Double-Transseptal, Double-Balloon Valvuloplasty for Congenital Mitral Stenosis

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Background. Eight patients with severe congenital mitral stenosis underwent double transseptal, double-balloon valvuloplasty; two had isolated congenital mitral stenosis, six had additional cardiac defects, and one had previous surgical valvotomy. Ages ranged from 0.6 to 36 years (median, 9 years).

Methods and Results. All procedures were tolerated well. After valvuloplasty, the left atrial wave minus the left ventricular end-diastolic pressure (LVEDP) gradient was reduced from 25±6 mm Hg to 9±3 mm Hg (p<0.001), the mitral valve mean gradient was reduced from 18±7 mm Hg to 8±3 mm Hg (p=0.003), and the LVEDP was unchanged. All patients had marked clinical improvement. Only one patient developed significant mitral regurgitation. Two of the first four patients underwent repeat balloon valvuloplasty 7 months later. Follow-up evaluation on six patients from 4 to 54 months revealed no recurrence of symptoms or increased mitral regurgitation.

Conclusions. Double transseptal, double-balloon valvuloplasty is an effective treatment for many forms of congenital mitral stenosis. Mitral regurgitation is uncommon after this procedure. The double transseptal approach results in less trauma to the atrial septum and femoral veins and allows easy assessment of any residual postvalvuloplasty gradient. (Circulation 1992;85:123–129)

Congenital mitral stenosis (CMS) is a general term describing a broad spectrum of developmental abnormalities resulting in an obstruction of flow from the left atrium to the left ventricle. The obstruction may occur at the mitral annulus, leaflets, chordae tendineae, or papillary muscles, separately or in combination. Four basic types of CMS are recognized: typical CMS (thickened leaflets, shortened chordae tendineae, decreased interchordal spaces), supravalve mitral membrane, parachute mitral valve, and hypoplastic left heart syndrome.1,2 CMS occurs in approximately 0.5% of patients with congenital heart disease and may have profound hemodynamic effects.3,4 The diagnosis of CMS often is obscured due to associated cardiac defects, commonly left-sided obstructive lesions.5,6 Medical management may provide temporary symptomatic improvement but often is unsatisfactory.7

Surgery for CMS is directed toward relieving the levels of mitral obstruction while providing a competent ativoventricular valve. Mitral valve reconstruction in infants and children is associated with significant morbidity and mortality. Mitral valve replacement in this age group prevents further growth of the mitral annulus and necessitates anticoagulation.

In efforts to provide symptomatic relief and allow continued growth of left heart structures, we performed percutaneous balloon valvuloplasty using two separate transseptal punctures and two balloons on eight patients with CMS.

Methods

Patients

From January 1986 to March 1990, eight patients with severe CMS documented by history, physical examination, and echocardiography underwent double transseptal, double-balloon valvuloplasty (DT-BVV). Patient demographics are listed in Table 1. Ages ranged from 7 months to 36 years (median, 9 years), and weights ranged from 5.4 to 84 kg (median, 20 kg). Five patients were female, and three patients were male. Informed consent was obtained before each procedure.

Valvuloplasty Technique

All patients received intramuscular premedication and local anesthesia. Using a percutaneous tech-
nique, sheaths (Argon Inc., Athens, Tex.) were placed in both femoral veins and a catheter was placed in one femoral artery. After a standard right heart catheterization, a transseptal procedure was performed as previously described, using a sideport transseptal sheath (Cook Inc., Bloomington, Ind., or USCI Inc., Billerica, Mass.).

Left atrial pressure was measured through the sheath. An end-hole catheter (USCI Inc.) one French size smaller than the sheath was placed through the sheath into the left ventricle; simultaneous left atrial and ventricular pressures were recorded. A left atrial angiogram was performed in the caudally angulated right anterior oblique and four-chamber projections using an NIH cardiomarker catheter (USCI Inc.), and the diameter of the mitral annulus was estimated. Using the same projections, a left ventricular angiogram was then performed.

To afford stability during catheter exchanges and to protect the left ventricular apex from the rigid balloon catheter tip, a 0.035- or 0.038-in. Teflon®-coated superstiff exchange wire (Medi-Tech Inc., Watertown, Mass.) was modified in the following manner: the soft tip of the wire was manually coiled, and a U-shaped curve was then placed at the junction of the stiff core with the soft tip. This allowed positioning of a smooth curve of stiff exchange wire in the left ventricular apex, with the soft wire tip extending into the outflow tract.

