Anatomical correction of complete transposition of the great arteries and ventricular septal defect in infancy

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British Medical Journal, 1976, **1**, 1112-1114

Summary

Two patients, aged 8 weeks and 5 years, with D transposition of great arteries and large ventricular septal defect were treated by transection of both aorta and pulmonary arteries and reattaching them to the appropriate ventricles. This included the origins of the coronary arteries. The ventricular septal defect was closed through a transverse ventriculotomy using a Dacron patch. The younger child was operated on as an emergency because of cyanosis and severe heart failure resistant to intensive medical treatment. The older child had had previous banding of the pulmonary artery at the age of 1 year. In both patients pulmonary artery pressure dropped to below half systemic pressure immediately after the operation. Postoperative progress was satisfactory with relief of cyanosis and heart failure. Early anatomical correction of transposition of the great arteries and ventricular septal defect is feasible and should play an important part in the management of these patients.

Introduction

Transposition of the great arteries is one of the commonest causes of cyanotic heart disease and accounts for 7-8% of all congenital heart disease.[1 2] If untreated the condition carries a grave prognosis.[3-5] Although the presence of ventricular septal defects slightly improves the initial survival rate,[3] it increases the risk of severe and possibly irreversible pulmonary vascular disease.[6-8] Recently Mustard's operation[9] or "inflow correction" has been accepted as the treatment of choice for transposition of the great arteries.[10-13] This, however, is not totally corrective. The most direct approach to the treatment of transposition is to correct the position of the aorta and pulmonary arteries in relation to the ventricles; this must include the coronary ostia. To have the greatest effect on the natural history of the disease this operation should be performed early in life. We describe here such an operation performed on an infant aged 8 weeks and on an older child.

Case 1

A 28-year-old mother gave birth to a boy weighing 3230 g on 24 August 1975. He was transferred to Harefield Hospital 12 hours later because of cyanosis and tachypnoea. Clinical findings were consistent with the diagnosis of transposition of the great arteries and ventricular septal defect. Cardiac catheterisation and angiography performed on 25 August confirmed the diagnosis; balloon septostomy was not performed. The peak systolic pressure was equal in both ventricles and there was no gradient across the pulmonary valve. The Pa_{O_2} was 5·6 kPa (42 mm Hg) while breathing pure oxygen. After initial improvement on medical treatment he deteriorated over the next two months with progressive cyanosis and tachypnoea. In spite of full medical treatment (digitalis and diuretics) his respiratory rate was 80/minute, his liver was palpable three fingers below the costal margin, and he required tube feeding. The chest radiograph showed increasing cardiomegaly with plethoric lung fields (fig 1). On 29 October 1975 his condition deteriorated further and it was decided that operation should be performed as soon as possible. This was performed the next day. At that time he weighed 4350 g.

Surface-induced profound hypothermia combined with cardiopulmonary bypass and circulatory arrest was used. The ascending aorta, aortic arch, and main pulmonary artery were mobilised. The ligamentum arteriosum was divided. The anteriorly placed ascending aorta was transected 3 mm above the top of the sinuses of Valsalva. The pulmonary artery, which was about twice the size of the aorta, was transected at the same level. The coronary ostia with a cuff of aortic wall about 1 mm wide were detached from the aorta and anastomosed to the corresponding sinuses of Valsalva of the pulmonary valve using 7/0 sutures. The proximal end of the pulmonary artery was then anastomosed to the distal end of the ascending aorta using 6/0 sutures. To match the size of the aorta to the large pulmonary artery the former was incised longitudinally on its posterior surface before starting the anastomosis. The defects in the aortic sinuses, produced by detachment of the coronary ostia, were then repaired using two patches of autogenous tissue (pericardium for one defect and free pulmonary arterial wall for the other). The proximal end of the aorta was then joined to the distal end of the pulmonary artery using a 10-mm Dacron graft.

A transverse right ventriculotomy was performed. This showed a large single infracristal ventricular septal defect measuring about 18 mm in diameter, which was repaired using a Dacron patch. A small secundum atrial septal defect was closed through a separate atriotomy. After rewarming bypass was discontinued without difficulty. The peak systolic pressure in the left ventricle and aorta was 90 mm Hg compared with 40 mm Hg in the right ventricle. Initially attempts at approximating the sternum resulted in a drop of systemic blood pressure. The postoperative course was complicated by one episode of sudden ventricular fibrillation six hours after operation, which was successfully treated. After that he made an uninterrupted recovery with complete relief of cyanosis and congestive cardiac failure.

