Current Management of Severe Congenital Mitral Stenosis
Outcomes of Transcatheter and Surgical Therapy in 108 Infants and Children

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Background—Severe congenital mitral stenosis (MS) is a rare anomaly that is frequently associated with additional left heart obstructions. Anatomic treatments for congenital MS include balloon mitral valvuloplasty (BMVP), surgical mitral valvuloplasty (SMVP), and mitral valve replacement (MVR), although the optimal therapeutic strategy is unclear.

Methods and Results—Between 1985 and 2003, 108 patients with severe congenital MS underwent BMVP or surgical intervention at a median age of 18 months (range 1 month to 17.9 years). Anatomic subtypes of MS were “typical” congenital MS in 78 patients, supravalvar mitral ring in 46, parachute mitral valve in 28, and double-orifice mitral valve in 11, with multiple types in ~50% of patients. Additional left heart anomalies were present in 82 patients (76%). The first MS intervention was BMVP in 64 patients, SMVP in 33, and MVR in 11. BMVP decreased peak and mean MS gradients by a median of 33% and 38%, respectively (P<0.001), but was complicated by significant mitral regurgitation in 28%. Cross-sectional follow-up was obtained at 4.8±4.2 years. Overall, Kaplan-Meier survival was 92% at 1 month, 84% at 1 year, and 77% at 5 years, with 69% 5-year survival during the first decade of our experience and 87% since (P=0.09). Initial MVR and younger age were associated with worse survival. Survival free from failure of biventricular repair or mitral valve reintervention was 55% at 1 year among patients who underwent BMVP and 69% among patients who underwent supravalvar mitral ring resection initially. Among patients who underwent BMVP, survival free from failure of biventricular repair or MVR was 79% at 1 month and 55% at 5 years, with worse outcome in younger patients and those who developed significant postdilation mitral regurgitation.

Conclusions—BMVP effectively relieves left ventricular inflow obstruction in most infants and children with severe congenital MS who require intervention. However, surgical resection is preferable in patients with MS due to a supravalvar mitral ring. Five-year survival is relatively poor in patients with severe congenital MS, with worse outcomes in infants and patients undergoing MVR, but has improved in our more recent experience. Many patients have undergone second procedures for either recurrent/residual MS or mitral regurgitation resulting from dilation-related disruption of the mitral valve apparatus. (Circulation. 2005;112:707-714.)

Key Words: mitral valve ♦ catheterization ♦ heart defects, congenital ♦ pediatrics ♦ balloon

Congenital mitral stenosis (MS) is a rare and morphologically heterogeneous lesion that affects both the leaflets and subvalvar tension apparatus of the mitral valve (MV). Frequently, congenital MS occurs in conjunction with additional left heart obstructions and/or a ventricular septal defect (VSD), which may complicate its management. In severe cases, congenital MS is difficult to manage medically and entails significant morbidity and mortality. Interventional therapies for medically refractory congenital MS include percutaneous transcatheter balloon mitral valvuloplasty (BMVP), surgical mitral valvuloplasty (SMVP), and mitral valve replacement (MVR). The optimal interventional treatment for congenital MS is unknown and in any given patient may depend on the severity and anatomic substrate(s) of MS, associated cardiovascular anomalies, and patient size. Although MVR can relieve left ventricular (LV) inflow obstruction, with little risk of procedure-related mitral regurgitation (MR), it carries high morbidity and mortality, particularly in infants and young children. SMVP may improve MV mobility and effective inflow area, but the success of plastic surgical procedures on the MV may vary according to the substrate of MS. In young children undergoing surgical intervention on the MV leaflet(s) and/or subvalvar apparatus for congenital MS, early mortality, morbidity, and need for MVR are relatively common. In contrast, surgical resection is typically an effective treatment for a supravalvar mitral
Balloon Mitral Valvuloplasty offers a less invasive alternative to SMVP or MVR for treatment of congenital MS in patients whose conditions are refractory to medical management. However, published data on BMVP for congenital MS are limited to small series with short-term follow-up. To determine outcomes of surgical and transcatheter management of severe congenital MS, and to assess whether outcomes have improved over time, we analyzed our experience with 108 infants and children treated for severe congenital MS at our institution since 1985.

**Methods**

**Patients**

The database of the Department of Cardiology was queried for patients with severe congenital MS who underwent BMVP, SMVP, or MVR for congenital MS at ≤18 years of age between 1985 and 2003. Institutional criteria for intervention in patients with congenital MS during this period included significant symptoms of congestive heart failure despite medical therapy, failure to thrive, and/or systemic right ventricular pressure. Patients with rheumatic MS, with postoperative MS after repair of an atrioventricular canal defect, or who had undergone interventions for congenital MS before referral to our center were excluded. The study was approved by the Children’s Hospital Committee for Clinical Investigation.

