Short reports

Congenitally corrected transposition of the great arteries with tricuspid incompetence: successful treatment by xenograft valve replacement in a 15-month-old child

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Congenitally corrected transposition of the great arteries (CCTGA) with tricuspid incompetence as the only associated anomaly is a rare occurrence. This is a report of a 15-month-old child with this condition who was successfully treated by xenograft replacement of the incompetent valve.

Case report

In 1977, a 16-week old boy was referred to the Brompton Hospital with a 10-week history of irregular feeding, breathlessness, and a pansystolic murmur.

On examination he was pale, acyanotic, below the third percentile for weight and in moderate congestive heart failure. There was a loud aortic component to the second heart sound and a grade 4/6 pansystolic murmur was heard maximally at the apex. The ECG showed sinus rhythm, a superior axis (-30°), left ventricular dominance and right but not left-sided Q waves. Angiography demonstrated discordant atrio-ventricular and ventriculoarterial connections—that is, congenitally corrected transposition of the great arteries with severe left (tricuspid) atrioventricular valve incompetence (figure). No rhythm disturbances were demonstrated by 24-hour ECG monitoring.

Subsequently the baby developed a mid-diastolic murmur, a third heart sound, and signs of increasing cardiac failure associated with sweating, vomiting, and loss of weight. He also had repeated chest infections. By the age of 15 months his condition had failed to improve despite treatment with digoxin and diuretics, and it was decided that valve repair should be undertaken.

OPERATION

The heart was approached through a median sternotomy. Biventricular hypertrophy and massive left atrial enlargement were noted. Cardiopulmonary bypass was instituted with two cannulae, one in the aortic root and the other in the right atrium. The patient was cooled to 15°C and the circulation stopped. The left atrium was opened posterior to and parallel with Waterston's groove. This afforded a good view

Figure Cineangiograms showing (a) reflux from the morphological right ventricle into the left atrium; the aorta fills from the right ventricle, right ventricular injection; (b) the morphological left ventricle filling from the right atrium and emptying into the pulmonary circulation, right atrial injection.

of the systemic atrioventricular valve. The valve cusps were dysplastic and tethered to the left ventricular wall. It was considered that repair would not be practicable so the valve was excised and a 25 mm Carpentier-Edwards aortic xenograft sited using multiple 4/0 Ethiflex sutures.

The postoperative recovery was complicated by a coliform chest infection but was otherwise uneventful and he was discharged three weeks after operation.

At present he is in excellent health. He is asymptomatic, gaining weight rapidly, and presents no abnormal cardiovascular signs.

Discussion

Congenitally corrected transposition of the great arteries is commonly associated with other intracardiac anomalies, particularly pulmonary stenosis, VSD, and tricuspid valve anomalies or a combination of these. Symptomatic tricuspid valve incompetence, however, is seldom found as the sole associated lesion and presents in only 4% of all cases.

Morphological studies¹ have shown that dysplasia of the tricuspid valve is common in CCTGA, occurring in 91% of all cases of corrected transposition. Of these, 76% have dysplastic tricuspid valves characterised by "short thickened chordae tendineae that attach the cusps, particularly the posterior, to the ventricular wall." As Allwork points out, this situation is frequently called Ebstein's anomaly though it varies from his original description since in her series although there was displacement of the tricuspid valve into the right ventricle, the wall of the ventricle was not thinned. Hence, the case we describe falls into the category of anomaly described by Allwork though it should not strictly be termed Ebstein's anomaly, despite close similarity. Evidently the tricuspid incompetence was a direct result of the dysplasia of the valve and the associated tethering of the cusps to the ventricular wall.

Prosthetic valve replacement for tricuspid insufficiency in CCTGA was first described in 1964.^{2,3} Since then two cases have been reported in which xenograft valve replacement has been used. Fox *et al*⁴ describe three patients who underwent valve replacement, with one death. Two received Starr-Edwards valves and a third a porcine xenograft. All these patients had associated ventricular septal defects. Bailey *et al*⁵ describe the successful use of a heterograft prosthesis in a four-year-old with corrected transposition, ventricular septal defect, and tricuspid incompetence.

Xenograft valve replacement for CCTA with tricuspid incompetence alone has not previously been described. We feel that the following points are of interest.

The child presented in heart failure at the age of 9 weeks and his clinical condition deteriorated sufficiently rapidly to necessitate valve replacement at the age of 15 months. This appears to be the earliest

reported age at which valve replacement has been successfully undertaken in a child with CCTGA.

A median sternotomy and left atriotomy provided excellent access for the repair of the isolated defect. Bailey *et al*⁵ have described the use of a left thoracotomy where a ventricular septal defect and left atrioventricular valve incompetence coexist. This is an alternative where VSD repair is anticipated but is not necessarily required when the tricuspid incompetence is present alone.

It has been well demonstrated that in CCTGA the conducting tissue is situated away from the left-sided tricuspid valve, thus reducing the possibility of intraoperative damage with resulting dysrhythmias. Anderson *et al*⁶ showed that the atrioventricular node is usually situated anteriorly in the right atrium at the lateral junction of the pulmonary and mitral valves and gives rise to a single anteriorly situated bundle that descends into the morphologically left ventricle, skirting the anterolateral aspect of the pulmonary outflow tract and then passing to the anterior septum before bifurcating. They also state that in some cases parts of the conducting tissue may be duplicated, resulting in a left atrial node and posterior conducting bundle in addition to the normal conduction pathways. If this fact is taken into consideration when tricuspid valve replacement is contemplated then, as the remaining conducting tissue is relatively distantly situated, it is unlikely that valve replacement will compromise conduction. There were no dysrhythmias at any time in the child we describe.

The porcine xenograft has produced an excellent functional result and, in addition, there was no need for postoperative anticoagulation in a young child.

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