Repeat cardiac catheterisation was performed six weeks after operation. This showed an arterial oxygen saturation of 95%. There were no residual shunts detected by oximetry. The right ventricular pressure was 50/0-3 mm Hg and the left ventricular pressure was

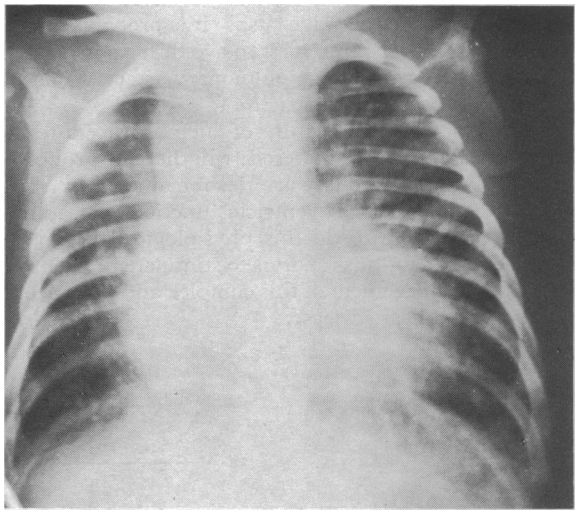

FIG 1—Case 1. Preoperative chest radiograph showing cardiomegaly and plethoric lung fields.

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90/0-5 mm Hg. There was a gradient of 35 mm Hg between the right ventricle and right main pulmonary artery. The left pulmonary artery was not entered. Left ventricular angiography showed normal appearances of the outflow tract, ascending aorta, and coronary arteries in contrast to the preoperative picture (fig 2). Right ventricular angiogram showed the relatively small pulmonary valve (originally aortic), the Dacron graft, and distal pulmonary arteries (fig 3).

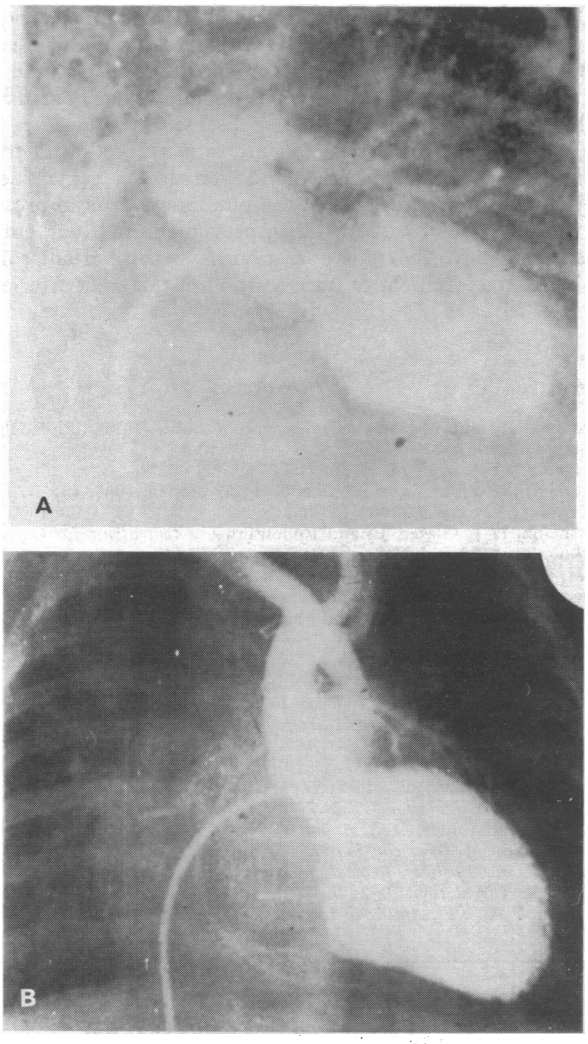

FIG 2—Case 2. (*a*) Preoperative left ventricular angiogram in showing pulmonary artery arising from left ventricle. (*b*) Postoperative left ventricular angiogram showing aorta filling from left ventricle with normal filling and distribution of coronary arteries.

