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Congenital choledocal cyst: a rare anomaly

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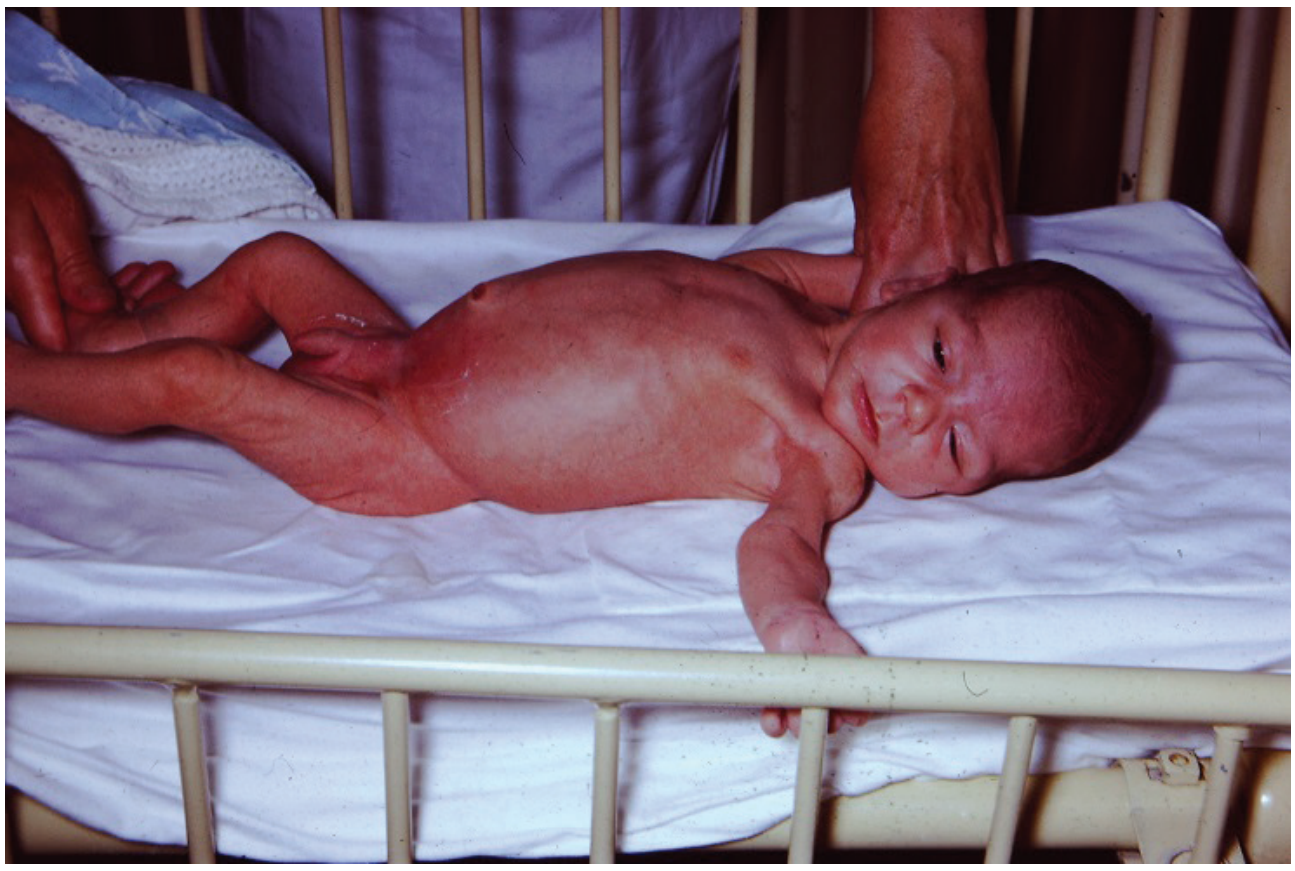

Fig 1: Baby AF aged 9 days (with permission)

During thirty years working in the newborn field I only encountered one case of this congenital anomaly:

CASE HISTORY

The mother was a health woman aged 25 with two children. The present pregnancy was uneventful. She went into labour spontaneously at 37 weeks gestation and had a normal delivery of a male infant weighing 3.2kg (head circumference 34cms and length 54cms). Examination at birth revealed no abnormality.

During the first week of life the baby became moderately jaundiced (maximum serum indirect-acting bilirubin 16mg%; direct-acting serum bilirubin < 1mg%). The jaundice was diagnosed as being due to AO incompatibility. At the age of 9 days, the infant commenced to vomit feeds and stopped passing stools. His upper abdomen appeared slightly distended and a soft lump was felt in the hypochondrium (Fig 1). Radiological studies including a barium meal and cholecystography revealed the presence of a choledocal cyst the size of a tangerine within the curve of the duodenum, laparotomy was undertaken by Mr. Arthur McPhearson, FRCS, (Fig 2).

