

Middle aortic syndrome as a cause of heart failure in children and its management

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ABSTRACT Two cases of middle aortic syndrome in children are described along with two other cases reported earlier. In childhood, this disease may present as incipient or overt cardiac failure. Surgical treatment should be undertaken based on an objective assessment of the severity of the stricture and after taking into account the future growth of the child.

Middle aortic syndrome has been described as an acquired coarctation of the descending aorta caused by Takayasu's disease causing proximal hypertension in the young adult.¹ It can, however, occur in the paediatric age group and two cases have been described previously.² The reasons for the difficulties encountered in the management of these two children were not apparent at the time. Two more cases have subsequently been studied and have thrown light on the presentation of this disease in childhood and the problems of management at that age.

Case reports

CASE 1

A girl (MK), aged 11 years, was admitted with grade 3 effort intolerance, palpitations, and cough with expectoration. Her symptoms had started four months earlier. There was no history suggestive of rheumatic fever. On examination, she was an underdeveloped and malnourished child weighing only 16 kg with engorged and pulsatile neck veins. Cyanosis and clubbing of the fingers were absent. There were no palpable peripheral pulses in the right upper limb nor in either lower limb. Arterial pressure in the left arm was 160/90 mmHg. On investigation she had a normal leucocyte count and an ESR of 50 mm. Other biochemical investigations were normal. The Mantoux test was positive. A chest radiograph showed an enlarged cardiac shadow without any notching of the ribs. An intravenous pyelo-

gram was normal and the ECG showed a normal QRS axis, biventricular hypertrophy and inverted T waves in leads V1, V2, and V3. An aortogram showed diffuse narrowing of the descending thoracic and upper abdominal aorta.

She was treated with digitalis, diuretics, steroids, and reserpine in appropriate dosage along with nutritional supplements and vitamins. Recovery with medical treatment was satisfactory and the child gained weight. There was only a marginal reduction in arterial pressure. After about a month she was considered for surgical treatment but it was decided to defer it in view of the improvement. Surgical treatment would have necessitated a long bypass graft from near the arch to the lower abdominal aorta,² and we thought that the child would outgrow her graft if it were inserted at this age. She was discharged after three months of hospital treatment but died suddenly four months later presumably from cardiac failure. There was no necropsy.

CASE 2

This girl, aged 9 years, was admitted with grade 2 effort intolerance and chest pain for one month. There was no history suggestive of rheumatic fever or exposure to pulmonary tuberculosis. She was a normally developed child for her age with moderate anaemia and multiple cutaneous furuncles. There was evidence of cardiac enlargement and pulmonary infection. Pulsations were absent in the arteries of both inferior extremities; the arterial pressure was 150/90 mmHg in the right arm and 105/90 mmHg in the left. Investigations showed a normal leucocyte count and an

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ESR of 35 mm. The Mantoux test was positive. Routine biochemical tests were normal. A chest radiograph showed moderate cardiac enlargement without any notching of the ribs. The ECG showed a normal axis and biventricular hypertrophy. An aortogram showed a diffuse stricture of the descending thoracic aorta with maximum constriction just above the diaphragm. The left subclavian artery was also occluded (fig 1). A withdrawal tracing across the aortic stricture showed a systolic pressure gradient of 80 mmHg (fig 2). A radioisotope renogram showed depressed arterial flow in the left kidney. A pulmonary angiogram showed no involvement of pulmonary vessels. The child developed cardiac failure while investigations were in progress.

Treatment was started with digitalis, diuretics, and small doses of reserpine along with vitamins and a high protein diet. The child improved remarkably although the arterial pressure remained

at about the same level. In view of our experience with case 1 surgical treatment was advised. Insertion of a long bypass graft was considered unsuitable since the child would outgrow the graft. We therefore decided to perform an aortoplasty, for which a modified technique was devised. With the patient lying on her right side, a transthoracic exploration was carried out through the left fifth intercostal space. The thickened fibrosed descending thoracic aorta and its intercostal branches were dissected out from the periaortic fibrous tissue. Tapes were passed around apparently healthy aorta above and below the diseased area. Since the aorta below the lower limit of the diseased area could be reached through the chest, the abdomen was not opened.

The repair was carried out by incising the narrowed area throughout its length and applying an onlay patch of preclotted 12 mm knitted Dacron arterial prosthesis. The distal circulation

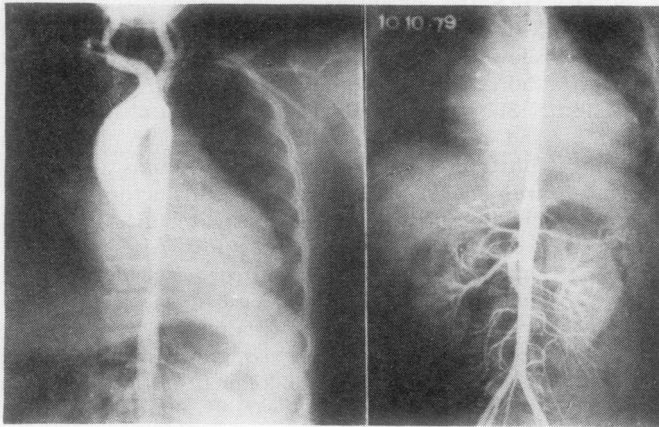


Fig 1 Preoperative aortogram of case 2. For description see text.