Case 2

A child aged 5½ years was admitted to Harefield Hospital on 9 January 1976. He was known to have transposition of the great arteries and a large ventricular septal defect, which had been proved by cardiac catheterisation in Tübingen at the age of 4 weeks. At that time balloon septostomy was performed. At the age of 1 year a second cardiac catheterisation showed the pulmonary artery pressure to be at systemic level. This was followed by banding of the pulmonary artery by Professor Hoffmeister of Tübingen. The child continued to have shortness of breath and was severely cyanosed. A third catheter, passed in Tübingen at the age of 5 years, showed equal pressures in both ventricles; the pulmonary artery pressure distal to the band was between 55/32 and 60/21 mm Hg (systemic pressure was 93 mm Hg). Angiograms (fig 4) confirmed the diagnosis of transposition, ventricular septal defect, and banding. The haemoglobin level was 23 g/dl.

Total correction was performed on 12 January 1976 using a technique similar to that described in case 1. Immediately after repair the peak systolic pressure in the anterior ventricle was less than half that in the posterior ventricle.

He made a smooth postoperative recovery and was discharged from hospital two weeks after operation.

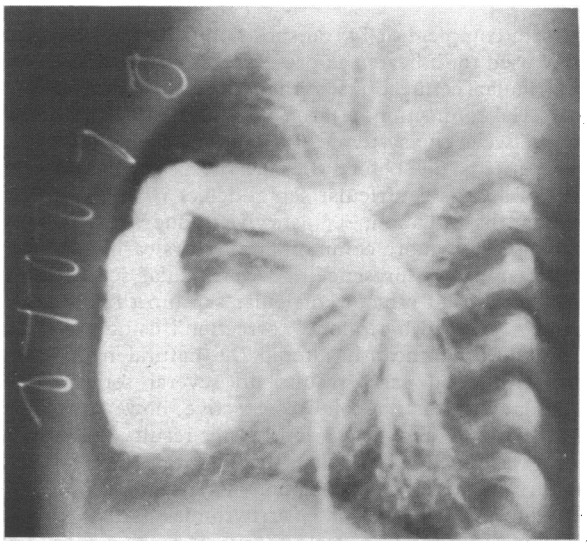

FIG 3—Case 1. Postoperative right ventricular angiogram showing small pulmonary (formerly aortic) valve, Dacron graft, and distal pulmonary arteries.