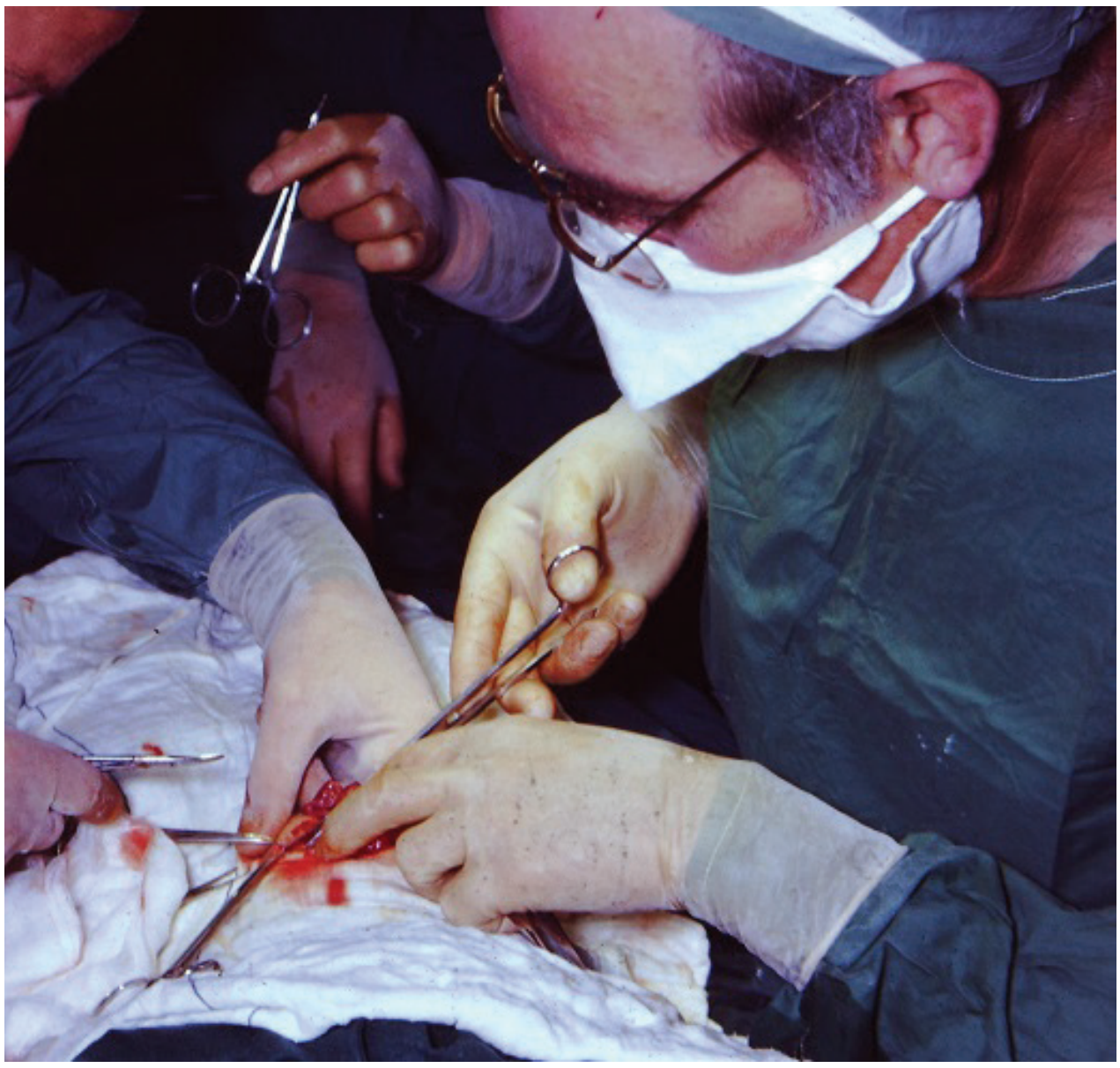

Fig 2: Mr. Arthur McPhearson at laparotomy

