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Total anomalous pulmonary venous drainage to superior vena cava associated with preductal coarctation of aorta[1]

Successful correction in a 12-day-old infant

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Successful repair of total anomalous pulmonary venous drainage to the innominate vein, associated with persistent ductus arteriosus, coarctation of aorta, and hypoplasia of the aortic arch in a 12-day-old infant is reported. To our knowledge this is only the second successfully operated infant with this combination of lesions.

Total anomalous pulmonary venous drainage is one of the lesions commonly requiring correction early in infancy. The mortality rate is high, especially in the presence of pulmonary hypertension (Gomes *et al.*, 1970; Behrendt *et al.*, 1972; Gathman and Nadas, 1970). Additional cardiac anomalies are often present (Brody, 1942, 20%; Bonham-Carter, Capriles, and Noe, 1969, 22%) and further complicate the treatment. Preductal coarctation of the aorta, hypoplastic aortic arch, or aortic arch interruption are among the most serious associated lesions. To our knowledge total anomalous pulmonary venous drainage associated with one of these malformations has not been successfully corrected, except in an 8-day-old infant recently reported by Barratt-Boyes and colleagues (1972).

We report here a case of a 12-day-old infant who underwent successful one-stage correction of total anomalous pulmonary venous drainage to left superior vena cava, associated with persistent ductus arteriosus and coarctation of the aorta.

Case report

Our patient, a boy, was born after a normal pregnancy and normal delivery in March 1972. His birthweight was 2·6 kg. Soon after birth he became breathless and signs of cardiac failure were noted. A heart murmur was heard 5 days after birth. Digoxin and diuretics were started and the infant was referred to our unit for further investigation and treatment at the age of 12 days.

On examination the pulses were bounding in both brachial arteries but the femoral arteries were not palpable. Respirations were shallow, 65 a minute. The blood pressure was 135 mmHg systolic in both arms, 25 mmHg systolic in the legs. A systolic murmur grade 2/6 was heard along the left sternal edge. The second sound was split. The liver was enlarged 2 cm below the costal margin. There was no peripheral oedema. The electrocardiogram showed moderate right ventricular hypertrophy with P pulmonale. Haemoglobin, electrolytes, and blood urea were within normal limits. Chest *x*-ray showed a large superior mediastinum with congested lung fields.

The clinical diagnosis of total anomalous pulmonary venous drainage and coarctation of the aorta was made. The child was catheterized on the day of admission. Cardiac catheterization (Table) showed a step-up in oxygen saturation in the right side of the heart and a systolic gradient between femoral and brachial artery of 43 mmHg. Angiocardiography from the left and right ventricle confirmed the diagnosis of total anomalous pulmonary venous drainage to the left superior vena cava, persistent ductus arteriosus, and coarctation of the aorta. There was severe pulmonary hypertension with the pulmonary artery pressure at systemic level.

The child gradually deteriorated and was intubated and ventilated on the Engström ventilator and indwelling venous and arterial cannulae were introduced. An emergency operation was performed 7 hours after cardiac catheterization.

Under general anaesthesia the child was placed between ice bags and cooled to 29°C nasopharyngeal temperature. The heart was then exposed through a bilateral thoracotomy in the 4th intercostal space. The pericardium was tense and contained fresh blood. When the pericardium was opened the heart fibrillated, and defibrillation was not successful (nasopharyngeal tem-

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TABLE *Cardiac catheterization at age 12 days*

	Oxygen saturation (%)	*Pressure* (*mmHg*)
Left innominate vein	94	
Superior vena cava	69	
Right atrium	87	10 (mean)
Inferior vena cava	53	
Right ventricle	85	80/10*
Pulmonary artery	73	100/55*
Left atrium	78	10 (mean)
Left ventricle	70	100/18
Femoral artery	70	55/45

* Pressures not taken simultaneously.

perature 29°C). The ascending aorta was cannulated with a right-angled metal cannula and a single cannula was introduced into the right atrial appendage. Cardiopulmonary bypass using a disc oxygenator was established and the child was further cooled. The circulation was stopped at 20°C. The persistent ductus arteriosus was ligated and the coarctation of the aorta resected. An end-to-end anastomosis was performed between the junction of the left subclavian artery and the aortic arch to the descending aorta, using 6–0 black silk (Fig.). The vertical anomalous pulmonary vein was then ligated and the heart was lifted up. The left atrium was opened posteriorly, the atrial septal defect was closed, and an open anastomosis between the left atrium and confluence of pulmonary veins was performed. After 45 minutes of total circulatory arrest, air was evacuated from the heart and perfusion was restarted. The patient was rewarmed on cardiopulmonary bypass and when normal temperature was reached bypass was discontinued without difficulty. A left atrial line was introduced through the right upper pulmonary vein, two epicardial pacemaker wires were inserted, and the chest was closed in a routine manner. A tracheostomy was performed at the end of the procedure because a difficult and prolonged postoperative course was expected.

