reliable cord blood samples for CKBB analysis the cord must be clamped in two places about 20 cm apart at delivery, and that by careful venepuncture only the first few millilitres of blood should be used for CKBB estimation.

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References


Obstructive jaundice secondary to chronic midgut volvulus

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SUMMARY A case of progressive extrahepatic biliary obstruction due to chronic midgut volvulus secondary to malrotation in a 5-month-old girl is presented. The obstruction to the bile duct was relieved after correction of the malrotation and division of the obstructing bands.

Malrotation with associated Ladd’s bands is a common cause of extrinsic obstruction to the second part of the duodenum in infancy and early childhood. This results in bilious vomiting which is usually the initial and may be the sole presenting feature of the anomaly. The jaundice that has been noted in association with malrotation is invariably described as a consequence of intestinal gangrene secondary to midgut volvulus. Mechanical obstruction to the extrahepatic biliary tract secondary to malrotation has not been previously reported.

Case report

A 5-month-old girl was referred for investigation of failure to thrive, persistent diarrhoea, and progressive jaundice. She was the first-born infant after a normal pregnancy and delivery, and weighed 3340 g. At age 2 days she vomited bile-stained material but a straight x-ray film of the abdomen was normal. The vomiting continued for a further 3 days but contained only milk. After discharge from the maternity unit she continued to feed poorly and vomit intermittently. At 3 months she developed explosive diarrhoea which persisted in hospital despite a variety of diets. A barium meal showed gross gastro-oesophageal reflux but was otherwise interpreted as normal. Jaundice was first noted at 5 months and serial liver function tests showed a progressive conjugated hyperbilirubinaemia.

Weight on admission was 5300 g at 5 months (<3rd centile). Abdominal examination showed a 2 cm globular-shaped cystic mass in the right hypochondrium and a 2 cm palpable hepatomegaly. Laboratory investigations showed a total bilirubin of 199 µmol/l (11.6 mg/100 ml), alkaline phosphatase 293 KA units/100 ml, AST 400 IU/100 ml, total protein 400 g/l (albumin 222 g/l), alpha-1-antitrypsin activity 3-2 (normal range 2-0-4-0) g/l, and a sweat sodium concentration of 10 mmol/l.

Ultrasound scan of the liver showed grossly dilated intra- and extra-hepatic bile ducts, and the common bile duct was grossly dilated proximally but distally narrowed to a fine stricture; the gallbladder was distended and contained 'sludge'.

Laparotomy via a right upper quadrant transverse muscle-cutting incision revealed a tense distended gallbladder and common bile duct to a maximum diameter of 1·5 cm. Cholangiogram via the gall-
bladder confirmed the gross dilatation of the intra- and extra-hepatic bile ducts. The distal portion of the common duct was narrow and tortuous (Fig. 1). Further exploration through an extended incision showed a midgut volvulus with a 360° twist. The mesenteric veins and lymphatics were distended but the intestine was normal. After reduction of the volvulus, Ladd’s bands could be identified crossing the second part of the duodenum, which was rotated to the right, and compressing the distal 2–3 cm of the common bile duct (Fig. 2). Proximal to this area of constriction the common duct was dilated to 1.5 cm in diameter. The compressing bands were divided allowing the common duct to dilate and permitting free drainage into the duodenum. The malrotation was corrected by a standard Ladd’s procedure. Repeat cholangiogram showed free passage of contrast into the duodenum.

The postoperative course was uneventful. There was a strikingly rapid resolution of the jaundice and within 2 weeks all liver function tests had reverted to normal. Histopathological examination of a wedge biopsy of the liver showed features consistent with extrahepatic biliary obstruction with bile stasis and increased periportal fibrosis. Review of the barium meal performed at 3 months showed non-rotation of the duodenal loop.

Discussion

Obstructive jaundice of the unconjugated type in infancy demands urgent investigation and treatment if irreversible liver damage is to be prevented. Biliary atresia and choledochal cysts are the most common ‘surgical’ causes of jaundice in this age group. Biliary obstruction in association with duodenal malformations⁴ and secondary to pancreatic haemangioendothelioma, hydronephrosis, and tumours⁵ has been described, but jaundice secondary to Ladd’s bands and chronic midgut volvulus has not been reported previously.

References

Hyponatraemia caused by repeated cerebrospinal fluid drainage in post haemorrhagic hydrocephalus

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SUMMARY Three hyponatraemic infants had post-haemorrhagic hydrocephalus which was treated by repeated drainage of cerebrospinal fluid. Each required oral sodium supplements. The concentration of sodium in the cerebrospinal fluid was such that as much as 3 mmol of sodium could have been removed with each ventricular tap. Serum sodium concentrations should be monitored closely in any infant requiring regular drainage of cerebrospinal fluid.

Periventricular haemorrhage is common in infants of very low birthweight. Hydrocephalus is a serious complication in many survivors. It is now possible to diagnose both these conditions early by the use of real-time ultrasound scanning via the anterior fontanelle. We report 3 patients whose post-haemorrhagic hydrocephalus was treated initially by repeated drainage of cerebrospinal fluid. Each developed hyponatraemia which was treated with oral sodium supplements.

Patients

Case 1. A boy weighing 1.25 kg was delivered at 28 weeks' gestation. He developed severe hyaline membrane disease and ventilatory support was required for the first 8 days of life. During this time the use of real-time ultrasound scanning enabled bilateral intraventricular haemorrhages to be detected. Progressive communicating hydrocephalus developed and after a total of 10 therapeutic lumbar punctures had been performed a ventriculoperitoneal shunt was inserted on day 37.

Between days 19 and 29, nine therapeutic lumbar punctures were performed (mean volume 22 ml). During the same period the serum sodium concentration declined from 133 to 122 mmol/l. Throughout this time the patient was fully enterally fed on expressed breast milk. There was no gastrointestinal upset and no diuretics were administered. During the next 4 days 3.5 mmol/kg a day of supplementary oral sodium were given and the serum sodium concentration rose to 140 mmol/l.

Case 2. A boy weighing 1.070 kg was delivered at 27 weeks' gestation. He developed signs of persistent fetal circulation and required ventilation for the first 3 days of life. Bilateral intraventricular haemorrhages were detected using real-time ultrasound scanning. Progressive non-communicating hydrocephalus developed and after a total of 16 therapeutic ventricular taps a ventriculoperitoneal shunt was inserted on day 36.

Before his first ventricular tap the patient was on full intravenous feeding. His serum sodium concentration was normal on a sodium intake of 4 mmol/kg a day. Four ventricular taps were performed during the next 5 days (mean volume 15.5 ml). His serum sodium concentration fell to 120 mmol/l, despite increasing his sodium intake to 8 mmol/kg a day. During the next 5 days daily ventricular taps (mean volume 18 ml) continued and the patient was fully enterally fed on expressed breast milk, with the addition of 23 mmol/kg a day of oral sodium supplements. The serum sodium concentration rose to 153 mmol/l. Sodium supplements were therefore stopped. Daily ventricular taps continued and 4 days later the sodium value had fallen progressively to 126 mmol/l. Throughout this time there was no gastrointestinal upset and the patient was not on diuretics.

Case 3. A boy weighing 1.48 kg was delivered at 30 weeks' gestation. Severe hyaline membrane disease developed and ventilatory support was required for the first 8 days of life. A unilateral intraventricular haemorrhage was detected using real-time ultrasound scanning. Progressive communicating hydrocephalus developed which was initially treated by lumbar puncture and later by the insertion of a ventriculoperitoneal shunt.