Surgical Management of Mitral Regurgitation After Repair of Endocardial Cushion Defects
Early and Midterm Results

Adrian M. Moran, MB, BCh; Sabine Daebritz, MD; John F. Keane, MD; John E. Mayer, MD

Background—Mitral regurgitation (MR) represents the principal indication for reoperation in patients after repair of atroioventricular septal defects (AVSD). Reports of mitral valvuloplasty (MVP) in such patients are few; the alternative, mitral valve replacement (MVR), necessitates commitment to future valve replacement and long-term anticoagulation. We sought to determine the outcome of those patients who underwent either MVP or MVR between January 1, 1988, and December 31, 1998, for significant MR after repair of AVSD. Furthermore, we sought to identify (a) morphological predictors necessitating MVR, and (b) predictors of future reoperation within the MVP group.

Methods and Results—Retrospective review of clinical, operative, and echocardiographic data were performed. There were 46 patients identified (37 MVP and 9 MVR). The median age at initial AVSD repair was 0.6 years, and the age at subsequent mitral valve operation was 2.8 years. The early postoperative mortality rate was 2.2%, and survival at 1 and 10 years was 89.9% and 86.6%, respectively. A high rate of complete heart block was noted within the MVR group (37.5%). Freedom from later mitral valve reoperation for both groups was similar. No significant morphological predictors necessitating MVR were found. Predictors of reoperation within the MVP group included the presence of moderate or worse MR in the early postoperative period. In both groups New York Heart Association class, degree of MR, growth, and ventricular volumes improved.

Conclusions—Mitral valve surgery significantly improves clinical status, with a sustained improvement in ventricular chamber size. MR can be successfully managed in patients after repair of AVSD independent of morphological type. Overall survival is acceptable, and further reoperation within the MVP group is influenced by early outcome of repair. (Circulation. 2000;102[suppl III]:III-160-III-165.)

Key Words: heart defects, congenital ■ mitral valve ■ mechanics ■ valvuloplasty

Hemodynamically significant mitral regurgitation (MR) with need for reoperation occurs in 4% to 15% of patients after repair of atroioventricular septal defects (AVSD) and is the primary cause for reoperation.1–5 The surgical procedures for this MR have been either mitral valve repair (MVP) or mitral valve replacement (MVR). To date, both the reported numbers of patients and reports of such therapy have been few.

MR leads to a volume-overloaded left ventricle. With increasing regurgitation, eccentric hypertrophy occurs, ultimately leading to left ventricular myocardial failure.6 Although the pediatric myocardium appears to tolerate this hemodynamic state better than the adult, if persistent, ventricular dysfunction will occur.7 Although the indications for surgery in adults appear well defined,8 they are less clear in the pediatric population, largely because of the unpredictable potential need for MVR and its attendant problems of size limitations and anticoagulation requirements.9–12

In this report, we have reviewed our experience of the surgical management of patients with hemodynamically significant MR after repair of AVSD. In particular, we report on the clinical outcome of these patients and have sought to examine the impact of surgery on ventricular function, the need for reoperation, and to identify morphological predictors of need for MVR.

Methods
This report is based on a review of patients who underwent mitral valve reoperation from January 1, 1988, to December 31, 1998, in whom the primary repair of the AVSD had been performed at Children’s Hospital, Boston. All patients were considered to have hemodynamically significant MR, based on clinical and echocardiographic features. All underwent surgery with an intention to repair while minimizing regurgitation and avoiding stenosis. MVR was performed at the discretion of the surgeon. Patients undergoing single-ventricle palliation, repair of tetralogy of Fallot–AVSD, and those who underwent primary AVSD repair in another institution were excluded.

Data Acquisition and Analysis
Except for the above exclusions, all patients identified from our institutional database coded with an AVSD and mitral valve surgery...
A total of 46 cases (27 female patients) were identified (Table 2). Of these, 37 had a MVP and the other 9 had an MVR. One third of patients had trisomy 21. The median age (range) at primary operation was 0.6 (0.03 to 10.6) years and at mitral valve reoperation was 2.8 (0.14 to 19.3) years. During the same period, a total of 498 (197 with trisomy 21) underwent primary repair of an AVSD. Of these patients, 36 are reported within our series, representing a 7.2% (95% CI 5.1% to 9.9%) minimum incidence of reoperation for hemodynamically significant mitral regurgitation.

