

Prosthetic Valve Implantation with Preservation of the Entire Valvular and Subvalvular Apparatus of the Tricuspid Valve in Congenitally Corrected Transposition of the Great Arteries

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ABSTRACT

Congenitally corrected transposition of the great arteries, which is characterized by atrioventricular and ventriculoarterial discordance, is a rare congenital heart disease. Most of the cases are diagnosed in childhood, owing to associated cardiac anomalies, such as ventricular septal defect, pulmonary stenosis or pulmonary atresia, and Ebstein-like malformation of the tricuspid valve. We present a patient with congenitally corrected transposition of the great arteries who underwent surgical replacement of the tricuspid valve with a bioprosthesis and reconstruction of the left ventricular outflow tract with bovine conduit.

INTRODUCTION

Congenitally corrected transposition of the great arteries, which is characterized by atrioventricular and ventriculoarterial discordance, is a rare form of congenital heart disease, constituting 0.5% of all congenital heart defects [Warnes 2006]. Most cases of congenitally corrected transposition of the great arteries are diagnosed in childhood, owing to associated cardiac anomalies, such as ventricular septal defect, pulmonary stenosis or pulmonary atresia, and Ebstein-like malformation of the tricuspid valve. Anatomic abnormalities of the tricuspid valve are common in patients with congenitally corrected transposition of the great arteries and negatively affect survival, whether treated surgically or not. Because of specific anatomic characteristics, the morphologic right ventricle functions as the systemic ventricle and the morphologic left ventricle functions as the pulmonary ventricle. Both ventricles have their unique atrioventricular valves, such as the tricuspid valve for the systemic ventricle and the mitral valve for the pulmonary ventricle. The management of a patient with corrected transposition of the great arteries is primarily determined by the presence and severity of the associated anomalies.

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We present a patient with corrected transposition of the great arteries who underwent surgical replacement of the tricuspid valve with a bioprosthesis (29-mm Mosaic[®]; Medtronic, Minneapolis, MN, USA) and left ventricular outflow tract reconstruction with 22-mm bovine conduit (Contegra[®]; Medtronic).

CASE DESCRIPTION

The patient was a 20-year-old-woman with corrected transposition of the great arteries. She was in New York Heart Association class II congestive heart failure and was admitted with fatigue and dyspnea. At 5 years of age, she had undergone ventricular septal defect closure and reconstruction of the left ventricular outflow tract with an 18-mm homograft.

A recent echocardiography evaluation revealed severe (4+) systemic atrioventricular tricuspid valve regurgitation (Figure 1A) associated with large dilation of the systemic (anatomic right) ventricle (ejection fraction, 60%). The pulmonary valve was severely stenotic, with a peak transpulmonary pressure gradient of 90 mm Hg.

The surgical reoperation was performed via a median sternotomy after cannulation of the right groin common femoral artery and vein. After establishment of extracorporeal circulation, the innominate vein was cannulated for venous drainage of the upper part of the body. The ascending aorta was cross-clamped, and antegrade cardioplegic cardiac arrest was obtained with cold blood cardioplegia. The systemic atrioventricular valve was explored via a left atriotomy. The exposure was absolute, and the systemic atrioventricular valve was Ebstein-like in appearance with the form of a tricuspid valve (Figure 2).

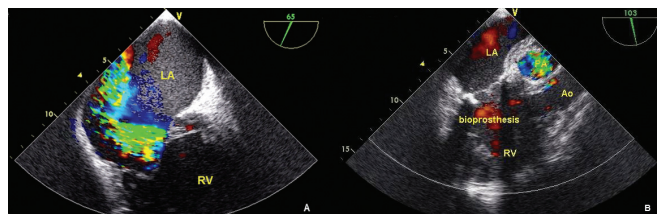


Figure 1. A, Preoperative midesophageal echocardiogram. B, Intraoperative midesophageal 3-chamber echocardiogram. LA, left atrium; RV, right ventricle (morphologic left ventricle); Ao, aorta; PA, pulmonary artery.

