# Long-Term Results After Starr-Edwards Mitral Valve Replacement in Children Aged 5 Years or Younger

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*Background.* **Mitral valve replacement with prosthetic valves in infants and children requires consideration of multiple variables. When we examined our late follow-up, the effect of the growth of the patient relative to the size of the prosthesis placed was the most important variable predicting late successful results. We reviewed our experience with mitral valve replacement using the Starr-Edwards ball valve in children aged 5 years or younger, focusing on the effect of valve prosthesis–patient mismatch on the long-term results in the growing patient.**

*Methods.* **From August 1974 to June 1986, 8 patients aged 5 years or younger underwent mitral valve replacements using the Starr-Edwards prosthesis size 0M in 3 patients and 1M in 5 patients. Model 6320 was used in 1 patient and Model 6120 in the remaining 7 patients.**

*Results.* **Follow-up was 100% from 15 to 27 years (mean, 20 years). No valve-related complications of thromboembolism, anticoagulant-related hemorrhage, or prosthetic valve endocarditis were seen. All patients normally developed to adult size. The range of the valve area index of the 3 patients who received the smaller Starr-Edwards valve (size 0M) was 0.97 to 1.24 $cm^2/m^2$. Although this size valve was adequate for patient growth to adolescence, in each case valve replacement with a larger valve was required.**

*Conclusions.* **Our long-term review of Starr-Edwards ball valve mitral valve replacement in children aged 5 years or younger shows that the Starr-Edwards ball valve (Models 6320 [1 patient] and 6120 [7 patients]) showed excellent durability, no thromboembolism, and no anticoagulant-related complications. Size 0M valves required replacement for hemodynamic reasons because of patient growth; larger size 1M valves remained hemodynamically satisfactory in spite of patient growth.**

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Mitral valve repair has improved and may provide acceptable intermediate-term and long-term results [1–3]. However, the procedures are not always feasible, especially in children whose valvular apparatus is frequently complex and fragile. Such cases may require mitral valve replacement (MVR), which involves the following specific considerations unique to this age group [4]: (1) size of the mitral annulus, left atrium, and left ventricle; (2) association with other cardiovascular anomalies; (3) maintenance of adequate anticoagulation; (4) the fixed prosthetic effective orifice size; and (5) prosthesis durability. When the long-term result is investigated in a growing child, the most influential concern is the valve–patient size mismatch that progresses with patient growth. Here we review our experience with MVR using the Starr-Edwards ball valve (SEBV) in children aged 5 years or younger and focus on the effect of valve prosthesis–patient size mismatch on the long-term results in the growing patient.

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## Patients and Methods

From August 1974 to June 1986, 14 children aged 1 to 5 years underwent 16 mitral valve operations (three mitral valve repairs, one open mitral commissurotomy, and 12 valve replacements [MVRs] using nine SEBVs and three Björk-Shiley valves at the Tokyo Women's Medical University Daini Hospital). Among these patients, 1 who had a valve repair, 1 who had an open mitral commissurotomy, and 1 each who had MVR with SEBV and Björk-Shiley valve died perioperatively. Another patient who received a mitral valve repair and 1 who received an MVR with a Björk-Shiley valve required a replacement of mitral (prosthetic atrioventricular) valve with SEBV 4 years and 1 year after initial operation, respectively. An additional 2 patients, 1 with mitral regurgitation and 1 with Björk-Shiley MVR failed their follow-up.

Therefore, in this study we investigated the long-term outcome of 8 children with MVR using the SEBV (Table 1). Of these patients, 4 had undergone previous procedures. Case 1 underwent ligation of a patent ductus arteriosus, and case 5 underwent closure of an atrial septal defect. Case 2 underwent MVR with a Björk-Shiley valve before re-MVR with the SEBV because of prosthetic valve failure caused by thrombosis. Case 3 underwent


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*Table 1. Patient Profiles*

| Case | Age (y) | Gender | Weight (kg) | Diagnosis | Associated Anomalies | Previous Procedures | Mitral Valve Replacement with Starr-Edwards Ball Valve | Size (Model) |
|---|---|---|---|---|---|---|---|---|
| 1 | 4 | F | 13.0 | MR (PML shortening) | BWG synd | PDA ligation | 8/5/1974 | 0M (6320) |
| 2 | 4 | M | 17.0 | Prosthetic valve failure (thrombosis) | Corrected TGA | MVR (Björk-Shiley 23M) | 2/2/1977 | 1M (6120) |
| 3 | 1 | M | 8.0 | MR (leakage) | ··· | MVP (AML, PML cleft) | 1/17/1979 | 0M (6120) |
| 4 | 1 | F | 6.4 | MR (AML chordal elongation) | VSD | ··· | 4/22/1981 | 1M (6120) |
| 5 | 5 | F | 14.6 | MR (PML cleft) | ··· | Closure of ASD | 9/6/1982 | 1M (6120) |
| 6 | 5 | M | 17.0 | MR (congenital tricuspid) | ··· | ··· | 7/16/1984 | 1M (6120) |
| 7 | 5 | F | 19.0 | MR (AML prolapse) | ··· | ··· | 6/10/1985 | 1M (6120) |
| 8 | 5 | F | 17.5 | MR (AML chordal elongation) | ··· | ··· | 5/28/1986 | 0M (6120) |

