Letter to the Editor
Tiny But Important

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Bile Duct Paucity (BDP) (syndromic – Alagille and non syndromic) is a well recognized, although uncommon cause of neonatal cholestasis(13-). As the clinical presentation can closely resemble extra hepatic biliary atresia, BDP will be missed unless an infant with Neonatal Cholestasis (NC) undergoes a comprehensive work up including a liver biopsy. We present our experience of 9 cases seen over 5 years.

Out of 600 hepato-biliary referrals, 210 (35%) comprised infants with NC. Idiopathic neonatal hepatitis (40%) was the commonest cause of NC followed by extra hepatic biliary atresia (25%). nine cases (4%) were diagnosed with BDP. All nine babies presented with prolonged neonatal jaundice and a history of pale stools. Mean age at presentation was 73 days (range 42110- days). five babies had classical features of Alagille syndrome – growth failure, abnormal facies, posterior embryotoxon, peripheral pulmonic stenosis and butterfly vertebra. Four babies had no features suggestive of Alagille syndrome.Only 3, in whom we manage to do HIDA scans on, had non excreting HIDA scans after 5 days of phenobarbitone. The ultrasound examination in all cases showed a gall bladder. In all patients there was no dilatation of the intrahepatic biliary radicles and nor was a choledochal cyst seen. The infection and metabolic screen was normal. The liver biopsy showed a paucity of bile ducts in all cases (ratio of interlobular bile ducts to arterioles in the portal tracts was less than 0.6). In view of the histology, no infant was subjected to a laparotomy for a peroperative cholangiogram. Supportive therapy in the form of appropriate nutrition (Calories 150 Kcal/kg/d, fat 3g/Kg/d 50% MCT, protein 2 g/kg/d), fat soluble vitamins (vitamin A 500010000- IU/d, 1,25 dihydroxy cholecalciferol 3050- nanograms/kg/d , vitamin E 200 mg/d, vitamine K 5 mg/d),calcium,zink and magnesium and Ursodeoxycholic acid 30 mg/kg/day were instituted. All babies were followed up (mean 24 months, range 1034- months). 4 developed progressive liver failure and were advised a liver transplant. One of them has a succesful liver transplant in Saudia Arabia and is well,two couldn,t manage to go abroad to have the transplant and died. Three cases of non syndromic BDP and two children with Alagille syndrome are well.

BDP constituted (4%) of all cases presenting with NC. A diagnosis of BDP would have been missed if a liver biopsy had not been performed. As presentation may be similar to extrahepatic biliary atresia and HIDA scan can show no excretion, such infants may be subjected to a laparotomy for a peroperative cholangiogram. Peroperative cholangiogram has been reported to show findings consistent with extra hepatic biliary atresia in upto a third of these patients and a Kasai portoenterostomy in such cases carries a poor prognosis necessitating liver transplantation(4).

It is, therefore, essential that infants with NC undergo a percutaneous liver biopsy and if bile duct paucity is identified a laparotomy can be avoided. It is imperative that a liver biopsy is performed urgently because if the biopsy reveals extra hepatic biliary atresia or is equivocal, a laparotomy for a peroperative cholangiogram should be offered without delay(5,6). As worsening of liver function is possible in patients with BDP at a later age (more likely in non syndromic BDP compared with Alagille syndrome), prolonged follow up is required.

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