Infra-diaphragmatic total anomalous pulmonary venous drainage presenting with rectal bleeding

A C ELIAS-JONES AND S V CORDNER

Department of Paediatrics and Department of Forensic Medicine, Guy's Hospital, London

SUMMARY A boy aged 1 month presented with profuse rectal bleeding. Chest *x*-ray film showed pulmonary oedema and at cardiac catheterisation infradiaphragmatic total anomalous pulmonary venous drainage to a dilated portal venous system was found. Ulcerated oesophageal varices were identified at necropsy after unsuccessful cardiac surgery. The late and unique presentation of this case is emphasised.

Case report

A boy was born at 38 weeks' gestation by normal delivery. His birthweight was 2·65 kg. He breast fed well, was discharged home at 1 week of age, but subsequently showed poor weight gain. At 1 month of age he presented to the referring hospital with profuse rectal bleeding. He was pale, shocked, but not cyanosed. His respiratory rate was 60/min, heart rate 100/min, and blood pressure 80/40 mm Hg, with normal heart sounds. The liver was palpable 6 cm below the costal margin. There was ascites, but no splenomegaly.

The chest *x*-ray film showed pulmonary oedema (Fig. 1) and his electrocardiogram showed evidence of right ventricular hypertrophy. Other investigations showed: haemoglobin value 7·5 g/dl; white cell count $2{\cdot}4 \times 10^9$/l (2400/mm^3); platelets 38×10^9/l (38 000/mm^3); clotting studies normal; arterial gases in F_IO_2 0·21; Pao_2 9·4 mm Hg; $Paco_2$ 5·1 mm Hg; and pH 7·36. After treatment with diuretics and a blood transfusion he was transferred to this hospital.

He continued to bleed profusely from the rectum and fresh blood was aspirated from the stomach. An echocardiogram showed a dilated right ventricle and evidence of right to left atrial shunting. The site of drainage of the pulmonary veins was not identified.

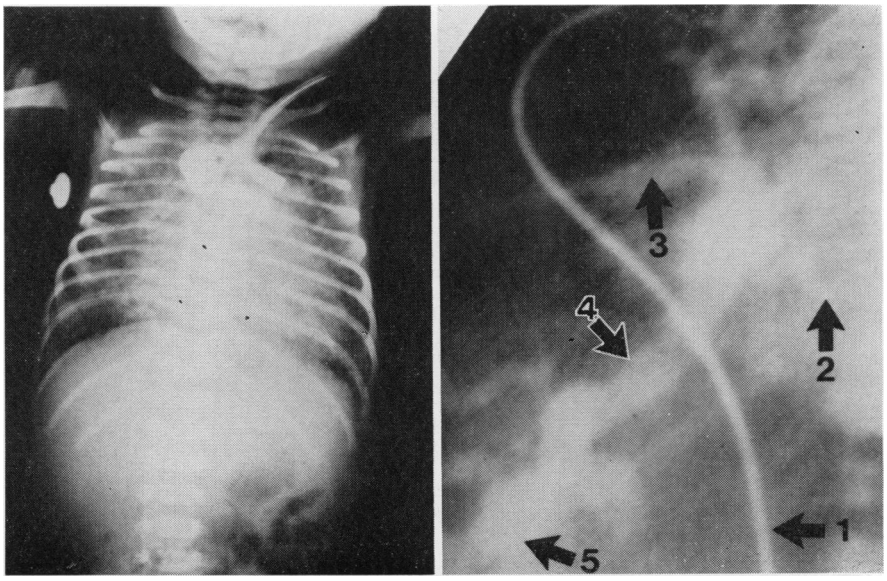

Fig. 1 *Left, chest x-ray film showing normal sized heart and pulmonary oedema. Right, lateral projection of the recirculation phase of a pulmonary arteriogram showing the catheter lying in the main pulmonary artery (1), left pulmonary vein (2), and right pulmonary vein (3) uniting to form common pulmonary vein (4) which descends below the diaphragm to anastomose with the hepatic portal venous system (5).*

