

Anomalous origin of the right pulmonary artery from the ascending aorta associated with aortopulmonary window

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Anomalous origin of the right pulmonary artery from the ascending aorta associated with aortopulmonary window. A rare case of anomalous origin of the right pulmonary artery from the ascending aorta associated with an aortopulmonary window and severe pulmonary hypertension in a 13-year-old girl is reported. The window was closed using a Dacron patch and the continuity between the anomalous vessel and the main pulmonary artery was restored with a tubular Dacron graft. After operation the pulmonary pressure dropped to about one-half of the systemic pressure, and equal perfusion and ventilation of both lungs was demonstrated. Although early recognition and surgical treatment of this condition are mandatory to prevent the onset of irreversible pulmonary vascular disease, the presence of severe pulmonary hypertension did not contraindicate surgical correction in our patient.

Anomalous origin of one pulmonary artery from the ascending aorta is a rare malformation, which was first described by Fraentzel in 1868. In this condition, one lung is supplied from the aorta, whereas the other is perfused through the main pulmonary artery, which continues as a single vessel. Other cardiovascular anomalies, typically persistent ductus arteriosus, may be present.

The prognosis of these patients is poor because of the tendency to early congestive cardiac failure, which could be fatal. In addition, irreversible pulmonary vascular disease may develop (Caudill *et al.*, 1969; Keane *et al.*, 1974), which precludes corrective surgery.

The presence of an aortopulmonary window (Fraentzel, 1868; Keutel *et al.*, 1973; Blieden and Moller, 1974) increases the risk of these two complications.

The purpose of this paper is to present the case of a 13-year-old girl with this association of defects and severe pulmonary hypertension, who underwent successful surgical correction.

The embryological, diagnostic, and surgical features of this condition are presented.

Case report

A 13-year-old girl from Nairobi was known to

have had a heart murmur since early childhood. Her only complaints were mild effort intolerance and repeated upper respiratory tract infections. On examination she was a small girl with no cyanosis. The jugular venous pressure was normal. The arterial pulses were all palpable and equal, collapsing in nature, and the heart rate was 76 per minute. The blood pressure was 110/60 mmHg. There was a marked anterior prominence of the chest wall with evidence of biventricular hypertrophy on palpation. On auscultation the first sound at the left sternal border was followed by a soft, late systolic murmur, which continued through the second sound into early diastole. The pulmonary component of the second sound was moderately accentuated.

Haematological investigations were normal. The chest radiograph showed cardiomegaly (CTR 11.5/19) with a left-sided aortic arch, prominent pulmonary artery, and plethoric lung fields. The electrocardiogram showed sinus rhythm with evidence of biventricular hypertrophy. At cardiac catheterisation only the left pulmonary artery could be entered by the catheter from the right ventricle, while both pulmonary arteries were reached from the aorta. Saturations were consistent with a bidirectional shunt at ascending aortic and main pulmonary artery level. The pul-

monary artery pressure was at systemic level. On cine angiography the contrast medium injected in the right ventricle opacified the main and left pulmonary arteries (Fig. 1). An aortic root injection showed a normal aortic valve and early opacification of the main and left pulmonary arteries through a large aortopulmonary window (Fig. 2). The right pulmonary artery was seen arising from the proximal segment of the ascending aorta and was also slightly dilated. Both pulmonary arteries were tortuous and freely pulsating.

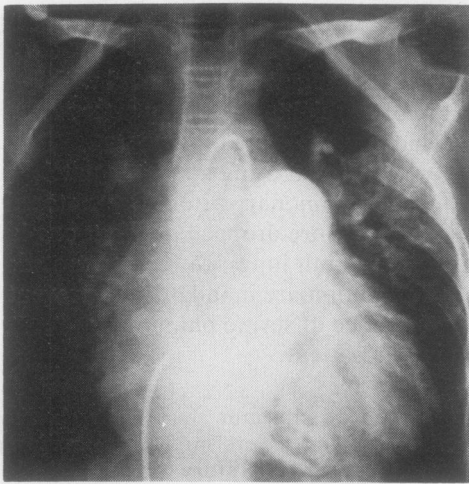


Fig. 1 Preoperative right ventriculogram showing the main and the left pulmonary arteries.

