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

Table Clinical details of patients with cleft palate and oesophageal atresia

Case No	Sex	Orofacial anomaly	Other anomalies	Surgical procedures	Outcome
1	M	Hard and soft CP; micrognathia		Ligation of TOF; primary oesophageal anastomosis; CP repair at 17 months; pharyngoplasty	Alive
2	F	Right complete CL and CP; hypoplasia middle third of face	Dislocated left shoulder	Ligation of TOF, oesophagostomy and gastrostomy; CL repair at 3 months; gastric tube oesophagoplasty at 1 year; CP repair at 18 months	Alive
3	F	Bilateral complete CL and CP	Hydrocephalus (Dandy Walker malformation)	None	Died
4	M	Cleft hard and soft palate	Pyloric stenosis	Ligation of TOF; primary oesophageal anastomosis; repeat oesophageal bougienage; CP repair at 18 months	Alive
5	F	Left complete CL and CP; absent columella and right nasal cavity; hypertelorism	Tetralogy of Fallot; solitary kidney; ureterocele (single ureter)	Ligation of TOF; primary oesophageal anastomosis; CP repair at 24 months	Alive
6	M	Hard and soft CP	Absent radii, thumbs; microcephaly; imperforate anus and rectovesical fistula; truncus arteriosus; horseshoe kidney; bilateral dislocation of hips; syndactyly of toes	None	Died

TOF = tracheal-oesophageal fistula; CP = cleft palate; CL = cleft lip.

incidence of associated CP or CL, or both, of 3.1% compared with 5.2% in this study.

The combination of limb, vertebral, anorectal, renal, and cardiac malformations in association with OA and TOF (Vater syndrome)⁵ suggest multifactorial damage during embryogenesis at a critical and early stage. Craniofacial malformations such as choanal atresia, macrostomia, and facial hypoplasia have been associated with OA.³ These malformations of face and oesophagus may be explained by disturbance of development of the embryo between the 5th and 10th weeks of intrauterine life. The patient in case 6 illustrates the possibility that a single insult in early fetal life may cause damage to all organ systems.

The possible presence of second and further hidden anomalies is always considered in a neonate with an obvious malformation and delay in diagnosis should therefore be minimised. The routine practice of attempted passage of a nasogastric tube after delivery led to rapid identification of OA and to measures to remove oropharyngeal secretions and obviate aspiration in 5 patients in this study: in the patient in which this was not done a milk feed caused immediate respiratory distress.

The 4 surviving patients each had considerable problems of repeated pulmonary aspiration and chest infections. While repaired OA may be complicated by aspiration pneumonia this is not a

consistent feature and, if persistent, is usually attributable to dysmotility or anastomotic stenosis. Only 1 patient required oesophagoscopy and bougienage but episodes of aspiration remained a problem after radiography had shown satisfactory oesophageal lumen and motility on barium swallow. Unless micrognathia is a notable feature, respiratory complications are usually minimal in the presence of CP—1 patient only had an obvious micrognathia.

Accumulation of pharyngeal secretions and subsequent lung aspirations may be avoided by frequent suction (depending on the rapidity of accumulation). When secretion accumulation is excessive continuous pharyngeal suction using a double lumen tube connected to a negative suction pump is effective. The infants are nursed better in an upright position in a special chair and physiotherapy and antibiotics should be repeated as necessary to treat episodes of chest infection. Feeding by nasogastric tube or via gastrostomy where applicable may allow pharyngeal incoordination or oesophageal dysmotility to improve and to maintain a satisfactory weight gain before re-establishing oral feeds.

We conclude that the 2 anomalies together lead to more chest complications than either alone and that their coincidence carries implications for morbidity and prognosis. This opinion is reinforced by the good progress and resolution of problems following repair of cleft palate in 3 patients.