**Anatomic and Hemodynamic Evaluation**

Preintervention and postintervention echocardiograms were reviewed to determine the anatomic substrate(s) of MS, mean and maximum Doppler-derived transmural gradients, severity of MR, and presence and severity of associated cardiovascular anomalies. In patients with catheterization data available, simultaneous left atrial (LA) and LV pressures were used to calculate mean and peak (LA A-wave–LV end-diastolic pressure) MS gradients. When pressure tracings were available, the effective orifice area of the MV was calculated according to the Gorlin method.22

**Balloon Mitral Valvuloplasty**

The basic technical details of BMVP in our practice have been described previously.21,22 Since these descriptions were published, several modifications have been adopted. The technique of snaring a guidewire in the aorta to direct sheaths/catheters across the MV is no longer employed. Improved sheath and catheter technology and preshaped curves in stiff wires have obviated this strategy, which has the potential to damage the MV or aortic valve. The size of the initial dilating balloon is determined by patient size rather than by the diameter of the MV anulus; the starting balloon diameter is typically 8 mm in patients with a body surface area ≤0.4 m², 10 mm in patients ≤0.4 to 0.8 m², 12 mm in patients ≤0.9 to 1.2 m², and 15 mm in patients ≥1.2 m². Balloon size is also varied by modulation of the inflation pressure. Thus, each balloon is generally inflated twice, first at low pressure (2 to 4 atm), which yields an effective diameter less than the stated balloon size, and then again at a higher pressure, which allows relatively fine gradation of inflation diameter. Most patients undergo at least 3 or 4 dilations before a satisfactory result is achieved.

**Data Analysis**

Acute outcomes assessed included relief of MS, development of moderate or severe MR, and in-hospital mortality. Time-dependent outcomes included survival, transplantation-free survival with a biventricular circulation, and several measures of reintervention-free survival. For comparison of preintervention and postintervention hemodynamic indices, paired t test analysis was used. For univariable analysis of factors associated with acute outcome and between-group comparisons of means and proportions, either independent-samples t test or 1-way ANOVA, and either χ² analysis or Fisher’s exact test were used, respectively, as appropriate. Time-dependent outcomes were assessed with the Kaplan-Meier product limit method. Factors associated with time-dependent outcomes were analyzed with Cox proportional hazards regression. In general, for Kaplan-Meier and Cox analyses of reintervention-free survival, death, conversion to a univentricular circulation, and cardiac transplantation were treated as events; however, in certain instances, patients were censored event-free at the time of interventions that precluded the event of interest (eg, in analysis of freedom from repeat BMVP, patients were censored at the time of MVR). Data are presented as mean±SD or median (range).

**Results**

**Patients**

Between 1985 and 2003, 108 patients ≤18 years of age underwent BMVP or surgical intervention for congenital MS at a median age of 18 months (range 1 month to 17.9 years). Thirty of these patients were included in prior reports.21,29 Demographic and anatomic details are summarized in Table 1.

The most common anatomic variant of MS (78 patients, 72%) was typical congenital MS,1 with thickened leaflets, short or absent chordae tendineae, obliteration of interchordal spaces, and 2 separate but often closely spaced papillary muscles. An SVMR was present in 46 patients (43%), a parachute MV in 28 (26%), and a double-orifice MV in 11 (10%), with >1 anatomic subtype of MS in nearly 50% of patients.

Additional cardiovascular defects were present in 86% of patients (n=93), and additional left heart anomalies were present in 76% (n=82). A VSD was present in 38 patients (35%), who were younger at the time of MS intervention than patients without a VSD (P=0.02) and more likely to have an SVMR (56%) or parachute MV (50%) than typical congenital MS (27%, P=0.01). A total of 91 prior cardiovascular interventions had been performed in 62 patients (57%; Table 1).

**Preintervention Echocardiographic and Hemodynamic Data**

Baseline echocardiographic and hemodynamic data are summarized in Table 1.

