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Perspective

# Perspective on Neonatal Hepatitis and Cholestasis

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## PERSPECTIVE

Neonatal hepatitis refers to many forms of liver dysfunction that affects foetuses and neonates. It is most often caused by viruses or metabolic diseases, and many cases are of an unknown cause.

The infant with neonatal hepatitis usually has jaundice that appears at one to two months of age, is not gaining weight and growing normally, and has an enlarged liver and spleen. Infants with this condition are usually jaundiced. Jaundice that is caused by neonatal hepatitis is not the same as physiologic neonatal jaundice. In contrast with physiologic neonatal jaundice, infants with neonatal hepatitis present with dark urine. Infants may also present with delayed growth.

Neonatal hepatitis is defined as a clinicopathologic disorder in the absence of an identifiable etiology and is among the most common causes of neonatal cholestasis. Previously, neonatal hepatitis accounted for up to 50% of all cases. Over time, the relative percentage of cases has decreased to 25–30% due to recognition of specific disorders mimicking neonatal hepatitis. Two distinct patterns have been identified, familial and sporadic. The sporadic form occurs more frequently and has a better prognosis; over 80% of infants will recover or have residual nonprogressive liver disease.

Familial neonatal hepatitis has a poorer prognosis, with up to 80% of patients developing end-stage liver disease. Most investigators believe that the familial form represents variants of PFIC and other undiscovered genetic disorders. Other diseases that originally were classified as idiopathic neonatal hepatitis include identified infections, α1-antitrypsin deficiency, Alagille's syndrome, Niemann–Pick type C, PFIC, bile acid synthesis defects, and other inherited metabolic defects.

Cholestasis in early infancy represents a diagnostic dilemma and most of these infants suffer either from extrahepatic biliary atresia or idiopathic neonatal hepatitis. Differentiation between the two conditions may be extremely difficult both clinically and biochemically, and a diagnostic liver biopsy is usually required. We report on a Sudanese infant who presented at the age of 4 weeks with prolonged cholestatic jaundice, abdominal ultrasound was inconclusive, HIDA scan was suggestive of extrahepatic biliary atresia and the diagnosis of idiopathic neonatal hepatitis was only reached by liver biopsy. The infant made full recovery on supportive treatment during a one year follow up period.

Cholestasis in early infancy is usually difficult to diagnose initially. Most of these infants suffer either from extrahepatic biliary atresia (EHBA) or idiopathic neonatal hepatitis (INH). In about 25% to 50% of infants presenting with conjugated hyperbilirubinemia within the first three months of life, no cause is found. Neonatal hepatitis may be caused by metabolic diseases, viruses or genetic disorders. In INH, however, the cause of inflammation remains unknown. The affected neonates have jaundice, dark urine, light or pale stools, hepatomegaly and varying degrees of coagulopathy may also be seen.

Since INH is a disease of the newborn and the natural history is of gradual resolution in most cases, differentiation from EHBA might be extremely difficult and liver biopsy is usually required for differentiation. Sporadic and familial cases of INH have been described.

We report on a Sudanese infant who presented at the age of 4 weeks with prolonged cholestatic jaundice, abdominal ultrasound was inconclusive, hepatobiliary iminodiacetic acid (HIDA) scan was suggestive of EHBA and the diagnosis of INH was only reached by liver biopsy. The infant made full recovery on supportive treatment during a one year follow up period.

A male infant was brought by his parents at the age of 4 weeks because of yellow discoloration of skin and sclera which started in the second week of life. The infant was born at term, pregnancy was uneventful and his birth weight was 2.6 kg. He was fully breast fed, his urine had deep yellow colour and stool was of normal colour. There was no consanguinity and no family history of neonatal jaundice. Clinical examination revealed a well-baby who was deeply jaundiced and was thriving well. The baby was not dysmorphic and had normal vital signs. Abdominal examination revealed hepatomegaly of 4 cm with liver span of 10 cm.

After consultation with the paediatric surgeon, a percutaneous liver biopsy was obtained which showed foamy degeneration of hepatocytes, giant cell transformation, prominent bile stasis and moderate mixed inflammatory cellular infiltrate in the portal tract with piece meal necrosis which was consistent with giant cell neonatal hepatitis.

The baby was commenced on fat soluble vitamins (A, D, E and K), ursodeoxycholic acid, zinc sulphate and multivitamin syrup. The baby started to show gradual improvement in his liver profile,

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remained stable and maintained normal synthetic liver function tests. He was discharged home after 8 weeks with liver function that showed total serum bilirubin of 14.9 mg/dl and direct bilirubin of 11.3 mg/dl, AST 340 U/L, ALT 219 U/L, alkaline phosphatase 356 U/L, total protein 5g/dl and albumin 3.8 g/dl. The baby was regularly reviewed in the outpatient clinic and when he was seen very recently at the age of one year, he was thriving well with normal development. The most recent liver function test was entirely normal (total protein 4.6g/dl, albumin 3g/dl, AST 11 U/L, ALT 18 U/L, total serum bilirubin 0.7mg/dl, direct 0.6mg/dl, and alkaline phosphatase 150 U/L).