequent non-invasive modality to evaluate bile duct patency. Patients with intrahepatic cholestasis either due to idiopathic neonatal cholestasis, infective hepatitis, metabolic liver disease, intrahepatic biliary atresia or Alagille syndrome have poor tracer uptake and drainage into the intestines during HIDA scintigraphy and those with extrahepatic cholestasis due to extrahepatic biliary atresia or choledochal cysts have good tracer uptake from the blood pool but poor or absent tracer drainage into the intestines. HIDA scintigraphy has sensitivity of 98.7% and specificity 70.4% of which specificity increases with the usage of pre-imaging phenobarbital and ursodeoxycholic acid (5). The visualisation of extrahepatic duct either CBD or

CHD on MRCP may ruled out biliary atresia.

The therapeutic effect of Kasai hepatopertoenterostomy among biliary atresia patients varies considerably. However, Kasai procedure in older age patients is associated with poorer prognosis and many centres opted for liver transplantation instead of Kasai procedure in patients older than 90 days old.

CONCLUSION

The prognosis of patients with biliary atresia depends on early surgical planning and intervention. Therefore, non-invasive diagnostic tools play important role in evaluation of child with neonatal cholestasis and distinguish biliary atresia from other causes of neonatal jaundice. HIDA scintigraphy is recommended to detect obstructive biliary lesion needing an urgent surgery.

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