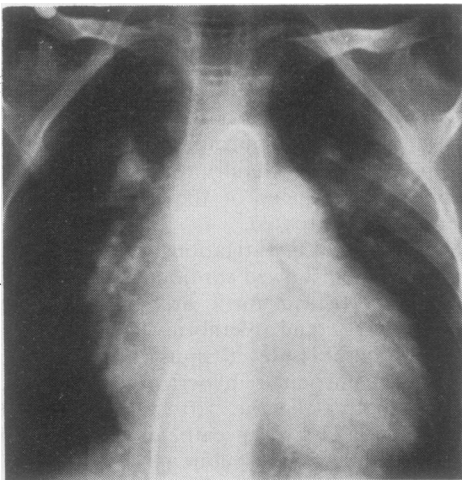


Fig. 2 Preoperative aortogram showing the aortopulmonary window situated just above the sinuses of Valsalva.

A diagnosis of anomalous origin of the right pulmonary artery from the ascending aorta associated with an aortopulmonary window was made. The patient was operated on in January 1976, with the aid of cardiopulmonary bypass and moderate hypothermia. The aorta was opened and a large aortopulmonary window 13 mm in diameter was found, together with an anomalous origin of the right pulmonary artery from the posterior surface of the ascending aorta, 2 cm above the sinuses of Valsalva. The right coronary artery orifice was located just below that of the main pulmonary artery. No patent ductus arteriosus was found. The left and main pulmonary arteries were separated from the ascending aorta using a Dacron patch inserted through the aortotomy. The right pulmonary artery, approximately 1 cm in diameter, was detached from its aortic origin and anastomosed end-to-side to the main pulmonary artery using a 10 mm Dacron tube running anterior to the aorta. At the end of the operation the pulmonary artery pressure dropped to about half of systemic pressure.

After the operation she made an uncomplicated recovery. Cardiac catheterisation one month later showed a pulmonary artery pressure of 45/15 with a systemic pressure of 130/80. There was no gradient across the patent graft. A pulmonary arteriogram showed normal main and left pulmonary arteries. The Dacron graft was patent although the filling of the right pulmonary artery was delayed (Fig. 3). A lung scan, however, demonstrated equal perfusion and ventilation of

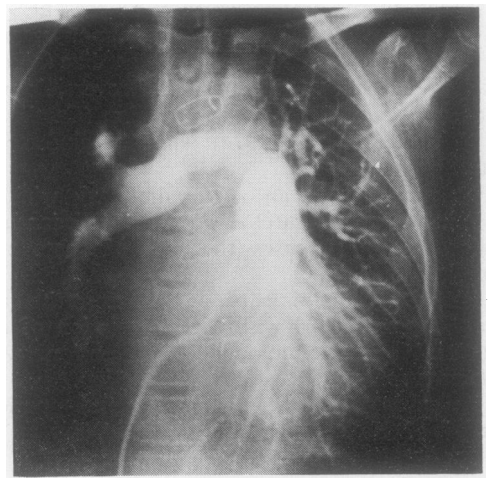


Fig. 3 Postoperative pulmonary angiogram showing good filling of both pulmonary arteries with some delay of filling on the right side.

both lungs (Fig. 4a, b). The patient was reviewed 21 months after the operation and was in excellent clinical condition.