**Interventions**

The initial intervention for congenital MS was BMVP in 64 patients (59%), SMVP in 33 (31%), and MVR in 11 (10%). In 38 of the 44 patients treated initially with surgical intervention, preoperative cardiac catheterization was performed, but BMVP was not attempted. In almost all patients who underwent initial SMVP or MVR rather than BMVP, there were circumstances that favored a surgical approach, including SVMR as a major substrate of MS (n=35) and/or the need for associated surgery (n=26, 59%), including VSD closure (n=15), relief of subaortic obstruction (n=14), MVR/SMVP for associated moderate MR (n=2), aortic valve replacement (n=1), and resection of right ventricular muscle bundles (n=1). There were no significant demographic differences between patients undergoing BMVP and surgical intervention, but there were several diagnostic and clinical differences (Table 1). Relatively more patients underwent SMVP during the latter half of our experience (Table 1), after analysis of our earlier experience led us to stop performing BMVP in patients with SVMR as a major MS substrate.
<table>
<thead>
<tr>
<th>Variable</th>
<th>Initial BMVP (n=64)</th>
<th>Initial SMVP (n=33)</th>
<th>Initial MVR (n=11)</th>
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</thead>
<tbody>
<tr>
<td>Demographic details</td>
<td></td>
<td></td>
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<tr>
<td>Age at intervention</td>
<td>17 (1−190)</td>
<td>23 (1−215)</td>
<td>19 (3−137)</td>
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<tr>
<td>&lt;1 year, n (%)</td>
<td>24 (38)</td>
<td>14 (42)</td>
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<td>&lt;2 years, n (%)</td>
<td>37 (58)</td>
<td>18 (55)</td>
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<td>&lt;10 years, n (%)</td>
<td>60 (94)</td>
<td>27 (82)</td>
<td>10 (91)</td>
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<td>Median weight (range), kg</td>
<td>8.4 (2.3−69)</td>
<td>7.9 (3.0−74)</td>
<td>8.5 (3.6−24)</td>
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<td>Weight &lt;5th percentile for age, n (%)</td>
<td>40 (63)</td>
<td>19 (57)</td>
<td>8 (73)</td>
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<tr>
<td>Intervention after 1994, n (%)</td>
<td>26 (41)</td>
<td>22 (66)</td>
<td>4 (36)</td>
</tr>
<tr>
<td>Anatomic details, n (%)</td>
<td></td>
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<td></td>
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<tr>
<td>Anatomic substrate of MS</td>
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<tr>
<td>Typical congenital MS</td>
<td>56 (87)a</td>
<td>15 (45)</td>
<td>7 (64)</td>
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<tr>
<td>Parachute MV</td>
<td>9 (16)b</td>
<td>16 (49)</td>
<td>3 (27)</td>
</tr>
<tr>
<td>SVMR</td>
<td>11 (17)b</td>
<td>30 (91)</td>
<td>5 (46)</td>
</tr>
<tr>
<td>DOMV</td>
<td>9 (14)</td>
<td>1 (3)</td>
<td>1 (9)</td>
</tr>
<tr>
<td>Combination of multiple MS substrates</td>
<td>20 (31)b</td>
<td>29 (88)</td>
<td>4 (36)</td>
</tr>
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<td>Associated anomalies and prior interventions, n (%)</td>
<td></td>
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<td></td>
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<tr>
<td>Additional cardiovascular anomalies</td>
<td>52 (81)</td>
<td>30 (91)</td>
<td>11 (100)</td>
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<tr>
<td>Additional left heart anomalies</td>
<td>44 (69)b</td>
<td>27 (82)</td>
<td>11 (100)</td>
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<td>Associated VSD</td>
<td>15 (23)b</td>
<td>20 (61)</td>
<td>3 (27)</td>
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<td>Prior cardiovascular interventions</td>
<td>31 (48)b</td>
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<td>COA repair or balloon dilation</td>
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<td>17 (52)d</td>
<td>6 (55)d</td>
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<td>Balloon or surgical aortic valvuloplasty</td>
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<td>4 (12)f</td>
<td>3 (27)f</td>
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<td>1 (9)h</td>
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<td>1 (9)</td>
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<td>1 (2)</td>
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<td>MS gradients</td>
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<td>Doppler maximum gradient</td>
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<td>23.7±5.9</td>
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<td>Doppler mean gradient</td>
<td>13.9±4.6a</td>
<td>11.9±3.9</td>
<td>11.6±3.0</td>
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<td>Catheterization peak gradient</td>
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<td>22.1±7.4</td>
<td>26.0±5.4</td>
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<tr>
<td>Catheterization mean gradient</td>
<td>14.9±4.3</td>
<td>15.1±4.9</td>
<td>17.3±5.3</td>
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<tr>
<td>Moderate or severe MR</td>
<td>4 (6)</td>
<td>1 (3)</td>
<td>1 (9)</td>
</tr>
<tr>
<td>Mean LA pressure</td>
<td>24.1±5.8</td>
<td>24.6±5.9</td>
<td>25.6±3.9</td>
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<tr>
<td>LV end-diastolic pressure</td>
<td>12.7±5.2</td>
<td>12.4±5.0</td>
<td>11.3±2.6</td>
</tr>
<tr>
<td>Mean pulmonary artery pressure</td>
<td>44.5±14.9</td>
<td>47.0±18.5</td>
<td>49.1±13.6</td>
</tr>
</tbody>
</table>

*aSignificantly more common/higher in patients who underwent BMVP than in those who underwent surgical intervention (all P<0.02).

