Short reports

Percutaneous transhepatic occlusion for bleeding oesophageal varices in polycystic disease

I A LAING, T A S BUIST, AND M S FRASER
Royal Hospital for Sick Children and Royal Infirmary, Edinburgh, and Victoria Hospital, Kirkcaldy, Fife

SUMMARY A 7-year-old boy with congenital polycystic disease of the kidneys and liver developed portal hypertension and gastro-oesophageal varices. After two episodes of upper gastrointestinal bleeding, percutaneous transhepatic occlusion of varices and of the left gastric vein was carried out. During the next year there was no evidence of further haemorrhage.

Polycystic deformity of the liver is present in up to one-third of patients with polycystic kidneys. Nevertheless, functional impairment of the liver is rare. If portal hypertension and gastro-oesophageal varices occur, the major threat to life is from upper gastrointestinal bleeding. Transfusion of blood, the use of intravenous pitressin, and compression with a double-balloon gastro-oesophageal catheter are the most important measures for the control of acute bleeding. Prophylactic porto-systemic shunting may be done as an elective procedure, but this is technically difficult in children because of the small calibre of vessels. Embolisation of the varices and of the principal supplying veins using the percutaneous transhepatic approach is a fairly simple, and less invasive technique and one which may delay the need for major surgery until the child has grown.

Case report

The patient was the result of the third pregnancy of a 24-year old woman. She had previously had a stillborn child at 28 weeks' gestation: no necropsy was carried out. She also has a normal son. There was no family history of liver or kidney disease. At his neonatal examination the patient was noted to have a palpable mass in either flank. Full blood count, urea, and electrolytes were normal. A sample of urine contained no pus cells, and there was no growth on culture. An intravenous pyelogram showed that both kidneys were enlarged, and the characteristic lobulated appearance confirmed the clinical diagnosis of polycystic disease.

Subsequent follow-up showed the boy to grow poorly, his height and weight being about 3 standard deviations below the mean for his age. He looked emaciated, and was also noticed to develop hepatosplenomegaly in the early months of life. His glomerular function remained satisfactory. At age 5 years gastro-oesophageal varices were present on barium swallow examination. The percutaneous splenic venogram carried out at that time is shown in Fig. 1. It shows a greatly enlarged left gastric

Fig. 1 Percutaneous splenic venogram shows flow to gastro-oesophageal varices from the greatly enlarged left gastric (coronary) vein.

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vein which supplies dilated varices in the stomach and lower oesophagus. Porto-systemic shunting was considered but was not carried out because the vessels were small.

At age 7 years the patient had a ‘coffee-ground’ vomit. Six weeks later he again vomited blood, and was admitted to hospital. On admission he was shocked and his haemoglobin level was 4.4 g/dl. After blood transfusion his condition improved and he was transferred to the Royal Hospital for Sick Children where he required further blood transfusion and intravenous pitressin (0.3 IU/kg over 30 min) to control haemorrhage. After 24 hours, endoscopy was carried out under general anaesthesia. This confirmed the presence of large gastro-oesophageal varices, and a presumptive area of bleeding from a varix was found in the proximal part of the gastric antrum. Immediately after endoscopy, percutaneous transhepatic portography and selective variceal and left gastric embolisation were carried out (by TASB).

A catheter was introduced percutaneously through the liver into the portal vein. It was then advanced into the splenic vein. The pressure in this position was 405 mm of saline, indicating a severe degree of portal hypertension. Contrast was injected and this gave much the same appearance as in the previous portogram (Fig. 1) with the greatly enlarged left gastro-oesophageal varices. The catheter was then positioned in the left gastric vein and progressive occlusion of the variceal bed carried out using fragments of gelatin sponge soaked in sodium tetradecyl sulphate. After this, the left gastric vein was completely occluded by wire coils with attached Dacron threads (Fig. 2). As the catheter was withdrawn through the liver, the track was also embolised with fragments of gelatin sponge.

After this procedure, prophylactic cimetidine was given for 6 weeks. The only complication noted was further enlargement of the spleen (by about 3 cm) during the week after embolisation. He remained in hospital for a further 3 weeks, and during the year since discharge there has been no evidence of further gastrointestinal bleeding. He has had a degree of hypersplenism requiring blood transfusion, but faecal occult blood has always been negative. Mild hypertension has been carefully controlled with propranolol.

Discussion

Although polycystic disease of the liver is commonly found in association with polycystic kidneys, it is rare for hepatic dysfunction to develop. Of 46 cases of polycystic disease of the kidney which came to necropsy at the Mayo Clinic, in 15 there was associated polycystic malformation of the liver. However, of the 207 patients with polycystic kidneys seen by the same workers between 1932 and 1947, none had symptoms referable to liver dysfunction. Bradford et al. described 2 children with polycystic disease of kidney and liver complicated by portal hypertension, and found 13 other reported cases. The aetiology of portal hypertension in these patients is unknown. While cystic malformation of the liver generally does not appear to disturb the structure of the hepatic lobule, both Parker and Kerr et al. described patients in whom biopsy showed replacement of normal architecture by portal connective tissue forming dense fibrous bands. It is suggested that these changes may cause intrahepatic portal venous compression. There is evidence that propranolol can be used to decrease portal venous pressure, and its use in this patient was a bonus while his systemic pressure was being controlled.

Our 7-year-old patient is small for his age and it was thought that shunt surgery would be very difficult to perform because of his small vessels and there would be an increased risk of subsequent thrombosis at the anastomotic site. In this case it appears that percutaneous transhepatic embolisation

Fig. 2 Arrow indicates site of complete occlusion confirmed by injection of contrast into left gastric vein via transhepatic catheter.
of varices has been a successful temporary measure which may allow the patient to grow before definitive surgery is carried out.

The technique was first described by Lunderquist and Vang in 1974,\(^2\) and has been used quite extensively in adults, but to our knowledge this is the youngest child to be successfully managed in this way.

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References


Correspondence to Dr I Laing, Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF.

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Benign paroxysmal torticollis in infancy

THIERRY DEONNA AND DOROTHEA MARTIN

Médecin-associé, Service de Pédiatrie (Unité de Neuropédiatrie), CHUV, Lausanne, and Basler Kinderspital (Neurologische Abteilung), Basel, Switzerland

SUMMARY Of 5 infants with benign paroxysmal torticollis, 3 had symptoms of infantile migraine at the same time.

Paroxysmal torticollis (PT) in infancy is a self limited and benign entity, and was first described by Snyder in 1969.\(^1\) It is characterised by recurrent episodes of head tilt (Figure), sometimes accompanied by vomiting, pallor, agitation, and ataxia which subside spontaneously within a few hours or days and entirely disappear within a matter of months or years. It is not widely known and is rarely reported, although diagnostic problems with more serious causes of abnormal head and trunk posture or intermittent dysequilibrium in young children are encountered.

Recent observations of familial occurrence of

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Figure (Case 5.) Head tilt in benign paroxysmal torticollis at ages 4 and 18 months.