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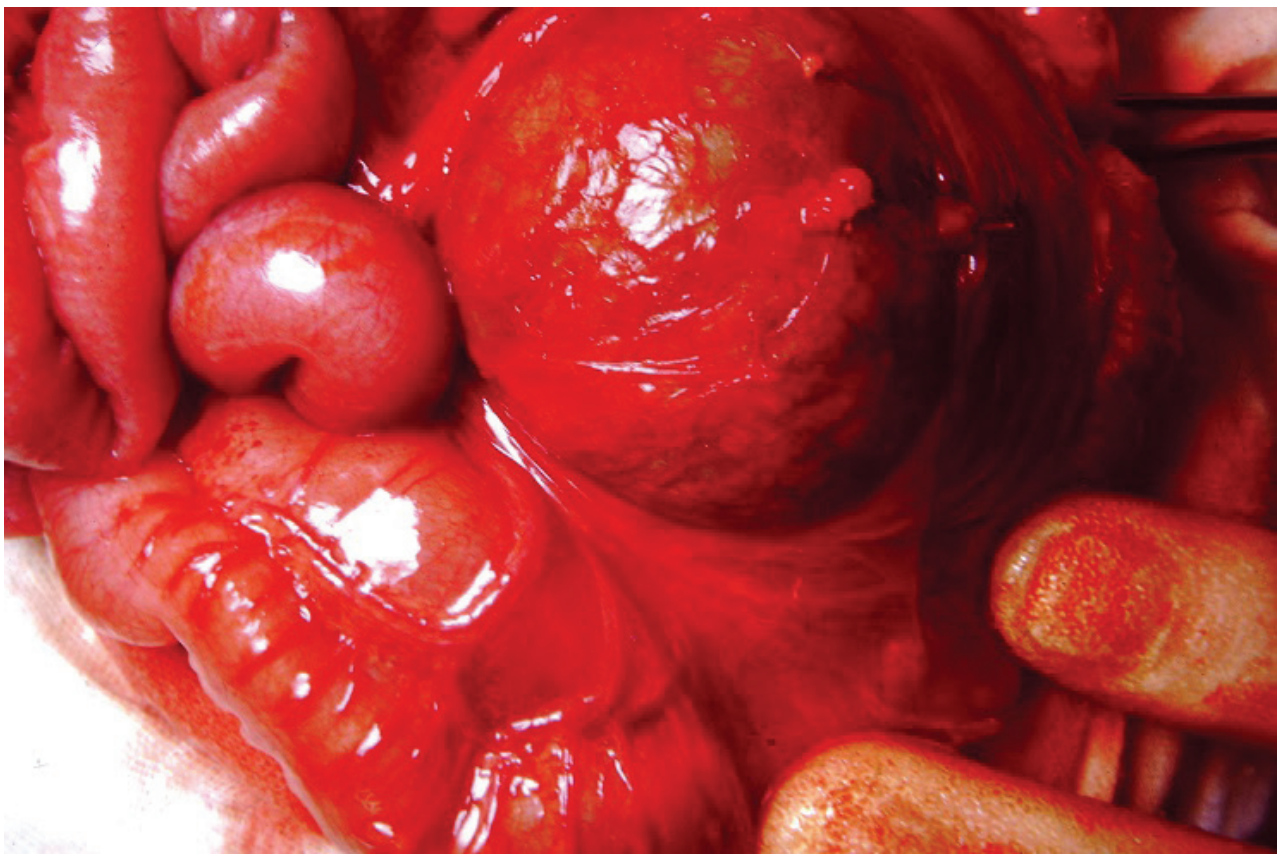