*bSignificantly less common in patients who underwent BMVP than in those who underwent surgical intervention (all P<0.02).

*cSurgical repair in 18 patients and balloon dilation in 1.

*dTwenty-eight prior interventions for coarctation of the aorta in these 23 patients: surgical repair in 22, primary balloon dilation in 1, and balloon dilation for recurrent coarctation of the aorta in 5.

*eFourteen interventions for aortic stenosis in these 10 patients: 11 balloon aortic valvuloplasty procedures and 3 surgical aortic valvuloplasty/valvotomy procedures.

*fTen transcatheater balloon aortic valvuloplasty procedures in these 7 patients.

*gThree interventions in this 1 patient.

*hTwo interventions in this 1 patient.

*iRepair of a hole in the MV leaflet after resection of subaortic stenosis.

*jTetralogy of Fallot repair in 1 patient, pacemaker placement for postoperative heart block after resection of subaortic stenosis in 2, atrial septal defect closure in 2, balloon atrial septostomy in 1, and vascular ring division in 1.

*kHemodynamic data are expressed as mean±SD mm Hg.

*lCatheterization data on peak and mean MS gradients were available for 23 and 28 patients, respectively, in the surgical intervention group.

*mCatheterization data on LA pressure, LV end-diastolic pressure, and pulmonary artery pressure available for 30 patients in the surgical intervention group.
Otherwise, there was no significant change in the distribution of intervention type over the course of our experience.

**Balloon Mitral Valvuloplasty**
BMVP was the first intervention in 64 patients. A balloon was passed through the MV and inflated in all 64 patients, with an average of 2.4±1.0 (median 2, range 1 to 5) balloon sizes and 3.9±2.4 (median 3.5, range 1 to 12) balloon inflations per patient. Additional interventions were performed in 12 patients (19%) during the same catheterization, including balloon aortic valvuloplasty in 7, embolization of a ductus arteriosus in 3, stenting of recurrent coarctation of the aorta in 1, and balloon dilation of a stenotic triple-orifice tricuspid valve in 1. Although we stopped performing BMVP in patients with an SVMR as a major cause of MS during the latter half of our experience, 3 patients with a minor SVMR along with typical MS underwent BMVP during that period.

**Surgical Mitral Valvuloplasty**
Of 33 patients undergoing SMVP, 91% (n=30) had an SVMR that was resected. Other valvuloplasty techniques were performed in 9 patients (6 who underwent SVMR resection), including commissurotomy or commissuroplasty in 5, leaflet thinning in 2, and chordal fenestration and/or resection in 5. Additional interventions were performed during the same operation in 19 patients (58%), including VSD closure in 13 (with pulmonary artery band removal in 2), relief of subaortic stenosis in 6, aortic valvuloplasty in 3, aortic valve replacement in 1, and coarctation of the aorta repair in 1. All 3 patients who did not undergo SVMR resection underwent concurrent VSD closure and/or procedures on the LV outflow tract.

**Mitral Valve Replacement**
Eleven patients underwent MVR. Five (45%) of the implanted valve prostheses were 17 mm or less, and 8 (72%) were implanted in the supraannular position. Additional interventions were performed during the same operation in 7 patients (64%), including relief of subaortic stenosis in 5, VSD closure in 2, aortic valve replacement in 1, resection of right ventricular muscle bundles in 1, and patch augmentation of the LA in 1. The 4 patients who did not undergo a concurrent procedure either had an SVMR (n=3) or were thought to have an SVMR (n=1), and underwent MVR because SVMR resection alone did not adequately relieve MS.

**Outcomes**

**Early MV Function After BMVP or SMVP**

**Balloon Mitral Valvuloplasty**
BMVP decreased the peak MS gradient by a median of 33% (from 21.8±7.1 to 13.3±7.5 mm Hg) and the mean gradient by a median of 38% (from 15.1±4.5 to 9.1±5.5 mm Hg; both P<0.001; Figure 1). Maximum and mean Doppler gradients after BMVP were 20% and 40% lower (median), respectively, than before intervention (both P<0.001; Figure 1). Calculated effective MV orifice area data were available both pre- and post-BMVP for 25 of 64 patients, in whom it increased by 74±69%, from 0.9±0.3 to 1.6±0.7 cm²/m² (P<0.001). Mean LA pressure decreased from 24.0±5.8 to 21.6±6.9 mm Hg (P=0.009), LV end-diastolic pressure increased from 12.7±5.2 to 16.4±7.7 mm Hg (P<0.001), and mean pulmonary artery pressure did not change acutely.