Anatomically, 9 patients (18.6%) had isolated primum atrial septal defects, 9 had a transitional canal defect, and the other 28 (60.8%) had a complete atrioventricular canal defect. The defect was unbalanced in 9, 6 in favor of the right ventricle, and 3 to the left ventricle. All patients were deemed suitable for a biventricular repair. The mitral valve apparatus was potentially stenotic in 10 patients, a parachute deformity being present in 6, and closely spaced papillary muscles in the other 4. One patient in the latter group had a double-orifice mitral valve.

The degree of MR increased from time of primary repair to mitral valve operation. A median increase of 1.5 occurred when initial postoperative regurgitation was less than moderate (grade <3), whereas an increase of 1 grade was noted within patients with moderate or higher grades (grade ≥3).

### Operative Repair/Replacement
At the initial AVSD repair cleft closure had been performed in 29 patients. At the time of the subsequent MV repair surgery, procedures included cleft closure in 32 (89%), annuloplasty or commissuroplasty in 15 (42%), chord shortening in 4 (11%), leaflet surgery in 4 (11%), and resection of portion of prolapsed anterior leaflet (2 patients). In 15, closure of an isolated cleft was the only procedure performed. In the MVR group, 1 patient received a supra-annular prosthesis (16-mm Carbomedics valve) and the other 8 received prostheses placed at the annular level, ranging in size from 16 to 31 mm (16, 18×2, 19, 21, 25, 31×2). Transthoracic echocardiography (TEE) was performed at the time of MR repair in 30 of 46 patients, being most frequently utilized in recent years (P=0.029).

### Results

#### Mortality
There was 1 early death, in the MVP group (2.7%), for an overall early mortality rate of 2.2% (Table 3). This patient died within 30 days of repair in the setting of a pulmonary hypertensive crisis. Late deaths occurred in 4 patients, 3 in...
the repair group. One patient died of bacterial endocarditis in the setting of varicella and bacterial superinfection and had developed recurrence of MR, necessitating insertion of a prosthetic valve. Another patient died of progressive pulmonary vein stenosis, the latter identified at the time of primary repair, and another, an MVP patient with trisomy 21, died of myelodysplasia. Both these latter patients were deemed to have a satisfactory MVP at their last evaluation. In the MVR group, 1 late death occurred as the result of acute valve dysfunction with prominent clot and pannus confirmed at autopsy.

Survival
The overall survival was 89.9%, 86.6%, and 86.6% at 1, 5, and 10 years, respectively (Figure 1). Survival rates in both groups were similar (MVP versus MVR, \( P = 0.97 \)), being 97.2% versus 85.7%, 92.6% versus 85.7%, and 86.6% versus 85.7% at 1, 5, and 10 years, respectively.

Freedom From Reoperation
Five patients within the series underwent a second mitral valve reoperation, 4 within the MVP group and 1 within the MVR group. The time to reoperation was not significantly different between groups (Figure 2). Freedom from reoperation was 96.9% versus 85.7% at 1 year, 78.5% versus 85.7% at 5 years, and 78.5% versus 85.7% at 9 years for the MVP and MVR groups, respectively.

Of the 4 patients requiring reoperation in the repair group, 2 received a repeat MVP for significant MR, whereas another patient was deemed unsuitable for repeat repair, and a mitral valve prosthesis was inserted. In the fourth patient, significant MR (not stenosis), a significant association was present (\( P = 0.25 \)). A trend toward significance leading to earlier reoperation included nonutilization of an annuloplasty technique (\( P = 0.06 \)). Pertinent factors that were not significant included MV morphology, location of regurgitant jet, suture of the cleft, and use of TEE.

Change in Clinical Status
A significant improvement in clinical status was seen after both MVP and MVR (Table 3), as assessed by New York Heart Association scale, degree of MR assessed by echocardiography, and growth as measured by weight and height percentile. The use of afterload reduction agents decreased from 69% to 41%. The degree of MR was also evaluated for the MVP group alone to avoid confounding from the MVR group, in whom the incidence of postoperative MR was low. In the MVP group, MR decreased from 3.6 ± 0.5 to 2.1 ± 0.6 (\( P = 0.004 \)) early after surgery and was unchanged at the most recent evaluation (2.3 ± 0.7). Other complications such as

<table>
<thead>
<tr>
<th>TABLE 3. Clinical Status in Response to Mitral Valve Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre</td>
</tr>
<tr>
<td>-----------------</td>
</tr>
<tr>
<td>NYHA class (I–IV)*</td>
</tr>
<tr>
<td>MR degree (0–4)*</td>
</tr>
<tr>
<td>Weight, %</td>
</tr>
<tr>
<td>Height, %</td>
</tr>
</tbody>
</table>

Values are mean ± SD. NYHA indicates New York Heart Association. * \( P < 0.05 \).
cerebrovascular accidents and infections occurred infrequently (Table 4).