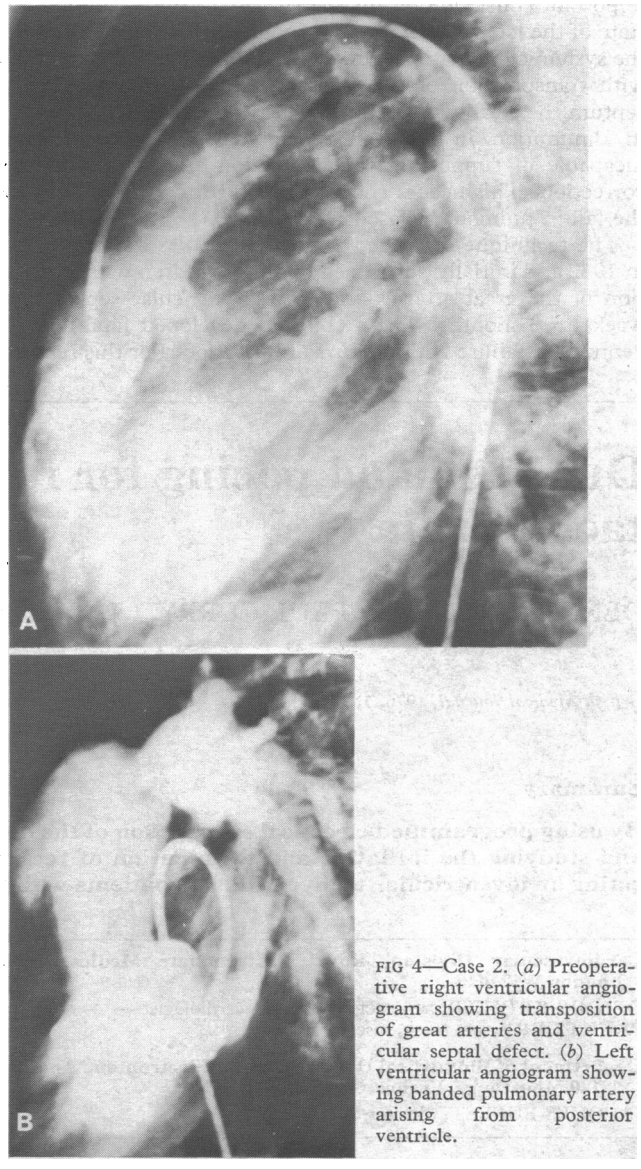

FIG 4—Case 2. (*a*) Preoperative right ventricular angiogram showing transposition of great arteries and ventricular septal defect. (*b*) Left ventricular angiogram showing banded pulmonary artery arising from posterior ventricle.

Discussion

The term complete or D transposition of the great arteries is generally used to indicate the presence of normal or concordant atrioventricular connections with abnormal (transposed) ventriculoarterial connections. In the absence of additional communication between the systemic and pulmonary circulation about 80% of patients die before the age of six months.[1-3] In the presence of a large ventricular septal defect 70% die within the first six months and about 20% survive to one year.[3] Pulmonary vascular disease is more common and occurs earlier (before the age of 1 year) in the presence of a ventricular septal defect.[6-8] In patients with an intact ventricular septum the use of atrial septostomy followed by inflow "correction" using the Mustard operation[9] has significantly altered the natural history of the disease, with good early results, in several series.[10-13] The Mustard operation is not totally corrective, however, and there is some concern about the long-term results. As the right ventricle continues to support the systemic circulation and tricuspid regurgitation,[14] obstruction of systemic and pulmonary venous drainage[15 16] and late arrhythmias[17] may constitute serious complications.

Total correction of transposition of the great arteries can be accomplished by transecting the aorta and pulmonary artery and reattaching them to the appropriate ventricles. One of the difficulties in applying this technique is the necessity of transposing the coronary ostia, which are very closely related to the aortic valve. The technique described in this paper illustrates the feasibility of this procedure very early in life. Another important consideration for the success of this operation is the state of the left ventricle, which should be capable of supporting the systemic circulation immediately after operation. In patients with transposition of the great arteries and intact ventricular septum the relatively low pulmonary vascular resistance results in diminution in left ventricular mass,[18] which renders it incapable of supporting the circulation after total anatomical correction. This occurs shortly after birth concomittantly with the fall in pulmonary vascular resistance.[19 20]

The technique described in this paper was developed by us in 1972 and initially performed in three children with transposition of the great arteries and intact ventricular septae aged 3 weeks to 3 months. These children developed fatal acute left ventricular failure within hours of operation. For this reason we believe that to be successful the totally corrective procedure in transposition with intact ventricular septum must be performed very early in life. In contrast, the functional state of the left ventricle is maintained in patients with transposition of the great arteries and ventricular septal defect. Nevertheless, the corrective procedure must be applied before the onset of irreversible pulmonary vascular disease, which is thought to occur between the ages of 6 months and 1 year in most cases. Banding of the pulmonary artery might prevent the occurrence of severe pulmonary vascular disease as in case 2. The creation of pulmonary stenosis in patients with transposition and ventricular septal defect, however, is not a guarantee against the development of pulmonary vascular disease.[6]

In the two patients described here total correction of transposition of the great arteries and ventricular septal defect resulted in an immediate fall of the pulmonary arterial pressure and was followed by dramatic improvement in their clinical states. We believe that this operation has an important role to play in the management of transposition of the great arteries.

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Dual-demand pacing for reciprocating atrioventricular tachycardia

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British Medical Journal, 1976, **1**, 1114-1116

Summary

By using programmed electrical stimulation of the heart and studying the initiation and termination of reciprocating atrioventricular tachycardia two patients with the Wolff-Parkinson-White syndrome were shown to respond rapidly and consistently to fixed-rate pacing. A demand pacemaker was implanted in each patient, having been modified so as to switch into the fixed-rate mode whenever the tachycardia began, thereby terminating the arrhythmia. This appears to be a promising form of treatment in patients with otherwise intractable paroxysmal tachycardia who have been shown by careful study to respond in this way.

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Introduction

While few patients with cardiac arrhythmias need therapeutic pacing, it may occasionally be the only means of controlling intractable paroxysmal tachycardia. In reciprocating tachycardias involving either the atrioventricular node or an anomalous