An abdominal aortogram showed no bleeding point. He was treated with further transfusion, platelet concentrate, and intravenous cimetidine. Cardiac catheterisation showed a step up in oxygen saturation from the superior vena cava at 69% to the inferior vena cava at 85% and mid right atrium at 87%. Both ventricles had a systolic pressure of 90 mm Hg equal to systemic level and the main pulmonary artery pressure was 90/45 mm Hg. Pulmonary artery angiography confirmed the presence of total anomalous pulmonary venous drainage (TAPVD) of an infradiaphragmatic type with drainage to the portal vein region (Fig. 1).

At cardiac surgery the common pulmonary vein was ligated at the diaphragm and an anastomosis was made from this vein to the left atrial appendage. Despite intermittent positive pressure ventilation and intravenous dopamine and isoprenaline, he remained hypothermic and hypotensive and died 9 hours later.

Necropsy findings

At necropsy the lungs were congested and oedematous. There was a valvular foramen ovale 0·7 cm in diameter. The right ventricular wall was hypertrophied to 0·7 cm. The pulmonary artery was dilated and had a thickened wall. The left atrium and ventricle appeared small. The aorta was narrow, measuring 0·7 cm in diameter. The pulmonary veins from both lungs united to form a common pulmonary vein which lay behind and was anastomosed to the left atrium. There was a ligature around the common pulmonary vein, as it coursed beneath the diaphragm, giving off no branches, and entered the left lobe of the liver. The portal vein entered the porta hepatis normally. The ductus venosus was closed. Further dissection showed an intrahepatic anastomosis of the anomalous vein to the portal venous system through many separate venous channels. The gastrointestinal tract contained altered blood. There was a healing ulceration of the oesophagus 2·5 cm from the gastro-oesophageal junction, with prominent submucosal venous markings that were confirmed histologically to be oesophageal varices (Fig. 2). The spleen weighed 10 g.

Discussion

Rectal bleeding is a previously unreported presentation of infradiaphragmatic TAPVD. These patients characteristically present shortly after birth with cyanosis and heart failure resulting from either pulmonary venous obstruction of the pulmonary veins into the systemic venous system, or the drainage of pulmonary venous return through the liver after closure of the ductus venosus.

Our case presented late on the 28th day. In the series of Duff *et al.*[1] the mean age of onset of

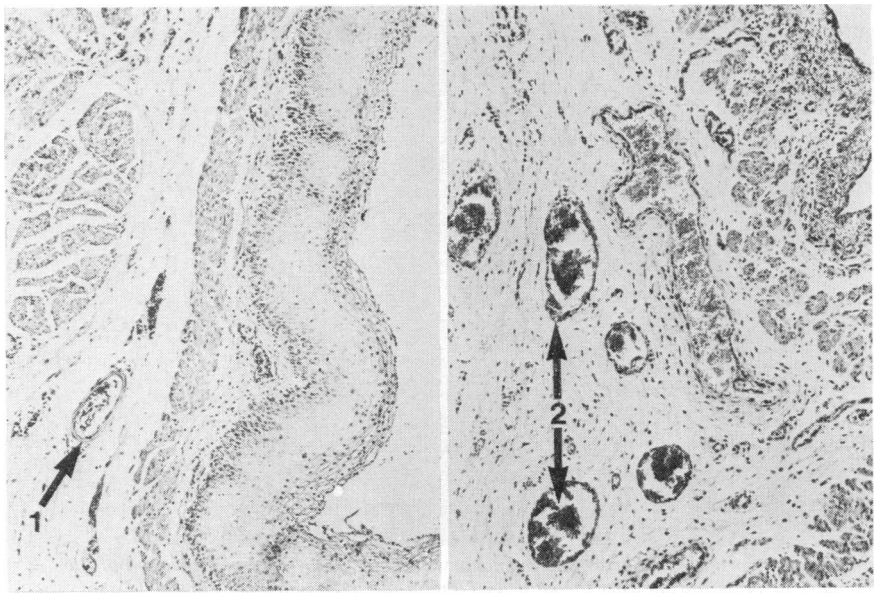

Fig. 2 *Left, histology of normal oesophagus of 1 month old infant showing normal size oesophageal vein (1), and right, the infant's oesophagus showing dilated oesophageal varices (2).*

