Case Reports

Valve Replacement Due to Tricuspid Valve Anomalies in Corrected Transposition

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SUMMARY

Left atrioventricular (A-V) valve anomalies are very frequent in corrected transposition, but left A-V valve replacement has seldom been reported. These anomalies are an important cause of morbidity and mortality in these patients, and due to anatomical presentation valve replacement may be difficult. We present 4 cases of left A-V value replacement with emphasis given to Ebstein's anomaly of the left A-V valve. (Jpn Heart J **34**: 103–107, 1993.)

Key Words:
Corrected transposition Valve replacement Ebstein's anomaly

CONGENITALLY corrected transposition is a combination of atrioventricular discordance with transposition of the great arteries. Anomalies of the left atrioventricular valve are seen in 90% of these cases and the most common anomalies are dysplasia and Ebstein's anomaly of the tricuspid valve.[1] Also, one-third of these patients have hemodynamic alterations due to these anomalies and some eventually need valve replacement.[2] Among 26 patients followed in our hospital since 1985 with the diagnosis of corrected transposition, 4 patients had Ebstein's anomaly of the left atrioventricular (A-V) valve. Two patients had important left A-V valve insufficiency and 1 patient had left A-V valve stenosis. One patient with Ebstein's anomaly and the 3 other patients with hemodynamically important tricuspid valve malformations underwent left A-V valve replacement in our clinic. Because there have been few reports of replacement of the left A-V valve, especially in those with Ebstein's anomaly, we have here presented these cases.

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Case Report

Since 1985, 26 patients were diagnosed in our hospital by cross-sectional echography and cardiac catheterization as having corrected transposition. Among these patients, 4 had Ebstein's anomaly (Table I). The diagnosis of Ebstein's anomaly was made with cross-sectional echography, and the displacement between the two A-V valves varied between 23 to 30 mm.

Despite the anomaly, only one of these patients had a significant degree of valvular insufficiency. This patient, a 4-year-old boy, first seen when he was a 16-month-old infant with congestive heart failure and failure to thrive, was diagnosed as having corrected transposition (SLL) with ventricular septal defect and left A-V valve insufficiency. His subsequent echocardiographies showed Ebstein's anomaly with a displacement of the tricuspid valve toward the apex (Fig. 1). At the age of 4 he was hospitalized again because of fever, and an echodense image was seen above the mitral valve. He was operated on in October 1989 with the additional diagnosis of possible infective endocarditis.

At operation the ventricular septal defect, a small one, was closed through the right atrium with a Dacron patch. The mitral valve was normal with no vegetations. The left A-V valve was exposed through the interatrial septum and was tricuspid with septal and posterior leaflets displaced into the morphologically right ventricle which was not atrialized, in a spiral fashion. The anterior leaflet was cleft with the leaflet barely maintaining its integrity. Exposure of the septal leaflet was inadequate so an aortotomy was performed and the valve was excised through it. A 23 mm Bjork-Shiley monostrut mitral valve was implanted with single sutures in the suprannular position. The patient maintained sinus rhythm and had an uneventful postoperative course.

Table I. Cases with Left A-V Valve Replacement

Anomaly	Age	Operation	Exposure	Valve	Follow-up
Cor. TGA, VSD Ebstein's Anomaly	4 years	VSD closure, Left A-V valve replacement, Epicardial pace lead	Right atriotomy aortotomy	23 mm Bjork-Shiley	18 months
Cor. TGA, Left A-V valve insufficiency	18 months	Left A-V valve replacement, Epicardial pace lead	Left atriotomy	25 mm Bjork-Shiley	1 year
Cor. TGA, Left A-V valve insufficiency	16 years	Left A-V valve replacement	Left atriotomy	29 mm Bjork-Shiley	3 years
Cor. TGA, Left A-V valve stenosis	14 years	Left A-V valve replacement	Left atriotomy	21 mm Bjork-Shiley	16 months

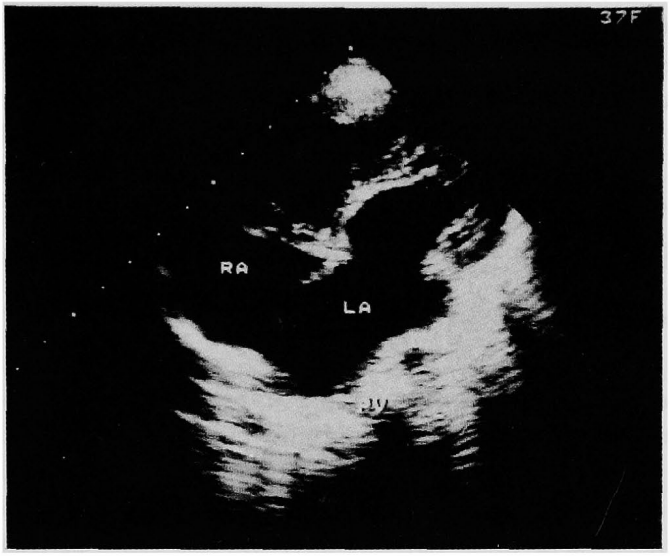

Fig. 1. Preoperative echocardiograph of the 4-year-old patient with Ebstein's anomaly.

