CASE REPORTS

Persistent Truncus Arteriosus with Survival to the Age of 38 Years

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The prognosis in persistent truncus arteriosus is very poor: the median age of survival of the 94 patients with the disease reported up to 1962 was only five weeks (Fontana and Edwards, 1962). Exceptionally, patients suffering from this disease live into the fourth decade. The longest survival reported is the case of the man described by Carr, Goodale, and Rockwell (1935) who lived to the age of 36 years and 2 months. We report here the case of a woman with persistent truncus who died at the age of 38 years; the anatomical changes in the pulmonary vasculature, which we believe account in the first place for her comparative longevity and ultimately for her death, are considered.

Case Report

A housewife, aged 38 years, was admitted to hospital in congestive cardiac failure of one month's duration with complaints of a choking sensation when lying flat. She had been cyanosed from birth but had led a normal life playing games at school and later doing housework without dyspnoea. At the age of 22 years she gave birth to a daughter, the pregnancy and labour being uneventful.

On examination she was spare and deeply cyanosed but was not dyspnoeic at rest. There was a striking increase in the jugular venous pressure and gross dependent oedema. Clubbing of the fingers and toes was evident. The radial pulse was regular at 124 a minute. Her brachial blood pressure was 100/70 mm. Hg. The apex beat could be felt in the 8th left intercostal space in the mid-axillary line and there was forcible pulsation at the left sternal edge. A presystolic gallop rhythm was audible but there was no significant murmur. There were bilateral crepitations at the bases of the lungs. The abdomen was distended, and the liver was palpably enlarged. An electrocardiogram showed sinus rhythm, pointed P waves suggestive of right atrial enlargement, and right ventricular preponderance. A radiograph of the chest showed over-all cardiac enlargement with

grossly dilated main pulmonary arteries. The haemoglobin was 135 per cent (19.7 g./100 ml.) and the PCV was 70 per cent. She made good progress at first, under treatment with bed-rest, digitalis, and diuretics, but subsequently she developed multiple venous thromboses in

FIG. 1.—The heart from the front showing a single large artery arising from the summits of the two ventricles.

spite of anticoagulant therapy, and she finally died about seven weeks after admission.

At necropsy there were small bilateral pleural transudates of a clear fluid. The enlarged heart weighed 550 g. (Fig. 1). The venæ cavæ were normal. The right atrium was dilated. The tricuspid valve was normal in structure and circumference (11 cm.). There was considerable hypertrophy of the right ventricular infundibulum. A large defect was present in the upper part of the ventricular septum. A single large atherosclerotic artery arose from the summits of the two ventricles (Fig. 1): this contained three large semilunar cusps (Fig. 2), and two coronary arteries arose from it. Two wide unobstructed pulmonary arteries arose at right angles from the major vessel and passed into the lungs. The pulmonary veins were normal and opened into a dilated left atrium. The mitral valve was normal

FIG. 2.—View looking into the single artery which contains three enormous semilunar valve cusps.

in structure. The left ventricle was dilated and hypertrophied and also communicated both with the right ventricle through the ventricular septal defect and with the truncus arteriosus. This cardiac anomaly was considered to be a persistent truncus arteriosus of the form classified as type 3 by Collett and Edwards (1949).

The elastic tissue pattern of the atherosclerotic main pulmonary arteries was of aortic type, consistent with pulmonary hypertension being present from birth, but in other areas the elastic tissue showed a "moth-eaten" appearance produced by cystic medial necrosis with loss of elastic fibrils (Wagenvoort, Heath, and Edwards, 1964). There was grade 6 hypertensive pulmonary vascular disease. The small pulmonary arteries showed medial hypertrophy with the development of longitudinal muscle adjacent to both internal and external elastic laminae. The degree of intimal fibrosis was slight. Angiomatoid dilatation lesions (Wagenvoort *et al.*, 1964)

were seen and there was evidence of healed necrotizing arteritis. Numerous hypertensive pulmonary arterioles with a distinct media of circular muscle were present.

Discussion

The prognosis in persistent truncus arteriosus is very poor but is related to its anatomical type. Fontana and Edwards (1962) found that of the 94 patients with the disease reported up to that time 80 per cent had died during the first year of life and only 15 per cent had survived for more than 10 years. In types 1, 2, or 3 on the classification of Collett and Edwards (1949), where there is no form of obstruction to the excessive blood flow in the pulmonary arteries, the median age of survival is less than one month. In type 4 truncus the pulmonary arteries are absent, the blood reaching the lungs through the bronchial arteries, and there is thus a pronounced degree of obstruction of blood flow to the lungs: the prognosis is better in such cases, the median age of survival being 5 years (Fontana and Edwards, 1962). Survival into the second decade and beyond in persistent truncus has been reported several times when there is obstruction to pulmonary blood flow, but it is very rare when there is no such obstruction (see reviews of Collett and Edwards (1949) and Fontana and Edwards (1962)).

We believe that this patient survived at birth because the pulmonary vascular resistance remained high and this prevented an excessive flow of blood into the pulmonary circulation directly from the persistent truncus. The anatomical counterpart to this was presumably a persistence of the thick-walled muscular pulmonary arteries normally found in the foetal lung. Had the normal neonatal thinning of the muscular coats of the pulmonary arteries occurred it is likely that fatal pulmonary oedema would have ensued as in cases of ventricular septal defect and widely patent ductus arteriosus (Heath *et al.*, 1958).

Such survival is achieved only at the price of subjecting the pulmonary vasculature to the effects of severe pulmonary hypertension, which produces further structural changes such as occlusive intimal fibro-elastosis. A vicious circle of ever-increasing pulmonary hypertension and arterial occlusion ensues. At first the effects are mainly beneficial, as the pulmonary blood flow is controlled and the moderate increase in pulmonary arterial pressure gives rise to few, if any, symptoms, as in the present patient. Eventually, however, the more severe grades of hypertensive pulmonary vascular disease develop with associated gross increase in pulmonary vascular resistance and diminution of pulmonary blood flow, which lead to the patient's

death, as in this case. On rare occasions structural changes in the pulmonary trunk brought about by pulmonary hypertension, such as cystic medial necrosis, as seen here, may prove fatal more immediately by bringing about rupture and torrential hæmorrhage from this vessel.

This case is of interest in that it offers an example of the natural history of a patient with a large bi-directional post-tricuspid shunt with a pulmonary arterial pressure of systemic magnitude.

Summary

We report the case of a woman with persistent truncus arteriosus who died at the age of 38 years, the longest recorded survival in this disease. There was no associated obstruction to the flow of blood to the lungs and as a result the patient developed severe hypertensive pulmonary vascular disease.

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