AML = anterior mitral leaflet; ASD = atrial septal defect; BWG synd = Bland-White-Garland syndrome; F = female; M = male; MR = mitral regurgitation; MVP = mitral valve plasty; MVR = mitral valve replacement; PDA = persistent ductus arteriosus; PML = posterior mitral leaflet; SEBV = Starr-Edwards ball valve; TGA = transposition of great arteries; VSD = ventricular septal defect.

previous mitral valve repair, but later suffered progressive mitral regurgitation requiring reoperation. During SEBV replacement, a concomitant procedure (closure of a ventricular septal defect) was required in case 4. Three of the replaced SEBVs were size 0M and five size 1M. Model 6320 SEBV was used in case 1, and model 6120 was used in the other 7 patients. Follow-up was strictly obtained by monthly outpatient visits in this institution for 7 patients and by contact with the referring physician for 1 patient. Anticoagulation was maintained with warfarin (Eisai, Tokyo, Japan). The therapeutic goal was to maintain the thrombotest at 10% to 25% (international normalized ratio, 1.6 to 2.8). Prosthetic valve-related morbidity was defined based on the Society of Thoracic Surgeon guidelines for reporting morbidity and mortality after cardiac valvular operations [5].

## Results

The follow-up was 100%, conducted from 15 to 27 years (mean, 20). All patients grew to adulthood with normal weight gain and development (Table 2). No valve-related complications such as thromboembolic events, anticoagulant-related hemorrhage, or prosthetic valve endocarditis were seen.

During the follow-up period, the 3 patients who received a size 0M SEBV replacement experienced no valve-related complications and developed normally, but required re-replacement with a larger prosthesis 9 to 20 years after the initial operation because of relative mitral stenosis.

In case 1, 9.5 years after the initial MVR at age 13, cardiac catheter examination revealed an increased pressure gradient through the prosthetic valve. In addition,

*Table 2. Long-Term Results After Mitral Valve Replacement With Starr-Edwards Ball Valve*

| Case | Starr-Edwards Ball Valve Size | Age (y) | Follow-Up (y) | Height (cm) | Weight (kg) | Body Surface Area ($m^2$) | New York Heart Association Class | Cardiac-Thoracic Ratio (%) | Remarks |
|---|---|---|---|---|---|---|---|---|---|
| 1 | 0 M | 31 | 27 [9.5] | 154 | 42 | 1.37 | II | 81 | Relative MS (MVR with Starr-Edwards 2M, 6/20/1984) |
| 2 | 1 M | 27 | 24 | 170 | 53 | 1.63 | I | 51 | No complications |
| 3 | 0 M | 23 | 22 [19.5] | 165 | 52 | 1.58 | I | 51 | Relative MS (MVR with SJM 27, 6/9/1998) |
| 4 | 1 M | 21 | 20 | 161 | 52 | 1.54 | I | 46 | No complications |
| 5 | 1 M | 25 | 19 | 153 | 47 | 1.42 | I | 48 | No complications |
| 6 | 1 M | 22 | 17 | 177 | 52 | 1.64 | I | 44 | No complications |
| 7 | 1 M | 21 | 16 | 157 | 47 | 1.44 | I | 50 | No complications |
| 8 | 0 M | 19 | 15 [14.5] | 155 | 49 | 1.46 | I | 54 | Relative MS (MVR with Carpentier-Edwards 27, 2/14/2000) |

# = years after initial mitral valve replacement; MS = mitral stenosis; MVR = mitral valve replacement; SJM = St. Jude Medical.

coronary angiography showed that the left coronary artery originated from the pulmonary artery. Repeat MVR with the SEBV (size 2M, model 6120) and closure of the coronary arterial ostium from the pulmonary artery were conducted. Although this patient's postoperative course was uneventful, echocardiography showed mild paravalvular leakage with the second SEBV and moderate tricuspid regurgitation, which demanded careful follow-up.

Case 3, who was followed by a referring physician, suffered shortness of breath at age 20. Because a catheter examination showed relative mitral stenosis, re-MVR was conducted with a 27-mm St. Jude Medical valve (St. Jude Medical, St. Paul, MN) at another hospital. This patient's postoperative course was uneventful.

Case 8 is an 18-year-old woman with New York Heart Association functional class I at 14.5 years of follow-up. In October 2000, she was admitted to our hospital with varying levels of consciousness, but with no new focal transient or permanent neurologic findings. Brain computed tomography revealed no findings consistent with thromboembolic events. After treatment for heart failure with dopamine and diuretics, cardiac catheterization revealed a normal functioning mitral valve prosthesis, but the effective orifice was calculated as 0.66 $cm^2$ by the Gorlin formula (Edwards Lifesciences LLC, Irvine, CA). Re-MVR with a 27-mm Carpentier-Edwards bioprosthesis was accomplished, and the patient's postoperative course was uneventful.