Moderate or severe MR developed in 17 patients (28% of 60 patients without preexisting moderate or severe MR) after BMVP. A tear in the MV, disruption of MV chordal structures, and/or a partially flail MV leaflet was visualized by echocardiography in 17 patients.

None of the patient-related or procedural variables analyzed were significantly associated with the degree of MS reduction or with new postdilation moderate or severe MR. However, 6 of 11 patients with SVMR who underwent BMVP (8 during the first half of our experience) developed significant MR.

**Surgical Mitral Valvuloplasty**
Postoperative Doppler MS gradients were available for 21 of the 32 early survivors of SMVP. The maximum and mean MS gradients were reduced by 68% and 70% (median), respectively, from 20.1±6.3 to 9.4±3.7 mm Hg and from 11.9±3.9 to 5.1±2.0 mm Hg (P<0.001). Moderate or severe MR developed after SMVP in 4 patients (12%), 3 of whom underwent SVMR resection along with commissuroplasty.

**Death or Failure of Biventricular Repair**
There were 7 early deaths (7%): 3 after BMVP (5% of 64), 1 after SMVP (3% of 33), and 3 after MVR (27% of 11). At cross-sectional follow-up of 4.8±4.2 years among the 101 early survivors (including 5 patients lost to follow-up), 14 additional patients had died, 9 within 1 year of the initial intervention. Eight of the 14 deaths during follow-up occurred acutely after reintervention (see below). Thus, 21 of 108 patients were known to have died at most recent

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Figure 1. Changes in mean (A) catheterization (Cath) and (B) Doppler echocardiography (Echo) gradients before and after BMVP for severe congenital MS. Line charts depict directly measured predilation and post dilation MS gradient for each patient, along with box plots representing median (central line), 25th–75th percentile (box), and 10th–90th percentile (error bars) MS gradient for entire patient cohort. ● indicates initial BMVP procedures; ○, reinterventional BMVP.
follow-up, including 13 of 64 patients treated initially with BMVP, 3 of 33 treated with SMVP, and 5 of 11 who underwent initial MVR. Failure of biventricular repair occurred in 6 patients, who either underwent conversion to a univentricular circulation or heart transplantation. Three patients were converted to a univentricular circulation within 2 months of BMVP, with a stage I palliation performed in 2 patients, at 3 and 6 months of age, and bidirectional Glenn anastomosis in 1 patient at 15 months of age. Three other patients underwent heart transplantation 1 to 3 years after BMVP (n = 64) or MVR (n = 1), at 1.5 to 3.5 years of age. One of the patients with a stage I procedure and 2 of the patients who received transplants died in the early postoperative period. Thus, a total of 24 patients had failure of biventricular repair and/or died during follow-up. Overall, Kaplan-Meier survival was 92% at 1 month, 84% at 1 year, and 77% at 5 years (Figures 2 and 3). Before 1994, survival was 89% at 1 month, 80% at 1 year, and 69% at 5 years, and from 1994 to 2003, it was 95% at 1 month, 91% at 1 year, and 87% at 5 years. There was a trend toward improved survival among patients treated during the latter half of our experience, with 87% 5-year survival since 1994 (P = 0.09; Figure 2B). Survival free from failure of biventricular repair was 89% at 1 month, 82% at 1 year, and 74% at 5 years. Table 2 summarizes the current status of patients according to initial BMVP or surgical repair and by age at initial repair.

Patients undergoing initial MVR were at increased risk of early death (P = 0.01), but within treatment groups, there were no predictors of early mortality. Independent predictors of decreased survival over time included age < 2 years at initial MV intervention (Figure 2A; P = 0.001) and MVR as the initial MV intervention (P = 0.02). Among patients who underwent BMVP, independent predictors of worse survival included younger age (continuous, P = 0.03) and higher predilation LA pressure (P = 0.05).