Two valvuloplasty balloon catheters (Medi-Tech Inc., or Mansfield Scientific, Mansfield, Mass.) were chosen so that the sum of the balloon diameters was approximately equal to the estimated diameter of the mitral annulus. Relatively long balloons (3 cm for small patients, 5–6 cm for larger patients) were chosen to minimize balloon movement during inflation. The balloons were prepared using iohexol (Winthrop Pharmaceuticals, New York) diluted 1:5 with saline and were then connected to a pressure monitoring inflation device (Sci-Med Life Systems Inc., Minneapolis, Minn.).

An end-hole catheter was placed through the transseptal sheath into the left ventricle, and the preformed exchange wire was positioned in the left ventricular apex. Using the other femoral vein, a second transseptal puncture was performed in the fossa ovalis approximately 1 cm in distance (superior or inferior) from the initial puncture followed by placement of another modified Teflon®-coated exchange wire. After the transseptal procedures, five of 10 patients received intravenous heparin (100 units/kg). The end-hole catheters and transseptal sheaths were removed. Sequentially, the balloon catheters were advanced over the exchange wires, through the separate punctures in the atrial septum, and centered across the mitral valve annulus; the atrial septal puncture did not require dilation before placing the valvuloplasty balloons. The balloon catheters were inflated rapidly to the maximum recommended inflation pressure (3–6 atm) for a 3–15-second duration as depicted in Figure 1. If the mitral “waist” did not appear with the balloons fully inflated, larger balloons were used until the mitral waist appeared and resolved with balloon inflation. The balloon catheters were replaced with transseptal sheaths over end-hole catheters, and the exchange wires were removed. Simultaneous and pullback left atrial and ventricular pressures were recorded, and angiography was repeated.

The mitral valve gradient was assessed in the catheterization laboratory by the left atrial a wave minus the left ventricular end-diastolic pressure (LVEDP). After catheterization, the mean gradient in six patients was calculated by computer-assisted planimetry of the pressure tracings. During all future procedures, cardiac output will be measured before and after valvuloplasty.

During catheterization, all catheters and sheaths were flushed continuously to minimize clot formation. The flush solution contained normal saline and heparin (3 units/ml). Because the patients received a quantity of heparin from the flush solution, the original protocol did not include a routine loading dose of heparin. The protocol was amended recently; all patients now receive a heparin loading dose (100 units/kg) after transseptal access to the left atrium. Serial activated clotting times were measured with a desired range of 350–400 seconds; a second heparin loading dose (10–30 units/kg) was occasionally required.

A noninvasive estimate of the mitral valve area was obtained in six patients. Because obstruction may occur below the plane of the mitral valve leaflet tips, echocardiographic two-dimensional valve area was not used; therefore, the pressure half-time method was used. Echocardiograms were obtained within 1 week before the procedure and were repeated the day after the procedure and at follow-up outpatient clinic examinations.

**Follow-up**

Three patients were reevaluated at regular intervals in our outpatient clinic. Two older patients were followed by local medical cardiologists. One patient moved and was followed at another institution, and two patients died.
Statistical Analysis

Data obtained before and after double-balloon valvuloplasty were compared using a two-tailed Student's t test. Statistical significance was assumed if the null hypothesis could be rejected at the 0.05 probability level.

Results

All procedures were tolerated well. Although all patients had dyspnea and/or orthopnea before the procedure, general anesthesia was not used and none of the patients required intubation. Two separate transseptal procedures were performed in each patient.

TABLE 2. Hemodynamic Data

<table>
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<tr>
<th>Patient</th>
<th>LA a (mm Hg)</th>
<th>LVEDP (mm Hg)</th>
<th>LA a–LVEDP (mm Hg)</th>
<th>Gradient (mm Hg)</th>
<th>Echo valve area (cm²)</th>
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<tr>
<td>Mean±SD</td>
<td>36±5 20±7 11±3 11±5 25±6 9±3 18±7 8±3 0.8±0.2 1.4±0.3 1.2±0.2</td>
<td></td>
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<tr>
<td>p</td>
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LA a, left atrial a wave; LVEDP, left ventricular end-diastolic pressure; Gradient, mean gradient.
without difficulty. The fluoroscopy time ranged from 48 to 86 minutes (mean, 63 minutes). The two-balloon diameter: annulus diameter ratio was 0.87–1.26 (mean, 1.13). No complications were noted from the placement of bilateral femoral venous transseptal sheaths.

**Immediate Results**

All eight patients had marked clinical improvement immediately after DT-DBV. As seen in Table 2, the left atrial a wave minus the LVEDP was reduced from 25±6 mm Hg to 9±3 mm Hg (p<0.001). The mitral valve mean gradient was reduced from 18±7 mm Hg to 8±3 mm Hg (p=0.003). LVEDP was unchanged. Figures 2 and 3 display the mitral valve gradient before and after valvuloplasty; a marked decrease in the transvalvular gradient was obtained in all procedures. The mean mitral valve area (pressure halftime) increased 75% from 0.8±0.2 to 1.4±0.3 cm² (p<0.001).