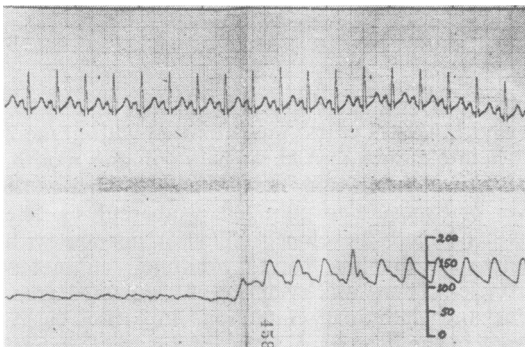


Fig 2 Withdrawal pressure tracing across the aortic stricture in case 2, showing a systolic gradient of 80 mmHg.

was maintained by an internal shunt of polyethylene tubing which was removed just before completing the suture line. Total aortic clamping time was about 45 minutes after insertion of the shunt and six to seven minutes before it.

The patient made an uneventful recovery with reversion of her hypertension to normal and equalisation of pressure between right arm and lower limbs. Diuretics were discontinued but she was kept on digitalis for a few months. An aortogram three months after operation (fig 3) showed that the anatomical correction of the defect could have been better although functionally there was no pressure gradient in the aorta. Eight months after operation the child is normal and asymptomatic.

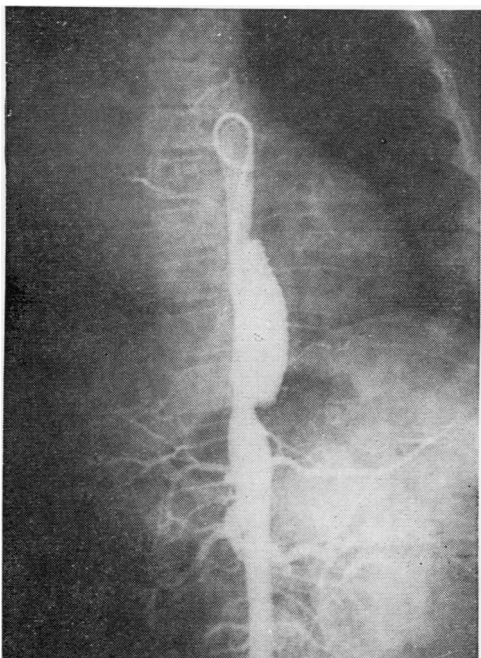


Fig 3 Postoperative aortogram of case 2. For description see text.

Discussion

Constriction of the descending aorta caused by Takayasu's disease is traditionally associated with the development of hypertension,^{1,3} but it has not been recorded that in children presentation with heart failure is more common. Any list of conditions causing heart failure in children should include acquired coarctation of aorta from Takayasu's disease as one to be considered in tropical countries.

In the first two children of this type reported earlier² the incipient cardiac failure was not recognised. One of them died of cardiac arrest on the operation table and the other recovered after a stormy postoperative period complicated by cardiac failure. In both patients the strictures were localised and were excised followed by graft replacement. Faced with the long stricture in the present case 1, we deferred operation since the child would have outgrown the long bypass graft. As a result she died from rapid worsening of the

cardiac failure in spite of medical treatment. Case 2 was not in heart failure at the time of admission but developed it later. She was therefore operated on and the aorta was enlarged. Anatomical correction of these defects could be improved with experience but the functional recovery has been satisfactory. The disease may extend and the patient may need further surgery in later life but she should grow normally through childhood and adolescence.

The main cause of heart failure in these children seems to be the hypertension caused by the rapidly progressive stricture of the descending aorta though there may be a primary cardiac affection as a part of a systemic disease. Irrespective of such a primary cardiac affection, it is essential to remove the aortic block by a method of surgical correction which will allow normal growth into adult life. To judge whether a stricture is critical enough to need surgical correction in children, objective assessment by angiographic appearance, measurement of pressure gradients, and estimation of renal blood flow is more important than the severity of proximal hypertension.

The technique of operation used in case 2, where the descending aorta had to be cross-clamped in the absence of adequate collateral circulation, is simpler than the other alternatives such as temporary bypass shunts or extra-corporeal circulation. We feel that this technique could be profitably used in some cases for surgical treatment of other diseases of the descending aorta.

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