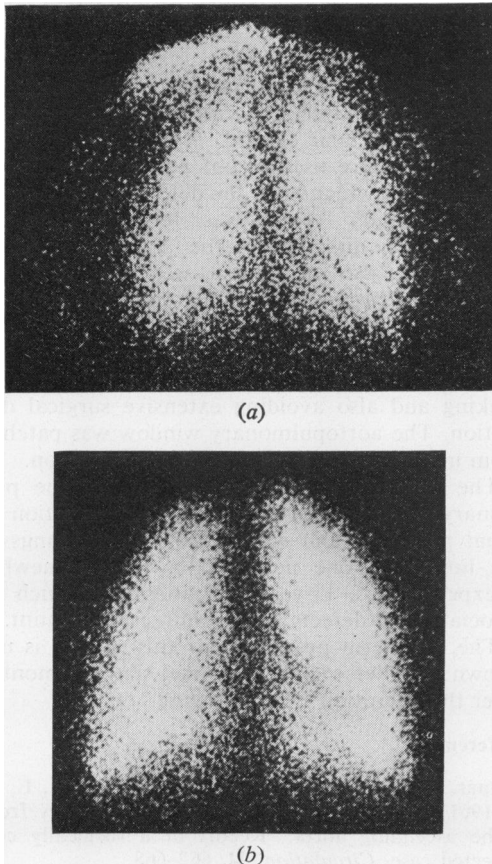


Fig. 4 Postoperative lung scan demonstrating equal ventilation (a—using ^{81m}Kr) and perfusion (b—using ^{99m}Tc) in both lungs.

Discussion

An anomalous origin of the pulmonary arteries from the aorta, apart from those associated with persistent truncus arteriosus, constitutes a specific clinicopathological entity. In most cases it is the right pulmonary artery which arises from the proximal half of the ascending aorta (Keane *et al.*, 1974). The embryological basis of this anomaly is not clear. According to Cucci *et al.* (1964), it might be a result of faulty septation of the truncus arteriosus. This septation process starts with the appearance of two ridges within the primitive

truncus, which extend from its cephalic end to the base of the conus and meet to form the truncoconal septum. It divides the aorta from the main pulmonary artery as well as the two ventricular outflow tracts. If the right truncoconal ridge originates from the primitive truncus more dorsally than normal, the proximal sixth aortic arch (and hence the right pulmonary artery) will arise from the ascending aorta.

The clinical picture is characterised by the early appearance of respiratory distress and congestive cardiac failure, the physical findings being those of a large left-to-right shunt. The electrocardiogram shows right ventricular hypertrophy with additional left ventricular strain in 25% of patients. The chest radiograph shows cardiomegaly and plethoric lung fields, more marked on the side of the anomalous vessel.

The anomaly is diagnosed by means of cardiac catheterisation and angiography. Haemodynamically, both pulmonary artery pressures are at systemic level in most of the cases.

The prognosis of these patients is poor since most of them die within the first year of life with congestive cardiac failure (Caudill *et al.*, 1969; Calazel and Martinez, 1975). Progressive obstructive pulmonary vascular changes may develop in the older patients, mainly in the 'systemic' lung (Caro *et al.*, 1957; Weintraub *et al.*, 1966; Winship *et al.*, 1967; Keane *et al.*, 1974). Endocardial fibroelastosis involving the right ventricle has also been described (Findlay and Maier, 1951; Griffiths *et al.*, 1962; Vásquez *et al.*, 1966).

Early recognition and surgical correction of this condition are advisable in view of the adverse natural history. In addition, it is hoped that surgery will prevent the onset of progressive, irreversible, pulmonary vascular disease.

Treatment consists of restoring the continuity of the pulmonary vessels and correcting any associated cardiovascular anomaly.

Primary anastomosis of the anomalous vessel to the main pulmonary artery (Kirkpatrick *et al.*, 1967; Winship *et al.*, 1967; Wilcox and Croom, 1968; Bjork *et al.*, 1970; Flege *et al.*, 1970; Blieden and Moller, 1974; Doty *et al.*, 1975; Neveux *et al.*, 1975) is feasible in the presence of a large, tortuous, anomalous vessel. This is passed behind the aorta and anastomosed to the main pulmonary artery using a running suture posteriorly and an interrupted suture anteriorly to allow future growth of the anastomosis.