All 5 patients undergoing size 1M SEBV replacement experienced no valve-related complications and achieved normal development. These patients are all in New York Heart Association functional class I and take no medication except their anticoagulant. The latest Doppler echocardiography at rest showed a mean transprosthetic valve pressure gradient of 18.5 mm Hg (range, 16.0 to 21.5 mm Hg).

## Comment

### *Prosthetic Valve Morbidity*

Starr-Edwards Model 6120 provides acceptable durability [6–9] as we previously reported [10]. No structural valvular deterioration such as poppet escape, ball variance, or cloth wear was observed in this series of 8 patients. In addition, no other valve-related complications such as thromboembolic events, anticoagulant-related hemorrhage, or prosthetic valve endocarditis were seen.

### *Management of Anticoagulant Therapy*

Maintaining appropriate anticoagulant therapy in children may be difficult. We have been fortunate to have good patient compliance and parental cooperation. We usually use the thrombotest (%) to monitor oral anticoagulant therapy in patients undergoing prosthetic valve replacement. The target thrombotest for MVR patients is 10% to 25%, equivalent to a prothrombin time international normalized ratio of 2.83 to 1.57 according to the regression equation [11]. This is a less intense therapeutic range than that adopted overseas. We saw neither documented embolic episodes nor anticoagulant-related hemorrhage during follow-up, suggesting that Asians may be less vulnerable to thrombotic disease than Caucasians [9, 11].

*Table 3. Valve Prosthesis and Valve Area Index*

| Starr-Edwards Ball Valve Size (Model) | Geometric Orifice Area ($cm^2$) | Valve Area Index ($cm^2/m^2$) | Range Valve Area Index ($cm^2/m^2$) |
|---|---|---|---|
| 0M (6320) | 1.54 | 1.24 | 0.97~1.24 |
| (6120) | 1.70 | 0.97, 1.05 | |
| 1M (6120) | 2.14 | 1.30, 1.31, 1.39, 1.49, 1.51 | 1.30~1.51 |

### *Valve Replacement in Children*

The prosthetic transvalvular gradient depends on valve size, mechanical efficiency, and blood flow rate, which is determined by the patient's size and activity. A perfectly functioning small size valve, for example, may produce an unacceptable transvalvular pressure gradient for an adult-sized adolescent. Dumesnil and Yoganathan [12] described the relationship between transprosthetic pressure gradients and effective orifice area indexed to body surface area, called valve area index. These authors demonstrated that a valve area index less than 1.3 $cm^2/m^2$ for MVR was associated with exponential increase in transprosthetic pressure gradients. According to the manufacturer's data, mounting diameters of size 0M and size 1M SEBV are 22 mm and 26 mm, respectively, and the respective effective orifice areas are 1.54 $cm^2$ and 2.14 $cm^2$. Thus there is a relatively large difference between sizes 0M and 1M. Valve area indexes with our patients are shown in Table 3. The range of valve area indexes of the patients replaced with smaller Starr-Edwards valve (size 0M) was 0.97 to 1.24 $cm^2/m^2$ at the time of the decision for replacement. Although these valves had been adequate earlier when the patients had less body surface area, the contribution of intimal growth and fibrous tissue encroachment may have additionally contributed to the reduced effective valve orifice. The problem of tissue overgrowth in valve dysfunction appears to be unique to the younger patient population. Although the 5 children who underwent MVR with the size 1M SEBV have not shown symptoms of relative mitral stenosis yet, continued follow-up is required, in spite of relatively stable body surface area.

There has been concern that placement of a small prosthetic valve in a child may inhibit annular growth and preclude larger valve replacement later. However, experimental studies in calves [13] that underwent MVR at 6 weeks old and were reexamined 1 to 3 years later demonstrated that the annulus had sufficiently grown to permit the insertion of a larger prosthesis. Braunwald and Castaneda [14] demonstrated that prostheses with a larger annular diameter could successfully be implanted after fibrous ridges associated with smaller initially

placed valves had been excised. These findings are supported by a large body of clinical experience since these reports. These results show that mitral annular diameter is not irrevocably determined by the placement of a valve substitute in early childhood.

This study does not specifically support the advantage of SEBV. We prefer the low-profile bileaflet mechanical valve in patients with a small ventricle. In clinical situations of the current era, we are faced with management of much younger and smaller patients than those encountered during the 1970s and early 1980s. Thus we need prosthetic valves of larger effective orifice area and lower profile. We believe that the data of valve area index in this study may serve as a reference.

In conclusion, we have reviewed our long-term results following MVR with the SEBV in children aged 5 years or younger. Model 6120 showed excellent durability, no thromboembolism, and no anticoagulant-related complications. The smaller valve (size 0M; valve area index, 0.97 to 1.24 $cm^2/m^2$) was adequate to permit patients to reach adolescence, but required elective re-replacement with a larger valve.

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