

# EXTRAHEPATIC BILIARY ATRESIA Aetiology and physiology

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Biliary atresia is present in approximately 1 in 12 000 live births and affects males and females equally. The aetiology is unclear; the extrahepatic bile ducts are progressively destroyed by an inflammatory process that starts around the time of birth. Intrahepatic changes also occur and eventually result in biliary cirrhosis and portal hypertension. Untreated, death from the consequences of liver failure occurs before the age of 3 years. The Japanese and Anglo-Saxon classification describes three main types (Kasai) (Figure 71.23): type I: atresia restricted to the CBD; type II: atresia of the common hepatic duct; type III: a patent gallbladder and a patent CBD are present; type IV: the gallbladder, cystic duct and CBD are also obliterated; type V: atresia of the right and left hepatic ducts and the entire extrahepatic biliary tree. Hats worn by the people of Phrygia, an ancient country in Asia Minor; they resemble the liberté cap of the French Revolution. Pablo Luis Mirizzi, 1893–1964, surgeon, Córdoba, Argentina. Morio Kasai, 1922–2008, Professor of Surgery, Tokyo University, Tokyo, Japan. -).

RP RP Figure 71.21 Patterns of cystic duct anatomy. Note segment VI drain

age into the cystic duct and drainage of the right posterior

# sectorial duct (RP) into the neck of the gallbladder or an accessory duct (duct of Luschka). Figure 71.22 Magnetic resonance cholangiopancreatography demon

strating low insertion of the cystic duct (thick arrow) into the common bile duct (thin arrow).

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