symptoms was 3 days, with a mean delay until hospital admission of 11 days. In this patient the previous poor weight gain may have been due to pulmonary oedema secondary to partial venous obstruction. Despite the absence of splenomegaly, the presence of oesophageal varices and rectal bleeding also suggests that any obstruction to pulmonary venous return was shared by the portal venous system, presumably as a result of the anastomosis between these 2 venous systems in the liver.

Oesophageal varices caused by infradiaphragmatic TAPVD to the left gastric vein was reported by Laurence and Brown,[2] in a newborn infant who presented within 24 hours of birth with haematemesis of 'bright red arterialised blood'. This led the authors to suspect the cardiac diagnosis. In our case the characteristic chest *x*-ray film showing a normal sized heart with gross pulmonary oedema led to the clinical diagnosis.

The poor outcome in our case reflects that found by Duff *et al.*[1] in a series of 28 cases in which the overall mortality rate was 93%, with an operative mortality of 66% in 9 cases. Similarly Clarke *et al.*[3] had an operative mortality rate of 50% in 6 cases.

We thank Dr O G Brooke, St George's Hospital for referring this patient; Dr M C Joseph for permission to report the case; Miss R Green for secretarial help.

References

1 Duff DF, Nihill MR, McNamara DG. Infra-diaphragmatic total anomalous pulmonary venous return. *Br Heart J* 1977; **39**: 619–26.
2 Laurence KM, Brown RJK. Total anomalous drainage of pulmonary veins into the left gastric vein. *Br Heart J* 1969; **22**: 295–9.
3 Clarke DR, Stark J, De Leval M, Pincott JR, Taylor JFN. Total anomalous pulmonary venous drainage in infancy. *Br Heart J* 1977; **39**: 436–44.

Correspondence to Dr A Elias-Jones, Department of Paediatrics, Addenbrooke's Hospital, Hills Road, Cambridge CB2 2QQ.

Received 3 February 1983

Orofacial clefts and oesophageal atresia

A F AZMY, P A M RAINE, AND D G YOUNG

Department of Paediatric Surgery, Royal Hospital for Sick Children, Glasgow

SUMMARY Of 114 children with oesophageal atresia, 6 had cleft palate and other craniofacial anomalies present included cleft lip, micrognathia, hypertelorism, microcephaly, and hydrocephalus. The difficulties encountered in the management of patients in whom these conditions present together are emphasised with special reference to swallowing problems and recurrent chest aspiration.

Congenital malformations occur in 1% to 3% of live births but the probability of additional anomalies in a patient presenting with a major malformation is higher than this. Additional malformations increase the risk of morbidity, residual disability, and mortality.

The incidence of other malformations with oesophageal atresia (OA) and tracheo-oesophageal fistula (TOF) is high,[1] although reports show that their extent and effect on ultimate prognosis varies.[1 2] The association of cleft lip (CL) and cleft palate (CP) with OA and TOF has not, however, been emphasised and although both CP and OA are surgically correctable, problems with swallowing and recurrent chest aspiration caused by CP, pharyngeal incoordination, and oesophageal narrowing and dysmotility add to the difficulties in management.

Patients

Between 1972 and 1981, 114 patients were referred to the neonatal surgical unit of this hospital with OA. Six also had CP with or without CL and 4 of these survived and underwent corrective surgical procedures. The Table shows the clinical details of these 6 patients.

Discussion

Reviews of large series of children with malformations associated with OA mention the occasional occurrence of CP or CL, or both, without giving details of incidence.[3 4] The extensive survey by Holder *et al.*[1] of 1058 patients with OA gave an