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Surgical Experience of Ebstein's Anomaly

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Summary. **From 1969 to 1995, 15 patients with Ebstein's anomaly underwent surgery at Niigata University Hospital. Two patients with an infantile form presented heart failure and cyanosis immediately after birth, and underwent a staged operation resulting in long-term survival in one of them (operative mortality 50%). For 13 patients with the adult form, 14 operations were performed on the tricuspid valve (10 valve replacements and 4 valvuloplasties) with 1 early death (operative mortality 7.7%). Five patients with bioprosthetic valve replacement had satisfactory results without any early or late deaths; however, 4 patients with mechanical ball or tilting disc valve replacement showed poor results with 1 early death, 1 reoperation, and 2 late deaths. Over the last four years, newly invented valvuloplasty techniques (Danielson's method and Carpentier's method) have been successfully performed on 4 patients, and we propose that the valvuloplasty should be considered in adult form cases whenever the anatomy is favorable.**

Key words— Ebstein's anomaly, tricuspid valve replacement, tricuspid valvuloplasty.

INTRODUCTION

Ebstein's anomaly is a congenital malformation of the tricuspid valve, which is characterized by a downward displacement of septal and posterior leaflets. The malformation usually includes abnormal chordal attachments and a sail-like redundant anterior leaflet, by which tricuspid insufficiency or stenoinsufficiency of varying degree is induced. Right ventricular dysplasia and inter-atrial communication are also common associations of this anomaly. The clinical manifestation depends upon the severity of the valve deformity and associated lesions. In the most severe form, the patient presents cyanosis and heart failure immediately after birth, and the prognosis is grave unless appropriate surgical treatment is undertaken. On the other hand, there are patients who lead an asymptomatic life until late adulthood, and there is one report of Ebstein's anomaly incidentally found at autopsy.[1)] Therefore, the indication for surgery is sometimes difficult to decide. In this paper, we review 15 cases of Ebstein's anomaly operated on at Niigata University Hospital, and discuss the treatment policy.

PATIENTS AND METHODS

From 1969 to 1995, 15 patients with Ebstein's anomaly underwent surgery, and were followed up. Patients with Ebstein-like malformation observed in corrected transposition of the great arteries were excluded from this study. Including reoperations, 18 surgeries for the tricuspid valve were performed on these 15 patients. The patients' data are summarized in Table 1. Because the patients with early onset (during neonatal period or early infancy) differ in anatomical features and in the clinical course from those patients with late onset (during late childhood or adulthood), we describe these two forms separately. Two patients of the infantile form both presented cyanosis and heart failure during the neonatal period, and underwent the initial surgery in early infancy. In 13 patients with the adult form, five patients had atrial septal defect, two patients had Wolff-Parkinson-White (WPW) syndrome, one patient had atrial fibrillation, and one patient had atrial flutter as an associated lesion (Table 1). The surgical indications of these adult form patients were heart failure in six patients, heart failure with arrhythmia in four, heart

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Table 1. Patient data summary

Patient	Age	Diagnosis	Op. procedure	Result
⟨Infantile form⟩				
1 TY	3 mos	Ebstein+PS+ASD	Staged correction	Surviving
2 HM	2 mos	Ebstein+PA+VSD	Staged correction	op. death
⟨Adult form⟩				
3 NF	11 yrs	Ebstein	TVR	Surviving (reop.)
4 KK	15 yrs	Ebstein	TVR	late death
5 ST	52 yrs	Ebstein	TVR	late death
6 KN	11 yrs	Ebstein	TVR	op. death
7 TS	9 yrs	Ebstein	TVR	Surviving
8 KY	27 yrs	Ebstein+ASD	TVR+ASD closure	Surviving
9 MY	18 yrs	Ebstein+ASD	TVR+ASD closure	Surviving
10 FJ	14 yrs	Ebstein+ASD+WPW	Danielson+Kent division	Surviving
11 SF	44 yrs	Ebstein	TVR	Surviving
12 KR	11 yrs	Ebstein+ASD+WPW	Danielson+Kent division	Surviving
13 YM	55 yrs	Ebstein+AF	Carpentier's repair	Surviving
14 FT	11 yrs	Ebstein+ASD	Carpentier's repair	Surviving
15 MM	49 yrs	Ebstein+Af	TVR	Surviving

