Type 3c truncus arteriosus

Case report with clinical and surgical implications

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An infant with type 3c truncus arteriosus of Collett and Edwards is reported. In this anomaly, the left pulmonary artery arises from the left lateral wall of the truncus and the right pulmonary artery originates from a right-sided ductus arteriosus. Only two reports of this malformation have been described previously. Precise anatomical diagnosis is of importance since some of these patients may have a small persistent ductus arteriosus and right pulmonary artery, so that surgical correction is not feasible. This rare anomaly is discussed and comment made on the surgical and clinical implications.

In truncus arteriosus a single great vessel arises from the base of the heart and supplies the coronary, pulmonary, and systemic circulation via the main vessels of these circulations (Collett and Edwards, 1949; van Praagh and van Praagh, 1965).

Collett and Edwards in 1949 classified truncus arteriosus into different major types and minor subsets. Type 3 truncus arteriosus was defined as a truncus arteriosus with one or both pulmonary arteries arising independently from either side of the truncus. Type 3 was further subdivided into 5 different subtypes.

In subtype 3a the truncal arch is left arching and pulmonary arteries arise from the right and left lateral walls of the truncus. In type 3b, also with a left arch, the right pulmonary artery arises from the right side of the truncus and left pulmonary artery from a normal persistent ductus arteriosus. Type 3c is a truncus with a left arch in which the left pulmonary artery arises from the left lateral wall of the truncus and the right pulmonary artery originates from a right-sided ductus arteriosus. Subtypes 3d and 3e are the mirror images of types 3a and 3b respectively.

Embryological implications

Subtype 3b differs from subtype 3c in that in the latter only the left main 6th arch is present: the left main pulmonary artery arises from the common trunk and supplies the left lung. The right lung is supplied by way of a ductus arteriosus, i.e. on the right side, the 6th primitive arch is poorly developed, and the 4th primitive arterial arch is well developed.

Subtype 3b represents an example of a mirror image embryological aberration.

Previously reported cases

We are able to find four previous reports of subtype 3b (van Praagh and van Praagh, 1965; Pott, 1879; Dickson and Fraser, 1914; McNamara and Sommerville, 1968) and two of type 3c truncus arteriosus (Shapiro, 1930; McNamara and Sommerville, 1968) (Table). In none was the diagnosis made during life.

Case report

A male infant was born after a normal pregnancy and delivery. The baby was admitted to the cardiac unit aged 8 days with mild tachypnoea and respiratory distress with rib recession. Moderate generalized cyanosis without clubbing was noted. The peripheral pulses were equal and of full volume in the arms and legs. The apex beat was localized in the 5th interspace just outside the midclavicular line and a right ventricular heave was palpable along the left sternal border. The first heart sound was normal and the second sound was single. No murmurs were heard. The infant had a weak hoarse cry. The liver was enlarged 2 cm below the costal margin.

The electrocardiogram showed right axis deviation and right ventricular hypertrophy and x-ray of the chest showed cardiomegaly, pulmonary plethora, right ventricular enlargement, and a right diaphragmatic hernia. Left and right heart cardiac catheterization was performed on the 10th day. Truncal saturation was 73 per cent and the angiographic findings are shown in Fig. 1a. Fig. 1b is a schematic diagram of the anatomical features of type 3c truncus arteriosus.

FIG. 1 a) Selective left ventriculogram in the anteroposterior and lateral projections. A ventricular septal defect is present; the common trunk arises above this. The left main pulmonary artery arises from the main stem of the common trunk while the right 'main pulmonary artery' arises from the right common carotid artery and right-sided persistent ductus arteriosus. b) Schematic diagram of the anatomical features of type 3c truncus arteriosus.

Surgery was not considered possible and the infant was discharged aged 12 days to continue with antifailure therapy. Death occurred suddenly at 18 days of age and the anatomical findings of the heart and vessels are shown in Fig. 2.

Surgical implications

Truncus arteriosus became a correctable cardiac malformation in 1967 when McGoon and others from the Mayo Clinic reported an operation for the correction of this anomaly (McGoon, Rastelli, and Ongley, 1968). Since then the lesion has been corrected in many other centres (Wallace *et al.*, 1969; Gomes and McGoon, 1971; McGoon, Wallace, and Danielson, 1972; Barratt-Boyes *et al.*, 1972; Rogers, Winship, and Coleman, 1971).

Prerequisites for the repair of truncus arteriosus are two adequate pulmonary arteries which will be able to take over the output of the right ventricle and the absence of obstructive pulmonary vascular disease. It is conceivable that types 3a and d may be surgically correctable, but in types b and c the size of the ductus arteriosus will determine whether surgical correction is feasible or not. Certain patients with type 3c truncus arteriosus may have a right ductus arteriosus and pulmonary artery of adequate size so that anastomosis of this vessel to a newly created pulmonary trunk may be possible. In type 3c truncus arteriosus, the left lung will bear the brunt of the initial flow of blood with the development of increasing changes in the arterioles of the lung. The right lung, however, because of the narrow and long arterial vessel supplying it, may conceivably be protected from the high pressure-flow dynamics of this anomaly for a much longer time. From previous descriptions of this rare form of truncus arteriosus and our own patient (3 patients in all), the right pulmonary artery is small, hypoplastic, and underdeveloped and makes surgery probably impossible even with presently refined techniques. Barratt-Boyes has indicated that if the ductus arteriosus or pulmonary artery measures more than 2 mm in diameter, surgical

TABLE Cases with type 3c truncus arteriosus

Author	Year	Aortic arch	PDA	LPA origin
Shapiro	1930	Left	R	Truncus
McNamara and Sommerville	1968	Left	R	Truncus
This patient	1973	Left	R	Truncus

FIG. 2 A view of the heart and vessels from the front showing the features typical of type 3c truncus arteriosus.

correction may be feasible (B. G. Barratt-Boyes, 1973, personal communication).

The mortality rate for the repair of truncus arteriosus from the Mayo Clinic group beyond 5 years of age is low (10%) but below this age, the mortality is greater than 50 per cent. Up to 1971 no infants under the age of 1 year had been submitted to repair of truncus arteriosus in their unit (McGoon and Wallace, 1972). Barratt-Boyes has undertaken surgical correction of truncus arteriosus in 3 young infants, with one survivor (B. G. Barratt-Boyes, 1973, personal communication).

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