

Clinical features

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About one-third of patients are jaundiced at birth; in all affected babies, jaundice is present by the end of the first week and deepens progressively. The meconium may be a little bile-stained, but later the stools are pale and the urine is dark. Pruritus is severe. Clubbing and skin xanthomas, probably related to raised serum cholesterol, may be present. Prolonged steatorrhea gives rise to osteomalacia (biliary rickets). Liver function tests show an obstructive pattern with elevated bilirubin and alkaline phosphatase (ALP). Associated anomalies occur in about 20% of cases and include cardiac lesions, polysplenia, situs inversus, absent vena cava and preduodenal portal vein. Biliary atresia may be suspected prenatally, when a cystic structure is observed in the porta hepatis on fetal USG.

Patent Atretic Atretic I IIa Figure 71.23 Classification of biliary atresia. Gallbladder filling provides a clue to the type of atresia.

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