PS, Pulmonary stenosis; PA, Pulmonary artresia; ASD, Atrial septal defect; VSD, Ventricular septal defect; WPW, Wolff-Parkinson-White syndrome; AF, Atrial flutter; Af, Atrial fibrillation; TVR, Tricuspid valve replacement; Danielson, Danielson's repair; Kent division, bundle division.

failure with cyanosis in two, and arrhythmia alone in one.

CLINICAL COURSE AND RESULTS

Infantile form

For both patients with the infantile form, a staged operation strategy was employed for the surgical treatment. Patient 1 presented heart failure and cyanosis soon after birth, and the cardiac catheterization confirmed the diagnosis of Ebstein's anomaly with severe tricuspid valve regurgitation complicated with atrial septal defect (ASD) and pulmonary valve stenosis. At the age of 3 months, with a body weight of 5.1 kg, pulmonary valvotomy under the beating condition (Brock's procedure) was performed to increase the cardiac output and arterial oxygen saturation. At the age of 4 years, under extracorporeal circulation, tricuspid valvuloplasty (Hardy's operation) and ASD closure were performed; however, moderate tricuspid regurgitation persisted afterwards due to the thin and scant valve tissue. At the age of 8 years, the patient underwent tricuspid valve replacement with a Carpentier-Edwards bioprosthetic valve. Since then, the patient has been well and leading a normal school life for 6 years.

Patient 2 had Ebstein's anomaly complicated with pulmonary atresia, ventricular septal defect (VSD) and patent ductus arteriosus. Brock's procedure was performed at the age of 2 months to relieve the cyanosis and heart failure. However, the circulatory state did not improve, and the patient required a reoperation at the age of 6 months. This time, under extracorporeal circulation, a corrective surgery was performed, which consisted of VSD closure, right ventricular outflow enlargement, and Hardy's tricuspid valvuloplasty. The heart failure still persisted after the reoperation and at the age of 7 months, the third operation, tricuspid valve replacement, had to be done. The patient unfortunately died suddenly 1 month after the third operation.

Adult form

The operative results of the patients with adult form Ebstein's anomaly are listed in Table 2. A total of 14 operations were performed on 13 patients. The first 4 patients (Patients 3 to 6 in Table 1) received mechanical valve replacement. Patient 3 received tricuspid valve replacement with a Starr-Edwards ball valve at the age of 11 years; the valve had to be replaced by a St. Jude Medical valve at the age of 22 years

Table 2. Surgery of adult form cases

Valve replacement (10 valves)	
a. Mechanical valve: 5 valves	
Starr-Edwards valve	3 : 2 late deaths, 1 reoperation
Bjork-Shiley valve	1 : early death
St. Jude Medical valve	1 : surviving
b. Bioprosthetic valve: 5 valves	
Carpentier-Edwards valve	5 : surviving
Valvuloplasty (4 patients)	
Danielson's method	2 : surviving
Carpentier's method	2 : surviving

because of valve thrombosis. Patient 6 received mechanical valve replacement (Björk-Shiley tilting disc valve) and died on the first postoperative day due to low output syndrome. Although Patient 4 and Patient 5 were discharged from the hospital in a satisfactory condition after mechanical valve (Starr-Edwards valve) replacement, they both expired suddenly in the second and the third postoperative year, respectively. Since the clinical results of these first 4 patients were poor (1 early death, 2 late deaths, and 1 late reoperation), the choice of the prosthetic valve was changed to bioprosthesis (Carpentier-Edwards porcine valve) in 1975. Since then, there have been no early or late deaths. Recent policy has been to repair the tricuspid valve whenever possible, and so far 4 patients have undergone reparative surgery by Danielson's method (Fig. 1) or Carpentier's method (Fig. 2). Their valvular regurgitation grade, which was 3/4 in one patient and 4/4 in 3 patients preoperatively, was decreased to 2/4 postoperatively in all patients. They have been leading normal lives without cardiac symptoms.