Mechanics After Valve Surgery
The impact of significant MR on ventricular mechanics and the beneficial effect of volume reduction is shown after both MVP and MVR in Table 5. Overall, the end-diastolic volume Z score decreased by 46.8% early after surgery ($P=0.0001$) and by 57.5% at last evaluation. The end-systolic volume Z score rose in the early postoperative period by 20.1% ($P=NS$), more noticeably in the MVR group, reflecting acute volume unloading and increase in afterload. The overall initial rise in end-systolic volume, more pronounced in the MVR group, resulted in a fall in ejection fraction in the early postoperative period by 26.6% ($P=0.0001$). The later rise in ejection fraction, again more obvious in the MVR group, from 47.6% to 58.2%, trended toward significance ($P=0.08$).

The mitral valve annulus Z score showed a significant fall in the early postoperative period in the MVP group (lateral: $1.46 \pm 1.97$ to $-0.95 \pm 3.28$, $P=0.001$, and AP: $2.65 \pm 2.21$ to $0.60 \pm 2.30$, $P=0.001$). However, an increase in annulus dimension was detected in both planes on long-term follow-up in the MVP group (lateral: $-0.95 \pm 3.28$ to $0.63 \pm 1.37$, $P=0.004$, and AP: $0.60 \pm 2.30$ to $1.29 \pm 1.85$, $P=0.36$).

Morphological Predictors of Risk of MVR at Time of MV Surgery
Preoperative and perioperative morphological factors were analyzed to determine their impact on the choice of surgery, namely repair versus replacement. Of these, TEE was the only significant factor detected by univariate analysis ($P=0.005$). Of the 37 patients who had MVP, 28 had TEE in the operating room, whereas only 2 of 9 patients in the MVR group underwent such a study. This suggests a possible role for TEE in directing feasibility of valve repair. Use of this technology increased during the study period, from 11 in the first 23 patients, to 19 in the last 23 patients ($P=0.029$). Although before 1988, MVP for MR was rarely used, since then no significant change has occurred in the rate of utilization of prosthetic valves. The significance of this finding is unclear and may reflect a prior decision to replace the valve rather than a contribution from TEE in the decision process. MV morphology1,3,5,14 and nonclosure of the cleft,3,15 previously reported as risk factors for postoperative MR after primary repair, were not predictors of MVP failure in the current study. In addition, the site of MR jet, left ventricular chamber size, and MV annular dimension were not predictors of MVR necessity.

Discussion
MR remains the principal indication for repeat operation in patients after repair of AVSD, with a reported incidence of 4% to 15%.1–5 Surgical options for management of this problem include MVP and MVR. Although the occurrence of MR and need for reoperation are identified in several reports, little attention has focused on the durability of MVP or the factors associated with successful MVP versus MVR. In this current report, the largest experience to date and spanning an 11-year period from a single institution, low mortality rates were found with the use both therapeutic options. In addition, low incidence rates of MV reoperation were also found in both groups together with reduced durability of repair being identified in those patients with moderate to severe MR after MVP. Double-orifice mitral valve,1,3,5 parachute mitral valve,14 non–Down syndrome patients,15 nonclosure of the cleft at primary repair,3,15 and preoperative4,5 and early postoperative MR1 have been shown to be associated with progressive, late postoperative MR. These factors in the current study were not found to be of significance in determining the durability of repair or need for valve replacement. We did observe, however, the previously reported high incidence of heart block within the MVR group,12 necessitating early pacemaker insertion.