The patient was ventilated on an Engström ventilator, blood gases being checked at frequent intervals. Tracheal secretions were aspirated ¼ to ½ hourly and intensive physiotherapy given at 2-hourly intervals. Fluid intake was restricted to 6 ml 5 per cent dextrose per hour for the first 24 hours after operation. Small intravenous supplements of potassium were given and the patient was given digitalis. Diuretics were started because of considerable lung congestion. Ampicillin and cloxacillin were given as a routine postoperative cover.

The chest *x*-ray cleared after a few days, and apart from a transient metabolic alkalosis lasting about 36 hours, the postoperative course continued uneventfully. The patient was ventilated for 8 days, the tracheostomy tube was removed on the 17th day, and he was discharged from hospital on the 24th postoperative day.

At the time of discharge the patient was in sinus rhythm and was feeding well. Haemoglobin and electrolytes were within normal limits. Electroencephalogram did not show any specific changes compared with the preoperative recording. Blood pressure, measured by Doppler ultrasound, was 122 mmHg systolic in the arms and 92 mmHg systolic in the legs. This residual gradient of 30 mmHg was believed to be due to hypoplasia of the aortic arch.

At the last follow-up 12 months after operation he continued to do well. His weight was 7·77 kg and height 72·3 cm. There were no signs of heart failure and all drugs had been discontinued.

Discussion

The mortality rate of patients with total anomalous pulmonary venous drainage or preductal coarctation of aorta is fairly high during the first year of life (Keith, Rowe, and Vlad, 1967; Bonham-Carter *et al.*, 1969; Tawes *et al.*, 1969; Mortensen *et al.*,

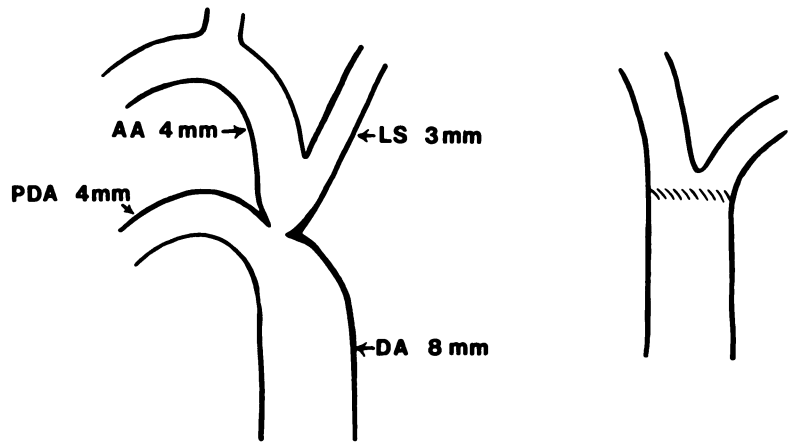

FIG. *AA, aortic arch; PDA, persistent ductus arteriosus; LS, left subclavian artery; and DA, descending aorta.*

1959; Sinha *et al.*, 1969). Early surgical treatment can offer better results, but the risk remains high (Cooley, Hallman, and Leachman, 1966; Thibert and Casasoprana, 1971; Behrendt *et al.*, 1972; Tawes *et al.*, 1969; Sinha *et al.*, 1969). The recent improvement in surgical treatment of congenital heart disease in early infancy cannot be ascribed to one single factor. Excellent results have been obtained by the method of deep hypothermia and circulatory arrest (Hikasa *et al.*, 1967; Barratt-Boyes, Simpson, and Neutze, 1971; Stewart *et al.*, 1973) but also with the use of cardiopulmonary bypass (Malm *et al.*, 1970; de Leval *et al.*, 1973). However, for the complicated lesions like the one described here, circulatory arrest under deep hypothermia undoubtedly offers better operative conditions and a better chance for survival. The other important factors are, in our opinion, early and complete cardiac investigation, intensive pre- and postoperative care, which includes monitoring of acid-base balance, blood pressure, central venous pressure, left atrial pressure, and serum and urinary electrolytes. Intensive respiratory care is very important. We prefer to ventilate every infant after open heart surgery at least for a few hours. Earlier in our experience this was achieved via tracheostomy which was recently replaced by nasotracheal intubation. Tracheostomy is thus reserved only for patients requiring long-term respiratory support or in whom the aspiration of secretions becomes difficult. Currently we would probably not do a tracheostomy on this patient at the end of the operation but would probably try to ventilate for a few days via nasotracheal tube.

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