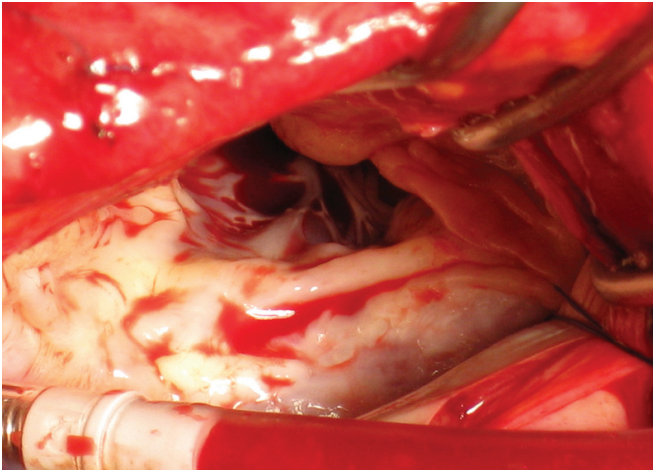


Figure 2. Intraoperative view of the Ebstein-like valve.

The tricuspid valve was not considered suitable for surgical repair, owing to poor midterm and long-term durability. A 29-mm bioprosthesis (Mosaic[®]; Medtronic) was implanted with 3-0 Prolene suture in a continuous fashion, while sparing the entire valvular and subvalvular apparatus of the tricuspid valve in situ. After the heavily calcified homograft was removed, a bovine conduit (Contegra[®]; Medtronic) was placed between the anterior wall of the left ventricle and the pulmonary artery. Extracorporeal circulation was withdrawn without difficulty. The durations of cardiac arrest and cardiopulmonary bypass were 40 minutes and 100 minutes, respectively. An intraoperative transesophageal echocardiography evaluation revealed no systemic atrioventricular valve regurgitation (Figure 1B).

The patient was extubated in 12 hours and transferred to the floor on the second postoperative day. Her postoperative period was uneventful. She was discharged from the hospital in 8 postoperative days.

DISCUSSION

Corrected transposition of the great arteries is a rare form of congenital heart disease [Warnes 2006]. The double discordance (atrioventricular and ventriculoarterial) results in a physiologically corrected circulation with the morphologic right ventricle serving as the systemic pump. Common associated conditions, which present in approximately 98% of cases, are ventricular septal defect, pulmonary stenosis, and congenital heart block. Most adult patients with diagnosed corrected transposition of the great arteries have undergone prior surgical repair in childhood for associated lesions that were hemodynamically significant. Our patient had received her diagnosis of corrected transposition of the great arteries and ventricular septal defect with pulmonary stenosis in childhood. She had undergone ventricular septal defect closure with placement of an 18-mm homograft valve between the left ventricle and the pulmonary artery. She had subsequently lived without any symptoms. Fifteen years later, however, heart failure with tricuspid regurgitation appeared, and an echocardiogram revealed severe tricuspid regurgitation associated with systemic ventricular dilatation.

The most common reported complications include tricuspid (systemic) valve regurgitation, right (systemic) ventricular dysfunction, congestive heart failure, and rhythm disturbances [Graham 2000]. The spectrum of valvular abnormalities ranges from minimal deformity, such as inferior displacement of the coaptation point, to severe dysplasia of one or more leaflets; valvular incompetence ranges from none to severe. A left-sided Ebstein-like anomaly is the most frequently observed malformation of the valve.

The surgical options for corrected transposition of the great arteries include tricuspid valve surgeries and a double-switch operation, in which the morphologic left ventricle becomes the systemic ventricle and the atrial blood is redirected accordingly. A double-switch operation, which is proposed as a more anatomic correction, is a technically challenging procedure with a significant mortality rate. Tricuspid valve surgical alternatives include repair, which is not recommended because of its poor durability, and conventional valve replacement [van Son 1995]. The conventional valve-replacement technique involves excision, with concomitant loss of the continuity between the valve annulus, valve tissue, chordae, and the ventricular wall. Clinical studies have shown, however, that preservation of the subvalvular apparatus plays an important role in maintaining ventricular performance after valve replacement [David 1983; Hennein 1990; Ghosh 1992]. Besides, the long-term outcome in corrected transposition of the great arteries is mainly determined by the function of the systemic ventricle, and consideration should be given to replacing the systemic atrioventricular valve at the earliest signs of progressive ventricular dysfunction. van Son et al reported that replacement of the tricuspid valve in a group of patients with ejection fractions <44% was associated with poor early and late outcomes [van Son 1995]. Consequently, we replaced the tricuspid valve at the beginning of ventricular dysfunction and spared the entire valvular and subvalvular apparatus.

In conclusion, implantation of a bioprosthesis in a young patient while preserving the tricuspid valve's entire valvular and subvalvular apparatus increases the probability that right ventricular function will be preserved over the long term.

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