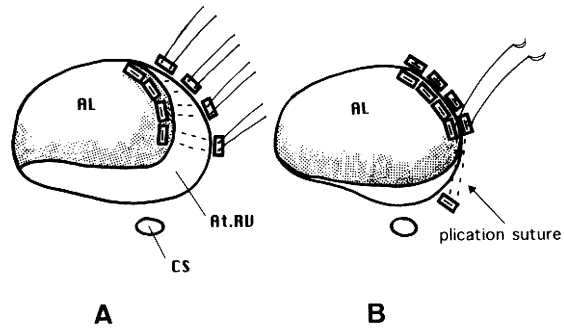

Fig. 1. Repair by Danielson's method. **(A)** Interrupted mattress sutures are placed to obliterate the atrialized right ventricle and to suspend the anterior leaflet to the original tricuspid annulus (right atrio-ventricular junction). **(B)** Aplication suture is placed in the posterior portion of the annulus for the annuloplasty. AL, anterior leaflet, At.RV, atrialized right ventricle; CS, coronary sinus.

DISCUSSION

The clinical presentation of Ebstein's anomaly cannot be described simply, because of the great variability of the valve deformity and the associated lesions.[2,3] The morphology and its hemodynamic consequence of Ebstein's anomaly that are presented during early infancy differ much from those which are presented during or after the school-age childhood. Therefore, these two forms should be considered as different clinical entities. The infantile form of the Ebstein's anomaly usually has a conspicuously malformed tricuspid valve which produces marked regurgitation, and is commonly associated with pulmonary artery lesions such as pulmonary stenosis or

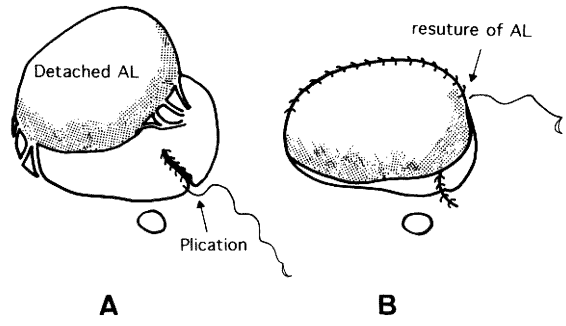

Fig. 2. Repair by modified Carpentier's method. **(A)** The anterior leaflet is detached from the annulus. The arialized right ventricle is plicated longitudinally. **(B)** The detached anterior leaflet is sutured to the original annulus.

