Valve Replacement for Ebstein’s Anomaly of the Tricuspid Valve*

Early and Long-Term Results of Eight Cases

Tomio Abe, M.D.; and Sakuho Komatsu, M.D.

The role of valve replacement in the treatment of Ebstein's anomaly of the tricuspid valve remains controversial. Since 1965, eight patients with Ebstein's anomaly ranging in age from seven to 22 years of age (mean 13 years) have undergone tricuspid valve replacement (TVR) in our institution. Seven were in functional class 3 or 4 (NYHA), while one was in class 2, but had an increase in cardiac size. All of these eight patients showed moderate to severe cardiomegaly (cardiothoracic ratio: 54-75 percent). The valve was placed on the tricuspid annulus in seven of the eight cases, the exception being one case in which it was placed above the coronary sinus. Seven artificial prostheses (two Starr-Edwards, one Harken, one Kay-Shlley and three Wadacutter) and one porcine valve (Hancock) were used. There was one operative death in an early case due to postoperative complete heart block. Follow-up (range three to 18 years) of the other seven survival patients showed three late deaths; the first a pulmonary embolus 2.6 years after TVR; the second with congestive heart failure two years following the operation; and the third with arrhythmias five years after TVR. The remaining four patients are now in functional class 1 (three cases) and class 2 (one case). Valve replacement for Ebstein's anomaly can produce good clinical improvement and provide excellent long-term results.

Ebstein's anomaly is a relatively rare congenital malformation, and there have been few reported series with a large number of patients undergoing surgical treatment. Optimal surgery for Ebstein's anomaly has not yet been established, and the method of choice between valve replacement and reconstruction remains controversial. Since 1965, eight patients have undergone tricuspid valve replacement for Ebstein's anomaly at Sapporo Medical College Hospital. This study assesses both the early and long-term results following valve replacement for Ebstein's anomaly.

Patients and Methods

The preoperative clinical status, age of the eight patients with Ebstein's anomaly surgically treated in our hospital, and their postoperative courses are summarized in Table 1 and 2. There were six male and two female subjects with a mean age of 13 years (range 7 to 22 years). All of the patients had moderate-to-severe dyspnea, and all but one exhibited cyanosis. The indications for operation were progressive subjective complaints of dyspnea, vertigo, and cyanosis in five patients and severe congestive heart failure in the remaining three patients.

Chest roentgenograms revealed moderate-to-marked cardiomegaly with a mean cardiothoracic ratio (CTR) of 62 percent and diminished pulmonary vasculatures.

Seven patients with intra-atrial shunt displayed increased hemoglobin (16.9±2.1 g/dl) and decreased arterial oxygen saturation (76.8±7.6 percent) as the result of the right-to-left shunt. Electrocardiograms demonstrated right ventricular hypertrophy in six patients and left ventricular hypertrophy in two patients. Normal sinus rhythm (NSR) was present in seven patients, complete right bundle branch block (CRBBB) was noted in three patients, and atrial fibrillation was present in one patient.

Cardiac catheterizations and right ventriculograms were performed on all of the eight patients and confirmed the final diagnosis of Ebstein's anomaly. One patient who had Hardy's operation 12 years before was studied by echocardiography.

Operative Findings and Procedures

All patients had median sternotomy, and the heart was induced electrically to fibrillate during operative procedure with moderate hypothermia (28 to 32°C). The operative procedures are summarized in Table 2. Pronounced valvular incompetence was noted in seven patients, with the most recent case (case 8), who had plication of the septal valve 12 years before, having paradoxical distension of the atrialized part of the right ventricle. The atrial septal defect (ASD) was of the foramen ovale type in one or was a secundum defect in six patients, ranging in diameter from 10 to 30 mm. The tricuspid valve was excised, and the prosthesis was inserted in the anatomic tricuspid annulus with interrupted mattress sutures in seven patients. In the most recent case, the porcine valve was placed above the coronary sinus on the atrial septum without the excision of the tricuspid valve. The ASD was closed simultaneously with a Dacron