Fig 3: Baby AF's choledocal cyst exposed at laparotomy

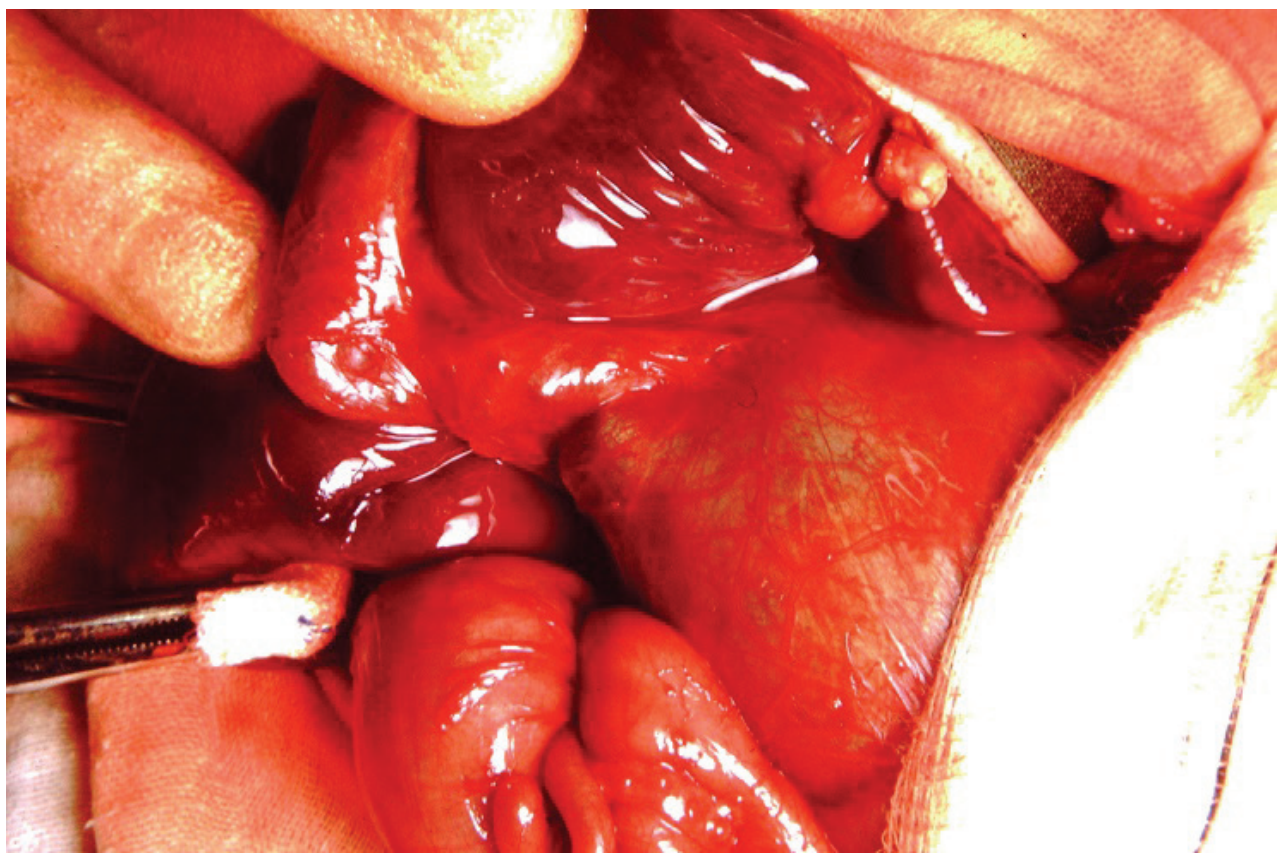

Fig 4: Choledocal cyst decompressed and anastomosed to duodenum

The cyst was identified (Fig 3), decompressed and then anastomosed to the duodenum (Fig 4). Post recovery was uneventful. The jaundice faded, there was no more vomiting and the infant started to pass pale yellow stools.

Follow up was without complication. Growth and development when last seen at the age of 3½ years was normal (weight 19.05 kg; head circumference 52cms and length 105cms) (Fig 5).

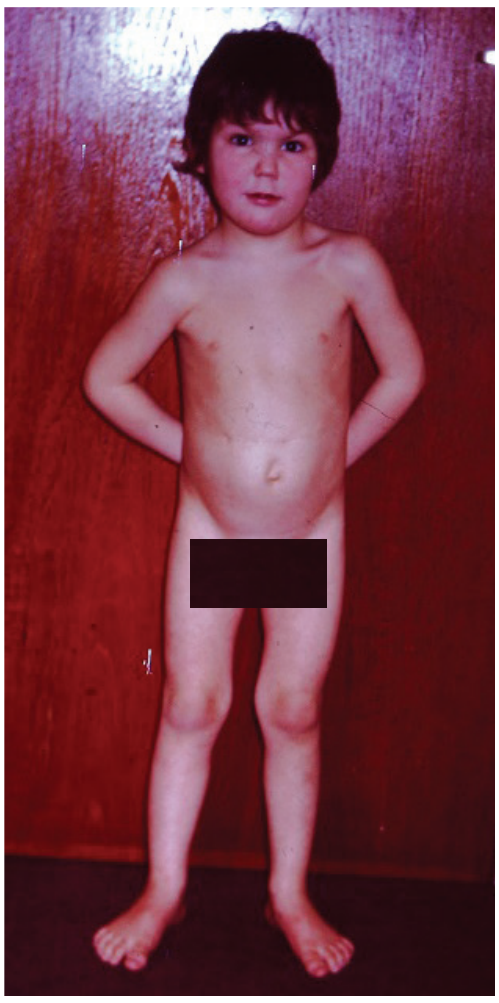

Fig 5:
AF at 3½ years (with permission)

COMMENTARY

The cause of this anomaly is atresia or stenosis of the terminal bile duct leading to a sharply demarked cyst of the common bile duct and moderate dilation of the proximal bile ducts as in the present case. The incidence of the condition is reported to be 1 in 250,000 with a male to female ratio of 1 to 4. The great majority of cases do well following cholecysto-duodenostomy, though cholangitis may occur. Typically such infants demonstrate obstructive jaundice; its absence in the present case may be explained by the early operative intervention.