The tubular graft, proposed by Maier (1954) has been used successfully by Armer *et al.* (1961) and others (Kuypers *et al.*, 1969; Keane *et al.*, 1974; Binet *et al.*, 1975).

The early and late results of surgery depend upon the degree of pulmonary vascular disease and the correction of the associated cardiovascular anomalies. Among 22 operated patients collected from the literature by Keane *et al.* (1974), 13 were under 1 year of age and all survived reconstructive procedures. Microscopical examination of the lung vessels did not show significant obstructive vascular disease in the majority of these patients, and the postoperative pulmonary artery pressure dropped to nearly normal values (Armer *et al.*, 1961; Redo *et al.*, 1965; Kirkpatrick *et al.*, 1967; Caudill *et al.*, 1969; Bjork *et al.*, 1970; Binet *et al.*, 1975; Neveux *et al.*, 1975).

The association of abnormal origin of the right pulmonary artery from the aorta with an aortopulmonary window is very rare.

To our knowledge, only three cases have been reported apart from the patient described in this paper (Fraentzel, 1868; Keutel *et al.*, 1973; Blieden and Moller, 1974). The age of the patients at the time of diagnosis varied from 2 years 9 months (Keutel *et al.*, 1973) to 24 years (Fraentzel, 1868).

According to Cucci's theory, the two anomalies may be related embryologically. It seems that if the dorsally displaced right truncal ridge delays correcting its sagittal growth, so as to meet the opposite ridge, it would fail to fuse with it at the distal level. The partial defect of septation would, therefore, result in a window between the aorta and the pulmonary trunk. This situation is, however, quite different from that which leads to a persistent truncus arteriosus, whose essential anatomical features are a common semilunar valve and a ventricular septal defect. This implies a total sagittal failure of septation of at least the most proximal segment of the truncus, as in type 1 of Collet and Edwards (1949). In our opinion, the term hemitruncus to describe this anomaly (Turpin *et al.*, 1962; Keith *et al.*, 1967) should be avoided as the essential features of truncus arteriosus (single semilunar valve and ventricular septal defect) are absent.

The clinical findings are suggestive of a truncus arteriosus with severe pulmonary hypertension. Both pulmonary arteries are typically entered by the tip of the catheter from the ascending aorta and are outlined following an aortic root injection. However, the presence of the pulmonary component of the second sound and the demonstration of two semilunar valves exclude the diagnosis of truncus arteriosus.

The prognosis of these patients is probably poor because of the risk of early development of obstructive pulmonary vascular disease, which may preclude successful corrective surgery. Our patient,

however, presented late in childhood, probably because of her low tendency to develop pulmonary vascular disease, in the presence of a large communication between the aorta and both pulmonary arteries. In spite of severe pulmonary hypertension she was considered suitable for operation. The haemodynamic findings and the angiographic appearance of the pulmonary vessels which were freely pulsating strongly suggested hyperkinetic pulmonary hypertension rather than irreversible pulmonary vascular disease.

The procedure used in our patient has allowed satisfactory correction of the defects. The anomalous pulmonary vessel was too short to be anastomosed directly to the main pulmonary artery. We also thought it was inadvisable to fashion a low-pressure tunnel inside the aorta, which might be subjected to possible distortion and compression. The graft was therefore placed anterior to the aorta, thus preventing its possible kinking and also avoiding extensive surgical dissection. The aortopulmonary window was patched from inside the aorta using a piece of Dacron.

The immediate striking reduction of the pulmonary artery pressure and the demonstration of equal perfusion and ventilation of both lungs in the lung scan one month later were somewhat unexpected in a 13-year-old patient with such an association of defects and a bidirectional shunt.

The long-term prognosis for this patient is unknown, but her excellent clinical state 21 months after the operation is encouraging.

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