**Reintervention**

At cross-sectional follow-up of 4.8 ± 4.2 years, 44 patients had undergone 61 reinterventions on the MV, 29 within 1 year of the initial intervention, with 8 early deaths after reintervention (2 after early reintervention, included above as early deaths). The first reintervention was primarily for MS in 26 patients and for MR with or without significant residual MS in 18. Among patients managed initially with BMVP, 34 underwent 45 MV reinterventions, including BMVP in 13 patients, SMVP in 8, and MVR in 18. Eight patients treated initially with SMVP underwent 14 MV reinterventions, including repeat SMVP in 2 patients, BMVP in 2, and MVR in 6. Redo MVR was performed in 2 patients who underwent...
initial MVR. Kaplan-Meier survival free from failure of biventricular repair or MV reintervention was 76% at 1 month, 55% at 1 year, and 39% at 5 years among patients who underwent BMVP (Figure 3A) and 96% at 1 month and 69% at 1 and 5 years among patients who underwent SVMR resection initially (Figure 3B). Among patients who underwent BMVP, independent risk factors for decreased survival free from MV reintervention included postdilation moderate or severe MR ($P<0.001$) and younger age ($P<0.01$). No risk factors were identified for decreased reintervention-free survival among patients treated with SMVP.

**BMVP or SMVP**

Among 15 patients treated with reinterventional BMVP for residual/recurrent MS, improvement in MS was similar to initial BMVP, with median peak and mean gradient reductions of 38% and 41% ($P<0.001$; Figure 1). Effective MV orifice area ($n=7$) increased by 82±24% ($P<0.001$). There were 2 early deaths after repeat BMVP, and significant MR developed in 3 patients (19%).

Eleven patients underwent some form of SMVP after BMVP or SVMR resection, for residual/recurrent MS in 6 patients, MS and MR in 2, and primarily MR in 3. Of the 3 patients who underwent SMVP after prior SVMR resection, only 1 was for a recurrent/residual SVMR.

**Mitral Valve Replacement**

MVR after primary BMVP ($n=18$) or SMVP ($n=6$) was performed during follow-up in 24 patients, either for combined MR and MS ($n=19$) or for MS without significant MR ($n=5$), with 1 early death and 1 case of postoperative heart block. MVR was supraannular in 17 patients (71%). Survival free from failure of biventricular repair or MVR is depicted in Figures 3A and 3B. Among patients treated initially with BMVP, independent risk factors for decreased survival free from failure of biventricular repair or MVR included significant postdilation MR ($P=0.001$) and younger age ($P=0.01$). Including the 11 primary and 24 reinterventional procedures, 35 of 108 patients underwent MVR at a median age of 22 months (range 3 months to 17 years). At follow-up, repeat MVR had been performed in 9 of these 35 patients, 2 for prosthesis dysfunction and 7 for upsizing of the prosthesis, with 2 early deaths and 1 case of postoperative heart block. Kaplan-Meier survival after MVR (including initial and reinterventional procedures, but not redo MVR) was 87% at 1 month, 77% at 1 year, and 60% at 5 years.

**Other Reinterventions**

Aside from MV reinterventions, conversion to a univentricular circulation, and cardiac transplantation, 25 patients underwent 44 additional cardiovascular interventions during follow-up, including relief of subaortic stenosis in 8 patients, balloon or surgical aortic valvuloplasty in 6, aortic valve replacement in 5, balloon angioplasty or surgical repair for coarctation of the aorta in 5, VSD closure in 5, pacemaker placement for postoperative heart block after MVR or resection of subaortic stenosis in 4, and other procedures in 4.

**Follow-Up MV Function**

At follow-up of $4.8±4.2$ years, 60 of 108 patients were alive with a biventricular circulation and their native MV (ie, they had not undergone MVR). In these patients, the peak and mean Doppler MS gradients were $17.1±7.2$ and $7.2±4.3$ mm Hg, respectively, which did not differ significantly from the early postintervention period.

**Complications**

**Balloon Mitral Valvuloplasty**

Including reinterventions after initial BMVP or SMVP, 80 BMVP procedures were performed in 66 patients. Aside from early deaths and iatrogenic MR, significant adverse events occurred in 16 patients, including cardiac arrest in 4 (2 early deaths after repeat BMVP), atrial ($n=2$) or ventricular ($n=1$) perforation in 3, transient rhythm abnormalities in 11, stroke

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**TABLE 2. Current Status Among Patients Undergoing Intervention for Severe Congenital MS**