References

- ¹ Holder TM, Cloud DT, Lewis JE, Jr, Pilling GP. Esophageal atresia and tracheo-esophageal fistula: a survey of its members by the surgical section of the American Academy of Pediatrics. *Pediatrics* 1964; **34**: 542-9.
- ² Waterston DJ, Bonham Carter RE, Aberdeen E. Oesophageal atresia: tracheo-oesophageal fistula. A study of survival in 218 infants. *Lancet* 1962; **i**: 819-22.
- ³ Brereton RJ. Skeletal anomalies in oesophageal atresia. *Z Kinderchirurgie* 1979; **26**: 258-70.
- ⁴ Cumming WA. Neonatal radiology: esophageal atresia and tracheo-esophageal fistula. *Radiol Clin N Amer* 1975; **13**: 227-95.
- ⁵ Quan L, Smith DW. The Vater association. *J Paediatr* 1973; **82**: 104-71.

Correspondence to Mr A Azmy, Royal Hospital for Sick Children, Yorkhill, Glasgow G3 8SJ

Received 7 April 1983

Disseminated arterial calcification associated with acardius acephalus

D ROYSTON AND F GEOGHEGAN

Department of Pathology, National Maternity Hospital, Holles Street, Dublin

SUMMARY Widespread arterial calcification was shown at necropsy in an infant who died at age 5 days and whose twin was acardiac. The changes resembled closely those described in idiopathic arterial calcification of infancy and the possible importance of haemodynamic factors in the production of these changes is discussed.

Case report

A 30 year old woman expecting twins, whose first pregnancy in 1975 had been completely normal, was admitted to this hospital because of ultrasound scan detection of a cystic malformation of 1 baby. Polyhydramnios was also present. There was no history of drug ingestion or smoking. Spontaneous premature labour began at 30 weeks' gestation and twin 1 was delivered by forceps because of cord prolapse at full dilatation. Delivery of twin 2 was by breech extraction under general anaesthesia and the acardiac fetus showed multiple subcutaneous fluid filled cysts, puncture of which was necessary to effect delivery. Twin 2 weighed 1940 g and displayed the typical features of acardius acephalus. The placenta was removed manually, weighed 920 g, and was bulky, pale, and fragmented and monoamniotic and monochorionic in type. The umbilical cord of twin 2 contained a single artery and vein and was inserted into the cord of twin 1 near its attachment to the placenta. The cord of twin 1 contained 2 arteries and 1 vein.

Twin 1 weighed 1500 g and had an Apgar score of 9 at 1 and 5 minutes. The baby was in excellent condition and did not require resuscitation. She was admitted to the intensive care unit because of

prematurity and remained well until 44 hours of age when her condition suddenly deteriorated and a clinical diagnosis of sepsis was made. Despite treatment she failed to improve and died at age 5 days. Cardiomegaly, hepatomegaly, and signs of neurological abnormality were present before death. Serum calcium values were normal and a metabolic and infection screen were negative.

Necropsy findings

Important gross findings at necropsy were confined to the heart, lungs, liver, brain, and blood vessels. The heart weighed 19.5 g (normal 10 g). Both ventricles were enlarged but no anatomical abnormalities were found. Both lungs were poorly aerated. The liver was firm and greenish brown in colour. Sections of the brain showed extensive bilateral intraventricular haemorrhage and kernicterus but there was no evidence of intracranial trauma. Patchy calcification was grossly evident in the pulmonary trunk, aortic arch, and in the thoracic and abdominal aorta. The major branches of the aorta were similarly affected.

Microscopic findings. The most striking findings were in the arteries. The elastic laminae of the aorta and pulmonary trunk showed patchy calcification and fragmentation. The media contained large focal calcium deposits that interrupted the elastic fibres (Fig. 1). In addition to the medial calcification, some large pulmonary branches showed focal intimal proliferation without appreciable obliteration of the lumen. The splenic artery showed, in addition to medial changes, an area of calcification along the internal elastic lamia but without intimal proliferation (Fig. 2). Intrathyroid and intrarenal arteries had similar