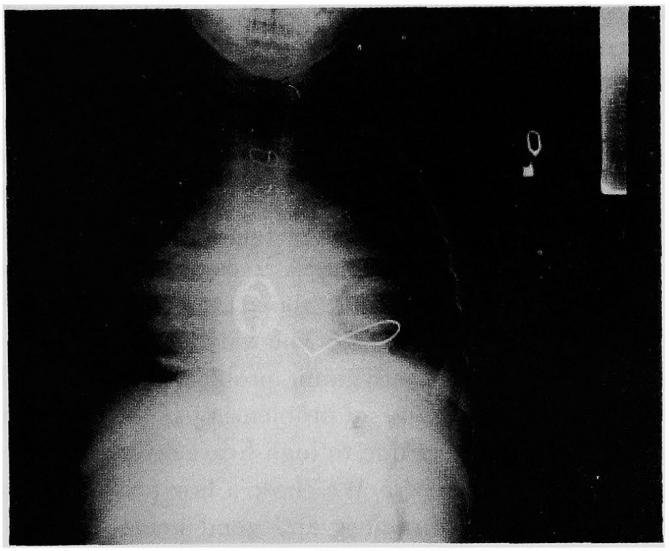

Fig. 2. Postoperative chest x ray of the 18-month-old patient with left A-V valve replacement and permanent epicardial pace lead implantation.

He is still well 2 years after the operation.

Two other patients required surgery because of left A-V valve insufficiency. One, an 18-month-old boy, received a 25 mm Bjork-Shiley mitral valve prosthesis (Fig. 2) and the other, a 16-year-old boy, received a 29 mm Bjork-Shiley mitral valve prosthesis. Another boy, 14 years of age, had

congenital stenosis of the left A-V valve with a gradient of 30 mmHg. The chordae were fused and attached to one papillary muscle resembling a parachute valve. A 21 mm Bjork-Shiley prosthesis was implanted.

All the patients had uneventful postoperative courses and were discharged on warfarin, acetyl salicylate and an occasional frusemide. Although all 4 patients were in sinus rhythm, epicardial permanent pacemaker electrodes were placed in 2 of the patients during surgery.

Discussion

The basic flow pattern in corrected transposition is normal, with the aorta carrying the saturated blood and the pulmonary artery carrying the unsaturated blood, but other underlying pathologies and the right ventricle working against high systemic pressure and resistance influence the flow pattern.[2] While 90% of these patients have various types of tricuspid valve anomalies, only one-third of these patients have clinically evident left A-V valve insufficiency.[1)-3)] Because of difficulties in diagnosis when seen with corrected transposition, the reported incidence of Ebstein's anomaly has varied, with figures of up to 75% of the cases in autopsy series.[1)-4)]

Left A-V valve insufficiency is progressive and is a serious problem encountered in the follow up of these patients and is difficult to alleviate without valve replacement.[3),5)] Ebstein's anomaly may limit surgical exposure of the septal leaflet of the tricuspid valve. Aortotomy in our patient helped to solve this problem. The anterior leaflet in these patients is not large as is the case in patients with Ebstein's anomaly and A-V concordance and it is cleft in 30% of the cases.[4)] The right ventricle above the displaced valve is not atrialized and plication is not necessary. The valve can therefore be sewn to the normal annulus with no hemodynamic problems. Valve replacement in small children is hazardous because of problems of small size, imperfect functioning of mechanical prostheses due to high heart rates and valve degeneration with biological prostheses.[6),7)] We chose a low profile mechanical prosthesis because of its proven durability and good hemodynamic function in children, despite problems such as pannus ingrowth, chronic anticoagulation and hemolysis.[6),7)]

Complete heart block, like left A-V valve insufficiency, is also a progressive and life limiting complication in corrected transposition. It is caused by an elongated A-V bundle that is connected to a posterior node which may undergo fibrosis in the course of the patient's life.[8)] We chose to place permanent epicardial pacemaker leads in 2 of our patients during surgery despite their being in sinus rhythm, and are planning to implant the generators

if rhythm problems occur in the future.

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