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Hyperfunction of Blalock anastomosis associated with Marfan's syndrome

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Hyperfunction of the subclavian-pulmonary artery anastomosis causing pulmonary vascular disease is uncommon. The mechanism for the late development of an overgenerous shunt is usually obscure. We report the case of a 31-year-old patient with Fallot's tetralogy and Marfan's syndrome who had undergone a right Blalock manoeuvre at the age of 5. At necropsy the Blalock anastomosis was widely patent and the changes of plexogenic pulmonary arteriopathy were noted. The association of Marfan's syndrome offers an explanation for the continued growth of the subclavian-pulmonary artery connection and subsequent pulmonary vascular changes.

Case report

A 31-year-old white woman presented with palpitation and increasing effort dyspnoea (New York Heart Association functional class III). Her early childhood had been characterised by poor growth and cyanosis on exertion and squatting. A clinical diagnosis of Fallot's tetralogy was made, and in 1954 at the age of 5 years she underwent a right subclavian-pulmonary artery anastomosis. After operation she began to grow and attended a normal school. During adolescence there was clinical evidence of shunt patency despite rapid growth. In 1966, at the age of 17 years, a radiograph of the chest showed cardiac enlargement with considerable pulmonary vascular plethora. Her haemoglobin concentration was 14·8 g/dl with packed cell volume (PCV) of 47%. In 1969 she bore a live healthy son following an uncomplicated pregnancy. Total correction of her tetralogy of Fallot was advised, but she declined further intervention and defaulted from review.

When seen again in 1980 the patient was centrally cyanosed with long, thin, clubbed fingers and a reduced right arm muscle mass. Her height was 172 cm and there was a dorsal kyphosis. The apex beat was in the fifth left intercostal space in the midclavicular line and on ausculation there was a split first heart sound, a loud systolic murmur at the left sternal edge, and a single accentuated second heart sound. No murmur was audible to suggest a functioning Blalock anastomosis.

Investigations showed a normal biochemical profile, a haemoglobin concentration of 18 g/dl, and a PCV of 53%. The resting electrocardiogram showed sinus rhythm, a mean frontal QRS axis of +30°, and a normal ventricular concordance. There was cardiomegaly with considerable dilatation of the main pulmonary arteries seen in the chest radiograph (fig 1). Cardiac catheterisation confirmed the diagnosis of tetralogy of Fallot. A catheter was passed from the aorta into both the right ventricle and the left ventricle. The pressures in the two ventricles were identical and no catheter could be passed from the right ventricle across the pulmonary valve and into the main pulmonary artery. There was no pressure gradient across the Blalock shunt. The peak left ventricular oxygen saturation was 85% and that in the region of the ventricular septal defect was 62%. The mean aortic oxygen saturation was 74% and the mean right atrial saturation 55%. Aortic arch angiography showed enormous flow through the Blalock anastomosis with contrast medium swirling in massively dilated pulmonary arteries. The right ventricular angiography disclosed a tiny main pulmonary artery with infundibular and valvu-

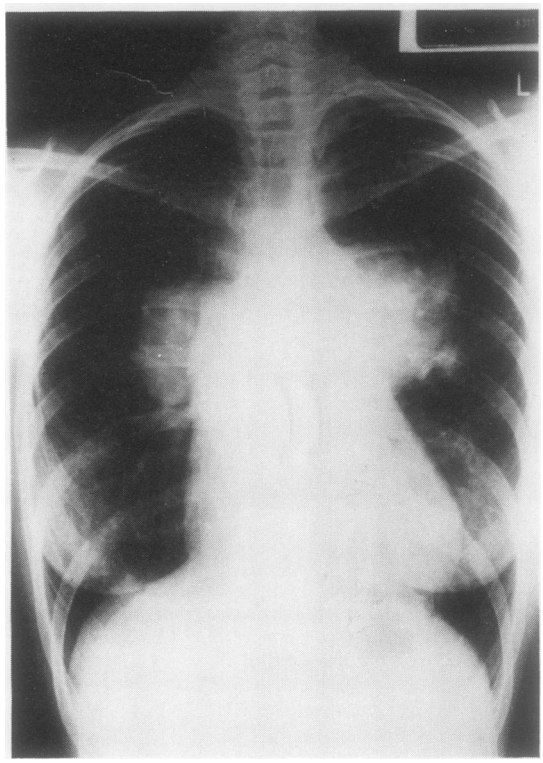

Fig 1 *Anteroposterior chest radiograph showing massively dilated main pulmonary arteries.*

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lar pulmonary stenosis. The bulk of the contrast injection was seen crossing a large ventricular septal defect into a smaller posterior left ventricle and passing into the large overriding aorta.

Ten months after these investigations the patient collapsed at home and died. Death was unrelated to the cardiac catheterisation and was caused by mediastinal haemorrhage from a circumferential tear in the left main pulmonary artery just proximal to its point of entry to the lungs. The heart showed typical anatomy of Fallot's tetralogy and the right Blalock anastomosis was widely dilated and patent. This histological appearance of the aorta and its main branches was typical of Marfan's syndrome. In the lungs changes associated with severe and irreversible pulmonary arterial hypertension were present. The larger muscular arteries had thickened muscular media with fasciculi of longitudinally orientated smooth muscle fibres in the intima and in the adventitia immediately adjacent to the media. The small muscular pulmonary arteries showed a pronounced concentric-laminar intimal fibroelastosis with the characteristic "onion skin" appearance and thinning of the underlying media. Plexiform lesions were seen (fig 2) and many of the muscular pulmonary arteries were totally occluded.

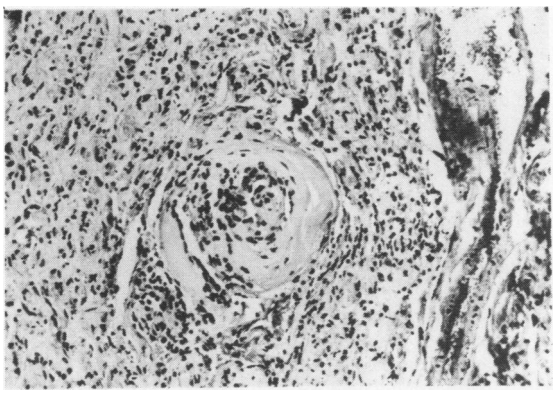

Fig 2 *Plexiform lesion (H and E × 77).*

Discussion

Pulmonary hypertension and pulmonary vascular disease are uncommon after subclavian-pulmonary artery anastomosis; 4% of 653 of Blalock's patients developed this complication.[1] When pulmonary hypertension does occur there is gross enlargement of the major pulmonary arteries.[2 3] Most patients with a Blalock-Taussig shunt have clinical evidence of diminishing pulmonary blood flow, suggesting restricted anastomotic growth with time.[4] Marfan's syndrome is recognised as a cause of arterial dilatation. Interruption of the continuity of elastic fibres and mucoid change in the media produce medial deficiencies allowing dilatation.[5] Our patient presented with pulmonary artery outflow obstruction and had pathological change in the pulmonary arteries typical of over-generous left-to-right shunting,[6] no doubt the result of dilatation of the Blalock-Taussig shunt. The mechanism of this dilatation and consequent pulmonary hypertension was the coexistence of Marfan's syndrome, and has not previously been reported. Indeed, the presence of Marfan's syndrome may be partly responsible for the enormous dilatation of the major pulmonary arteries, although the histopathological change in these vessels is indistinguishable from that caused by sustained pulmonary hypertension.[7]

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