atresia. The natural history of this form has been reported to be extremely poor,[4,5,6] and the surgical result, while improving, is still unsatisfactory. Primary total corrective surgery during infancy is not feasible in most of the cases. We previously reported a rare case of Ebstein's anomaly complicated with double tricuspid orifice and severe steno-insufficiency, which succumbed without corrective surgery.[7] The autopsy of this case revealed an irreparable tricuspid valve. Therefore, surgical treatment during infancy should be directed to the alleviation of the main symptoms presented at the time. In 2 patients of the infantile form in our series, one was successfully treated by multi-staged surgery. In spite of a gradual exacerbation of the tricuspid regurgitation after 2 operations, the patient survived to 8 years of age, when prosthetic valve replacement was performed. However, the other patient required prosthetic valve replacement at the age of 7 months because of severe tricuspid regurgitation, and died. Tricuspid valve replacement during infancy is known to carry an extremely high risk.[8] In general, dysplasia of the tricuspid valve is prominent and hypoplasia of the right ventricle is frequently associated with the infantile form. Therefore, the primary total anatomical correction is often unfeasible. In such cases with irreparable anatomy, the Starnes' procedure, which consists of tricuspid valve closure and an aorto-pulmonary shunt creation, may be indicated for the preparation of Fontan type operation, a physiological corrective surgery, in the future.[9] For the adult form of the Ebstein's anomaly, valvuloplastic surgery has lately been performed with satisfactory results in many institutions.[10,11,12] The recent trend is for the Carpentier's reparative method, which consists of the detachment of the anterior and posterior leaflets and re-suture at the atrioventricular junction combined with longitudinal plication of the atrialized right ventricle.[13] Originally, Carpentier's method was completed by placing a Carpentier ring at the new annulus; however, the use of the ring is still controversial, since the fixation of the annulus may reduce the right ventricular contractility.[14] In 2 patients we employed Quaegebeur's modification of Carpentier's method in which no ring was utilized.[15] Danielson's method, which consists of an obliteration of the atrialized right ventricle by suspending the anterior and posterior leaflets to the atrioventricular junction followed by annular plication at the posterior portion, is another choice for the tricuspid valvuloplasty.[11] In our series 2 patients underwent valvuloplasty by Danielson's method. Although these two reparative methods were equally effective in reducing the regurgitation—provided that the anterior leaflet is large and billowing—mild to moderate regurgitation was detected in all of them by echocardiography during the long-term postoperative period. The regurgitation mostly originated from the septal portion, where a complete leaflet coaptation is unatainable due to the plastered septal leaflet. Reports from other institutions also indicate that a complete disappearance of the tricuspid regurgitation could not be expected by these methods.[15] Technical advancement and refinement is needed to construct a completely sufficient valve. For the 9 patients who underwent tricuspid valve replacement, the decision to perform replacement instead of valve repair was primarily based on the following reasons: (1) extremely thin septal and posteior leaflets with marked plastering to the ventricular wall in 7 patients; (2) valve stenosis rather than insufficiency in 1 patient; and (3) a small anterior leaflet in 1 patient. Because the principle of valvuloplasty in both Carpentier's and Danielson's methods is to use the large anterior leaflet as a "monocusp valve", patients with a stenotic valve or small anterior leaflet are not indicated for these procedures, and they have to undergo valve replacement. However, the advanced deformity of septal and posterior leaflets does not necessarily obviate valvuloplastic surgery at present. Therefore, the 7 patients with valve replacement might have been fitted without a prosthetic valve if they were treated today using the recent valvuloplastic technique. Ten prosthetic valves were implanted in 9 patients in our series, and as is shown in Table 2, better clinical results were obtained in the bioprosthesis than for the mechanical ball or tilting, disc valve. As the number of the valves is limited and the surgical era is different, it may be inappropriate to conclude the superiority of the valve. However, we do think that a bioprosthetic valve such as Carpentier-Edwards porcine valve is the valve of choice in the tricuspid position. Satisfactory clinical results of tricuspid valve replacement with bioprosthesis have been reported,[16,17] and we assume that the bioprosthetic valve has an advantage in preventing thromboembolism in the right side of the heart, especially when the patient is likely to complicate ventricular and supraventricular arrhythmia as in Ebstein's anomaly. The indication for surgery was decided by the presence of heart failure, cyanosis, or tachy-arrhythmia attack. Since favorable surgical results have been obtained lately, we propose that the surgery should be indicated at an earlier stage before degenerative changes of the myocardium and valvular tissue ensue, so that a more precise valvuloplastic technique can be accomplished. At present it is our policy to surgically treat a patient who has a cardiothoracic

ratio greater than 65% and more than 3/4 degree of tricuspid regurgitation as detected by echocardiography, since this condition inevitably deteriorates with time.

In summary, surgery for the Ebstein's anomaly was performed on 2 patients with the infantile form and on 13 patients with the adult form. The operative mortality was 50% (1/2) for the infantile form, and 7.7% (1/13) for the adult form. The morphological complexity of the infantile form makes primary total intracardiac repair unfeasible, and staged operations should be considered. In adult forms, the clinical result of valvuloplastic surgery is improving, and it is the surgery of choice in current practice. If valve replacement is necessary, a bioprosthetic valve should be chosen.

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