Table 1—Valve Replacement for Ebstein's Anomaly

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>NYHA</th>
<th>Symptoms and Signs</th>
<th>CTR (%)</th>
<th>Preop ECC</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>12</td>
<td>4</td>
<td>Dyspnea, cyanosis</td>
<td>70</td>
<td>NSR CRBBB</td>
</tr>
<tr>
<td>2</td>
<td>14</td>
<td>3</td>
<td>Dyspnea, vertigo, cyanosis</td>
<td>58</td>
<td>NSR</td>
</tr>
<tr>
<td>3</td>
<td>10</td>
<td>2</td>
<td>Dyspnea, fatigue, palpitation</td>
<td>54</td>
<td>NSR</td>
</tr>
<tr>
<td>4</td>
<td>13</td>
<td>3</td>
<td>Dyspnea, cyanosis, vertigo</td>
<td>60</td>
<td>NSR</td>
</tr>
<tr>
<td>5</td>
<td>8</td>
<td>4</td>
<td>Dyspnea, cyanosis, CHF</td>
<td>65</td>
<td>NSR CRBBB</td>
</tr>
<tr>
<td>6</td>
<td>7</td>
<td>3</td>
<td>Dyspnea, cyanosis, vertigo</td>
<td>56</td>
<td>NSR</td>
</tr>
<tr>
<td>7</td>
<td>22</td>
<td>3</td>
<td>Dyspnea, cyanosis, CHF</td>
<td>58</td>
<td>NSR CRBBB</td>
</tr>
<tr>
<td>8</td>
<td>16</td>
<td>4</td>
<td>Dyspnea, af, CHF</td>
<td>75</td>
<td>Af</td>
</tr>
</tbody>
</table>

CHF = Congestive heart failure; NSR = normal sinus rhythm; CRBBB = Complete right bundle branch block; AF = atrial fibrillation

diastolic hypertension.

*From the Department of Cardio-Thoracic Surgery, Sapporo Medical College, Sapporo, Hokkaido, Japan.

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patch in four and with suture method in three patients to discontinue the right-to-left shunt. No attempts were made to plicate the atrialized right ventricle.

Results

Operative death occurred in the first of the eight patients. The patient, who had severe congestive heart failure and marked cardiomegaly, died on the second day after surgery with complete heart block. The other seven patients, however, survived the operation with a mean follow-up period of 12.6 years (range 3 to 18 years). Hemodynamic data, preoperative, and one month after valve replacement, are shown in Figure 1.

The major complication after operation was dysrhythmia in these remaining seven patients, with three suffering late postoperative deaths. One patient, who was in functional class 2 with normal sinus rhythm, but who had complete right bundle branch block with left axis deviation (LAD), died suddenly on the way home from school five years after the operation. Another patient, who had congestive heart failure preoperatively, died two years after the operation because of bradycardia and progressive myocardial dysfunction. The third patient had a pacemaker because of postoperative complete heart block, but was active for three years as a clerk. He then developed chest pain and hemoptysis and died suddenly at another hospital. Pulmonary embolus due to the mechanical prosthesis was strongly suspected in this patient.

At present, there are four surviving patients and at follow-up of 3, 13, 16 and 18 years after the operation, respectively; all four patients were acyanotic and clinically much improved, with three in functional class 1 and one in functional class 2. One patient was in

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class 1 for nine years until complete heart block developed. He had a permanent pacemaker implanted seven years ago. He is now, however, very active as a carpenter.

**DISCUSSION**

The clinical features and natural prognosis of Ebstein's anomaly are strongly dependent upon abnormalities in the attachment of the tricuspid valve. There are few reports of patients who have survived for relatively long-term periods without surgical treatment. The natural prognosis of this disease is not so satisfactory due to associated cardiac lesion, increased cyanosis, progressive cardiomegaly, and the appearance of congestive heart failure.

