Anatomic correction of Ebstein anomaly

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Between December 1997 and February 2001, 23 consecutive cases of Ebstein anomaly underwent anatomic correction.

Clinical Summary

There were 10 male and 13 female patients, aged 3 to 32 years (mean age, 14 years). Patients were in New York Heart Association

functional class II ($n = 12$) or III ($n = 11$). The cardiothoracic ratio ranged from 0.50 to 0.73 (mean, 0.65). Tricuspid incompetence was moderate in 10 and severe in 13 patients. According to the operative inspections, 2 patients were classified as having Carpentier type A, 3 type B, and 18 type C.

Most of the atrialized chamber was excised, and the cut edges were sutured with 4-0 polypropylene sutures. Meanwhile, the tricuspid anulus was plicated. The displaced posterior and septal leaflets, with their chordae tendineae and papillary muscles, were detached from the anulus and the ventricular wall, modified, and joined together to produce new leaflets. The newly produced leaflets were reattached to the plicated anulus, and the papillary muscles were reimplanted in an appropriate position. In 5 patients the septal leaflet was severely hypoplastic, with the appearance of membranous remnants. A piece of fresh autologous pericardium was sutured to the near-normal septal anulus level. The chordae transferred from the anterior leaflet or made by clipping membranous remnants were sutured to the free edge of the pericardium to make a new septal leaflet (Figures 1 and 2). Associated procedures included 1 Rastelli operation, 6 cases of atrial septal defect repair, 4 cases of patent foramen ovale closure, 2 cases of ven-

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Figure 1. Chordae made by clipping membranous remnants.

tricuspid septal defect repair, and 1 case of division of a patent ductus arteriosus.

All patients survived and recovered uneventfully; their New York Heart Association functional class improved to class I. Postoperative echocardiography showed that tricuspid incompetence disappeared in 19 and was mild in 4 patients, the displaced tricuspid leaflets were near their normal level, the right ventricle was reduced in size remarkably, and the atrialized chamber vanished. In our follow-up study (range, 1-27 months; mean, 12 months), exercise tolerance improved to normal, all patients were in sinus rhythm, and none had complete atrioventricular block. Echocardiography indicated that tricuspid incompetence disappeared in 19 and was mild in 4 patients.

Discussion

The proposed technique of treating Ebstein anomaly differs from previous techniques in some important ways. First, the atrialized chamber is excised longitudinally. Therefore, the right ventricular geometry is nearly restored, and the anatomic atrialized chamber is completely obliterated.

Second, a trileaflet valve is created, whereas in the techniques of Danielson,¹ Hetzer,² Vargas,³ and their colleagues, the anterior leaflet functions as a monocusp valve, and in the techniques of Carpentier,⁴ Quaegebeur,⁵ and their coworkers, the anterior leaflet and posterior leaflet function as a bileaflet valve. Whether it is monocusp or bileaflet, it is not the true anatomic structure of a tricuspid valve.

Third, there is almost no possibility for the tricuspid annulus to enlarge after annuloplasty because of the effective plication of the tricuspid annulus and competence of valve function and lightening of right ventricle pressure, and therefore the reconstructed tricuspid annulus was not reinforced with a prosthetic ring in any patient.

Figure 2. Autologous pericardium was sutured at near-normal septal annulus level. Chordae made by clipping membranous remnants was sutured to the free edge of the pericardium to make a new septal leaflet.

We think that the rigid tricuspid ring may reduce right ventricular function.

Because of complete excision of the atrialized chamber, a trileaflet mechanism covering the entire orifice area of the tricuspid valve, and restoration of the tricuspid annulus to its appropriate position and size at the atrioventricular junction, the valve function is competent. The incidences of right ventricular dysfunction and arrhythmia decrease, and a satisfactory result is achieved. The majority of patients with Ebstein anomaly could be free of tricuspid valve replacement. The indications for the conservative procedure have expanded. Long-term results require further observation.

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