<table>
<thead>
<tr>
<th>Current Status*</th>
<th>Initial BMVP (n=64)</th>
<th>Initial Surgical Intervention (n=44)</th>
<th>Age &lt;2 y (n=64)</th>
<th>Age ≥2 y (n=44)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Died, n (%)</td>
<td>13 (20)</td>
<td>8 (18)</td>
<td>19 (30)</td>
<td>2 (4)</td>
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<td>&lt;1 mo</td>
<td>3</td>
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<tr>
<td>1–5 y</td>
<td>2</td>
<td>2</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>&gt;5 y</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Alive, single ventricle or heart transplant, n (%)</td>
<td>3 (5)</td>
<td>0 (0)</td>
<td>3 (5)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Alive, no MV reinterventions, n (%)</td>
<td>25 (39)</td>
<td>29 (66)</td>
<td>25 (39)</td>
<td>29 (66)</td>
</tr>
<tr>
<td>Symptomatic improvement</td>
<td>23</td>
<td>28</td>
<td>23</td>
<td>28</td>
</tr>
<tr>
<td>Symptomatic, awaiting MV reintervention</td>
<td>2</td>
<td>1</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Alive, MV reinterventions, n (%)</td>
<td>23 (36)</td>
<td>7 (16)</td>
<td>17 (27)</td>
<td>13 (30)</td>
</tr>
<tr>
<td>&lt;1 mo</td>
<td>6</td>
<td>1</td>
<td>6</td>
<td>1</td>
</tr>
<tr>
<td>1 mo to 1 y</td>
<td>6</td>
<td>3</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>1–5 y</td>
<td>7</td>
<td>1</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>&gt;5 y</td>
<td>4</td>
<td>2</td>
<td>3</td>
<td>3</td>
</tr>
</tbody>
</table>

*<1 mo, 1 mo to 1 y, 1–5 y, and >5 y indicate duration after initial MV intervention.
in 1, and a femoral arteriovenous fistula in 1, who developed extensive venous thrombosis of the right leg and underwent embolization of the fistula.

**SMVP or MVR**
Overall, 44 patients underwent SMVP and 35 underwent 44 MVR procedures. Aside from death and MR requiring MVR, the most notable adverse events in this cohort were postoperative heart block requiring a pacemaker, which occurred in 3 patients after MVR (7% of MVR procedures), and 1 episode of subacute prosthesis thrombosis requiring emergent redo MVR 2 months after the initial MVR.

**Discussion**
Severe congenital MS is a rare and challenging condition for which the optimal treatment is unknown. Between 1985 and 2003, 108 infants and children—the majority young, failing to thrive, and with additional left-sided cardiovascular anomalies—underwent transcatheter or surgical intervention for symptomatic congenital MS not responsive to medical therapy. In general, BMVP was our first-line therapy for most forms of congenital MS, whereas surgery was typically reserved for patients with SVMR, patients with associated intracardiac lesions necessitating cardiotomy (such as a VSD or subaortic stenosis), and patients with significant MR at baseline. Overall, 1-month survival was 92%, and intermediate survival with a biventricular circulation was relatively poor (74% at 5 years). However, there was a trend toward improved survival during the latter half of our experience, with 5-year survival of 87% from 1995 to 2003 compared with 69% between 1985 and 1994. Reinterventions on the MV were common. The majority of deaths and reinterventions occurred within 1 year of the first procedure, reflecting a subgroup of patients who received minimal or no benefit from the initial MV intervention. Another sizable subset of patients was free from reintervention at most recent follow-up or underwent reintervention at least 5 years after the initial procedure, which indicates substantial and possibly definitive relief after the initial procedure. As might be expected, younger age at the time of primary intervention was associated with both worse survival and need for reintervention.

**BMVP for Congenital MS**
The goal of BMVP for children with congenital MS is to reduce LV inflow obstruction and LA pressure, hopefully producing lasting relief but at a minimum improving symptoms and delaying MVR until the patient is older and larger. In this regard, BMVP is effective. Among 64 children undergoing BMVP in this series, the MS gradient was reduced by a median of ~40%, and the calculated effective MV orifice area almost doubled. More than half of the patients received symptomatic improvement sufficient to avoid further MV intervention for at least 1 year, and 40% were alive and free from MV reintervention 5 years after the initial procedure for MS. A substantial number of patients were free from significant recurrent MS up to 13 years after BMVP, and survival without MVR was 55% at 5 years. Nevertheless, BMVP is not curative, and even when successful acutely, it does not provide definitive relief of MS in many of these high-risk patients: 19 patients required repeat BMVP (n = 13) or surgical reintervention (n = 6) for residual/recurrent MS, 10 within 1 year of the initial procedure, with younger patients at higher risk for reintervention. In an earlier analysis, we found that BMVP was not effective for patients with SVMR as a major substrate of MS and frequently resulted in MR. Thus, in the latter half of our experience, BMVP was performed in only 3 patients with SVMR, in whom the major cause of LV inflow obstruction was typical MS.