Mortality Rates
Mortality rates of 26% to 50% for valve replacement in the setting of AVSD in the early postoperative period and 10% when reoperation occurs >6 months after primary repair have been reported, considered in part to reflect the high rate of complete heart block necessitating pacemaker insertion.12 We report low mortality rates of 10.1%, 13.4%, and 13.4% at 1, 5, and 10 years, respectively, and an early postoperative value of 2.2%, with similar rates between the MVP and MVR groups. Within the repair group, freedom from cardiac-

**TABLE 4. Complications**

<table>
<thead>
<tr>
<th>Complication</th>
<th>MVP (n=37)</th>
<th>MVR (n=9)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Early mortality</td>
<td>1 (2.7)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Total mortality</td>
<td>4 (10.8)</td>
<td>1 (11.1)</td>
</tr>
<tr>
<td>CHB</td>
<td>1 (2.7)*</td>
<td>3 (37.5)†</td>
</tr>
<tr>
<td>CVA</td>
<td>0 (0)</td>
<td>1 (11.1)</td>
</tr>
<tr>
<td>Minor</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pneumonia</td>
<td>1 (2.7)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Wound infection</td>
<td>1 (2.7)</td>
<td>1 (11.1)</td>
</tr>
</tbody>
</table>

Values are n (%). CHB indicates complete heart block; CVA, cerebrovascular accident.

*Heart block after reoperation and prosthetic MV insertion.
†Three of 8 patients had heart block. One patient had complete heart block and pacemaker from primary repair.

**TABLE 5. Echocardiographic Ventricular Mechanics Before and After Mitral Valve Surgery**

<table>
<thead>
<tr>
<th></th>
<th>Preoperative</th>
<th>Early Follow-Up</th>
<th>Late Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>MVP</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>LVEDV (Z score)</td>
<td>$3.7 \pm 3.6$</td>
<td>$1.5 \pm 1.9^*$</td>
<td>$1.7 \pm 2.3$</td>
</tr>
<tr>
<td>LVESV (Z score)</td>
<td>$2.3 \pm 2.3$</td>
<td>$2.3 \pm 2.2$</td>
<td>$1.4 \pm 1.8$</td>
</tr>
<tr>
<td>Ejection fraction, %</td>
<td>$64.6 \pm 8.5$</td>
<td>$50.1 \pm 13.7^*$</td>
<td>$59.7 \pm 10.1$</td>
</tr>
<tr>
<td><strong>MVR</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>LVEDV (Z score)</td>
<td>$3.3 \pm 1.6$</td>
<td>$3.3 \pm 3.1$</td>
<td>$1.1 \pm 3.3$</td>
</tr>
<tr>
<td>LVESV (Z score)</td>
<td>$2.2 \pm 1.3$</td>
<td>$4.4 \pm 3.4$</td>
<td>$0.1 \pm 2.9$</td>
</tr>
<tr>
<td>Ejection fraction, %</td>
<td>$66.3 \pm 6.3$</td>
<td>$38.7 \pm 12.7^*$</td>
<td>$52.5 \pm 8.2$</td>
</tr>
</tbody>
</table>

*$P<0.002$. 

Downloaded from http://circ.ahajournals.org/ by guest on November 25, 2017
related valve mortality was 97.2%, 92.6%, and 92.6% at 1, 5, and 10 years, respectively.

Although survival has improved with time for MVR, the incidence of early complete heart block in our group remains high, at 37.5%, suggesting that something other than AV node dysfunction previously contributed to the earlier higher mortality rates. Although an earlier age of reintervention may be associated with less AV valve distortion and thus perhaps enabling higher chance of repair, age of MV repair was not significantly different from prior studies from our own institution.22 However, 2 important differences have occurred since that era: (a) the earlier age at primary AVSD repair,1,16 with its associated lower mortality rates, avoidance of AV valve distortion, and lower incidence of pulmonary vascular obstructive disease are likely to play a role,17 as are (b) the improvements in myocardial preservation.18

Prior studies of pediatric MVP have concentrated on congenital MV regurgitation. Aharon et al19 reported on an early experience of 79 patients and reported survival rates of 94% at 1 and 82% at 5 years. Ohnio et al20 reported on 41 patients with congenital mitral regurgitation, without any early or late deaths. A similar experience was described by Matsumoto et al.21 Our study reports favorable survival to 10 years after MVP for MR in patients who previously had repair of AVSD defects being 97.2%, 92.6%, and 92.6% at 1, 5, and 10 years, respectively.

Complete Heart Block

The reported incidence of complete heart block after mitral valve replacement varies between 20% and 30%.12 and may reflect mechanical pressure from the prosthesis on the conduction system.22 We report a similar experience in our series, namely 37.5% in the early postoperative period. Within the repair group, however, no AV node dysfunction occurred, representing a clear advantage to reparative techniques by avoiding the need for generator changes and the influences of dyssynchronous ventricular contraction during pacing. The only patient within the repair group who required a pacemaker for complete heart block did so after insertion of an MV prosthesis for recurrent MR.