Therefore, the indication for surgery seems to be strong for patients in functional classes 3 and 4, those patients with moderate-to-severe cyanosis, paradoxical emboli, or arterial thrombosis and right ventricular obstruction. A few patients in class 2 who show a rapid increase in cardiac size, dyspnea, and cyanosis should also be recommended for surgical treatment. Even after surgery for Ebstein's anomaly, however, there still remain the disturbing problems of arrhythmia and sudden death.

The optimal surgical correction for this condition has not yet been established, and the modality of repair of valvular dysfunction in Ebstein's anomaly remains the most controversial problem. Our experiences and other reports suggest that Ebstein's anomaly has a wide spectrum involving abnormalities of the tricuspid valve and the right ventricular geometry. Since most patients suffering from Ebstein's anomaly, however, have marked dilated right ventricle, with this dilatation involving not only atrialized ventricle but also the right ventricular infundibulum, the method of choice, either reconstruction or valve replacement for valvular dysfunction, must be decided upon. In 1958, Hunter and Lillehei suggested valvular reconstruction, and then in 1964, Hardy and his colleagues reported practical application and further development of this procedure. It has not always been indicated for Ebstein's anomaly because of the variations in this disease and because of the continuing high morbidity and mortality. Even our present report, one patient, who had plication of the septal valve 12 years before, was admitted with congestive heart failure with atrial fibrillation. Her condition was much improved after prosthetic valve replacement.

On the other hand, in 1963, Barnard and Schrine reported the first successful treatment of two patients with Ebstein's anomaly by the insertion of a prosthetic valve. Most of cases, however, have numbered less than ten in the majority of reports, and there have been few long-term follow-up studies of successfully-treated cases. Watson reported, in an international cooperative study of 505 patients with this anomaly, that nine of 16 patients (56 percent) undergoing valve replacement died after surgery. On the other hand, Barbero-Marcial and his colleagues, and Bove and Kirsch, reported excellent results with valve replacement for Ebstein's anomaly. In our present report, only one early patient, who had valve replacement, died following operation.

There are two important technical problems concerning the fixation of the prosthetic valve. Although Barnard and Schrine and others recommended the fixation of the prosthesis above the coronary sinus to avoid injury to the bundle of His, there is a high incidence of postoperative heart block with this method. On the other hand, Barbers-Marcial et al. and Melo et al. mentioned that they placed the prosthetic valve on the anatomic ring, and that there were two of 20 patients with complete heart block postoperatively. On the basis of our experience and that of others, the fixation of the prosthesis should be made in the anatomic ring to produce the most excellent long-term results.

The other technical problem is that the atrialized right ventricle interferes with improvement of right ventricular filling and pumping after the prosthetic valve replacement. Some authors have insisted that it is not always necessary to do plication at the time of valve replacement. The authors believe that this procedure is unnecessary and somewhat dangerous.

In four of our eight patients, at follow-up periods from three to 18 years after valve replacement, especially in careful evaluation of the right ventricular segmental size by echocardiogram, the paradoxical movement of the atrialized ventricle was diminished, and the functional right ventricular cavity was reduced.

A long-term follow-up study has been completed in the seven surviving patients. All of them improved in functional class and have been leading active lives for two to 16 years following valve replacement. Two of three patients with late follow-up, in whom sudden death occurred two to five years postoperatively, suggested that valve replacement does improve the quality of life and that it is possible to prevent the sudden death with careful postoperative management.

It is concluded that valve replacement produces good and long-lasting clinical improvement in patients with Ebstein's anomaly.

**REFERENCES**

National Conference on Advances in Cancer Therapy

The American Cancer Society will present this national conference at the Waldorf-Astoria Hotel, New York City, December 8-10. For further information, please write Nicholas G. Bottiglieri, M.D., American Cancer Society, 777 Third Avenue, New York 10017.