Although early mortality after BMVP was only 5%, decreasing the LV inflow gradient with BMVP may entail a significant cost, namely, tearing of an MV leaflet or disruption of the tension apparatus, resulting in moderate or severe MR, which occurred in nearly 30% of patients in the present series. Many of these patients required MVR, typically within 1 year of BMVP, but they were not at increased risk of poor outcome after MVR compared with patients who underwent MVR as a first MS intervention (there was only 1 early death among 18 patients undergoing MVR after BMVP).

The mechanism by which BMVP increases the effective MV orifice area in patients with congenital MS is not clear and almost certainly varies according to the particular anatomic features of the MV. Despite the fact that significant MR due to leaflet tears or disruption of the tension apparatus occurred in a substantial subset of patients, the therapeutic benefit of BMVP in this population most likely derives from such tears. In the malformed MV in patients with congenital MS, the chordal apparatus is grossly abnormal, and the functional contribution of the various chordal structures is less clear than in the normal heart. Although it is difficult to predict whether BMVP-induced tears in MV leaflets/tension apparatus are therapeutic or detrimental, our impression has been that gradually increasing balloon size and inflation pressure decreases the likelihood of inducing significant MR.

**Surgical Therapy for Congenital MS**
The initial intervention for MS was SMVP or MVR in 44 of the patients in the present series, and 35 others underwent MV surgery after prior BMVP or SMVP. Surgical intervention was undertaken for 1 of 2 primary reasons: the presence of an SVMR as a major substrate of MS, or residual MS and/or dilation-induced MR after BMVP. A small cohort of patients underwent primary surgical intervention for MS in the absence of an SVMR, in all cases because surgery was indicated for treatment of hemodynamically important associated anomalies such as subaortic stenosis or a VSD, or because there was significant associated significant MR.

Surgical resection is the preferred method of treatment for SVMR. Whereas resection is effective and safe, with little risk of significant MR, the risk–benefit ratio of BMVP in such patients is relatively low, because BMVP is unlikely to have a significant mechanical effect on an SVMR and is more likely than resection to induce MR. Among 33 patients with SVMR in the present series, resection was almost universally successful in improving excursion of the affected leaflet(s) and relieving LV inflow obstruction, as other investigators have reported. Significant MR developed after SVMR resection in only 10% of patients, all
of whom underwent concurrent commissuroplasty, and recurrent
SVMR requiring intervention was uncommon, occurring in only
1 patient, in contrast to the experience of Tulloh et al.16
Among 35 infants and children who underwent a total of
44 MVR procedures, 1- and 5-year survival rates were 78%
and 57%, respectively, and 3 patients developed heart block
necessitating pacemaker placement. In the majority of pa-
patients, supraannular implantation of the prosthesis was
required. Although survival among children undergoing MVR
appears to be improving, we believe that MVR should be
avoided as first-line intervention for congenital MS whenever
possible. MVR is particularly undesirable in infants, who
often require supraannular implantation of the MV prosthesis,
which limits LA compliance, resulting in LA and pulmonary
artery hypertension even in the absence of prosthetic valve
obstruction25 and which has been associated with higher
mortality than annuloplasty.5

Conclusions
BMVP effectively relieves LV inflow obstruction in the
majority of cases and is our preferred mode of treatment in
most infants and children with severe congenital MS due to
typical MS, double-orifice MV, or parachute MV. In patients
with SVMR as the primary substrate of MS, however,
resection is preferred. Patients with significant associated MR
and/or hemodynamically important associated anomalies re-
quiring cardiectomy (eg. VSD or subaortic stenosis) are re-
ferred for surgical treatment. In such cases, maintenance of
the native MV is prioritized, but the decision between SMVP and
MVR ultimately depends on intraoperative assessment of the
reparability of the MV. With respect to reinterventional
procedures, patients with recurrent MS are typically referred
for repeat BMVP, unless the initial BMVP was ineffective or
significant MR is present, in which case surgery is performed,
with the decision between SMVP and MVR dependent on
intraoperative evaluation of the MV. Unfortunately, owing to
baseline differences between patients treated with BMVP,
SMVP, and MVR in the present series, our data do not allow
for conclusions about the relative outcome benefits of any
treatment strategy over another.

Procedural mortality is uncommon in children undergoing
BMVP or SMVP for congenital MS, and intermediate sur-
vival continues to improve. A substantial proportion of our
patient population has remained free from reintervention,
with no recurrence of MS, but many patients, particularly
those who underwent BMVP, required reintervention for
either recurrent/residual MS or MR. Ultimately, additional
follow-up will be required to assess long-term outcomes and
optimal application of BMVP and SMVP for infants and
children with severe congenital MS.

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