Repair Prolongs the Time to Ultimate Valve Replacement

MVR operations are associated with significant mortality and morbidity. The risk of thromboembolism, need for long-term anticoagulation, hemodynamic dysfunction, complete heart block, bacterial endocarditis, and absence of growth potential cannot be discounted. We noted several of these side effects, even within our own small series. Although a need for reoperation for small valve size was not seen within the time span of this study, the sizes used will undoubtedly require future reoperations. MVP, in contrast, avoids the need for anticoagulation and provides annular growth potential. The aim of repair is to optimize the functioning potential of the valve and to at least prolong the interval before replacement. The true long-term durability of these repairs is unknown. Prior studies have reported various reparative techniques, confirming the possibility of successful repair in patients with AVSD and in those with congenital MR.19–21,23

In our series, freedom from reoperation at 9 years was 78.5% in the MVP group and 85.7% in the MVR group. Furthermore, we report the ability to re-repair the valve in 2 patients. We identified 1 risk factor, which appeared to shorten the durability of repair, namely significant residual MR immediately after that operation. It is not surprising that significant MR on echocardiography before discharge predicts cardiac reoperation, and use of this transthoracic tool is of clinical utility in follow-up, in contrast to TEE, which has been shown to correlate poorly with degree of MR seen on long-term follow-up.24 Our report thus suggests a greater role for transthoracic echocardiography early after surgery, whereas TEE is more appropriate at the time of actual surgical repair. In our study, the use of annuloplasty indicated a trend toward longer durability of repair.

In non–Down syndrome patients with AVSD,15 additional defects such as double-orifice MV1,3,5 and parachute MV14 have been reported to be associated with early postoperative regurgitation and need for repeat operation. Although these additional defects occurred in some of our patients, they did not appear to adversely affect the repair outcome.

The use of intraoperative TEE has been reported to assist surgeons in more satisfactory valve repairs,24 a finding also noted in our study with a higher utilization rate in the latter half of our cohort as well as a higher repair rate when used.

Clinical Status

Early surgical intervention to correct MR before the onset of significant symptoms of heart failure has been recommended in adults to avoid irreversible left ventricular dysfunction, and specific echocardiographic indexes have been used as indicators for reintervention.8 Similar echocardiographic guidelines are not currently available for pediatric patients, such that symptoms often constitute the surgical indication, this latter approach being reported by the observations of Krishnan et al.7 MR is associated with a volume-overload state with low wall stress in early and late systole. The repair of the regurgitant valve, however, leads to a state of afterload mismatch with elevated afterload and fall in ejection fraction.6 We report a similar fall in ejection fraction within the early postoperative period, particularly in the MVR group. This outcome reflects a decrease in end-diastolic dimension without a concomitant reduction in end-systolic dimension, similar to that seen in adults undergoing mitral surgery.25 However, on follow-up, the end-systolic dimension decreased, with resultant normalization in ejection fraction.

The results of this study are encouraging. Echocardiographic and clinical measures of cardiac status show improvement, with an acceptable duration of freedom from reoperation. A decrease in MR degree during follow-up is supported by the ventricular volumes measured, and their use presents a more objective means of assessing regurgitation and supports the interpretation in color jet width seen in 2 orthogonal planes.

Conclusions

Repair of AVSD in infants is associated with a low mortality rate. MR remains a significant reason for reoperation. Although options include MVR and MVP, we find that repair is
possible in all anatomic subtypes including parachute mitral valves, with significant improvement in patient status. Similar results are achieved with replacement, although a high incidence of complete heart block is noted. Replacement also carries with it the need for future prosthesis replacement in the setting of growth.

Acknowledgments

We gratefully acknowledge the many cardiologists and cardiac surgeons involved in the care of these patients. Furthermore, we thank Dr. Kim Gauvreau for her assistance in the statistical analysis.

References

Surgical Management of Mitral Regurgitation After Repair of Endocardial Cushion Defects: Early and Midterm Results
Adrian M. Moran, Sabine Daebritz, John F. Keane and John E. Mayer

Circulation. 2000;102:iili-160-Iii-165
doi: 10.1161/01.CIR.102.suppl_3.III-160
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2000 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/102/suppl_3/iili-160

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/