Angiographic improvement was noted in six patients (Figure 4), as evidenced by increased diastolic excursion of the mitral leaflets, loss of late diastolic leaflet doming during atrial systole, and a broad flow of contrast across the valve; similar findings were noted on the echocardiogram. There was no correlation between the decrease in mitral valve gradient and the balloon:annulus ratio, the number of inflations, or the duration of inflation.

Patient 4 underwent thermodilution cardiac output measurement before and after valvuloplasty during the initial and repeat procedures. Both valvuloplasty procedures resulted in a small increase in cardiac output: 1.0–1.2 l/min initial valvuloplasty, and 0.9–1.0 l/min repeat valvuloplasty.

Only patient 6 developed mitral regurgitation (2+/4+). The regurgitation was likely due to the complex nature of her stenosis (supravalve membrane, parachute mitral valve). Clinically she has improved, the left atrial size has decreased, and the mitral valve area has remained stable.

**Long-term Results**

Six patients have continued clinical improvement, with follow-up ranging from 4 to 54 months (mean, 24 months); two patients died. Serial mitral valve area measurements by echocardiography were obtained in five patients. The mitral valve area decreased slightly, from 1.4±0.3 cm² the day after balloon valvuloplasty to 1.2±0.2 cm² on follow-up exam; this remained a 50% increase (p=0.004) from the pre-valvuloplasty mitral valve area. Follow-up invasive studies on three patients (two repeat valvuloplasty and one postmortem) revealed no defect in the atrial septum.

Patients 3, 5, 6, and 7 continued to have improved exercise tolerance and growth. Patient 5, who had been unable to walk to school, returned to school and participated in sports. Patient 3, who had the least dramatic immediate hemodynamic result, had a repeat catheterization 24 months later; the mitral valve gradient further decreased, and his exercise tolerance continued to increase. Patient 7 had DT-DBV 8 years after a Blalock-Taussig shunt; although he was mildly cyanotic, he maintained an improved level of activity.

Two patients had an immediate reduction in the mitral valve gradient, but they developed restenosis. Repeat DT-DBV was performed 7 months after the initial procedure. In both patients, the mitral valve gradient again was reduced significantly. Patient 1, the first patient to undergo this procedure, underwent repeat DT-DBV with the use of larger-diameter balloon catheters and has had no restenosis in over 4 years. Patient 4, who weighed only 5.4 kg and had ring X chromosome and congenital nephrotic syndrome, underwent successful repeat DT-DBV but again developed restenosis over the next 12 months. She was referred for surgery, where the mitral annulus was noted to be small: both leaflets were thickened, the chordae tendineae were fibrotic and shortened, and the papillary muscles were separate but nearly fused at the base; this was a forme fruste parachute mitral valve. Extensive mitral valve reconstruction was performed. An echocardiogram performed the day after surgery revealed mild mitral regurgitation. The patient died on the second postoperative day from low cardiac output.
Patient 2 was doing well clinically, and the mitral valve area remained stable for 13 months after valvuloplasty. She died suddenly from a fulminant multisystem viral infection documented by postmortem examination. At autopsy, the left atrium was enlarged and sclerotic, the mitral leaflets were thickened, and there were no interchordal spaces; the leaflets were fused with the papillary muscles. The only evidence of previous balloon valvuloplasty was the rather sharp demarcation separating the two dysplastic leaflets.

Complications

Two patients experienced complications. Patient 7 had a transient ischemic attack; he was polycythemic (hemoglobin, 19 g/dl) and had not received a loading dose of heparin. Patient 2, who received heparin, was noted to have an enlarged cardiac silhouette by fluoroscopy after completing the procedure. An echocardiogram performed in the catheterization laboratory revealed a pericardial effusion; a hemopericardium was evacuated uneventfully and did not recur.

Discussion

Previous Studies

Various surgical techniques have been described for CMS, depending on the type of mitral obstruction. Mitral valve repair is difficult because of the dysplastic nature of one or more of the valve components. Two recent reviews reported 17–19% operative mortality for mitral valve repair.11,12 Mitral valve...
replacement in children is complicated by the inability to place an adequate size prosthetic valve in the annulus and has problems associated with anticoagulation. Mitral valve replacement in children has an operative mortality of 24–48%.1,11–13

An alternative approach, using a left atrial to left ventricular conduit with a prosthetic valve, has been reported.14–16 This procedure is not without risk and requires long-term anticoagulation and subsequent replacement as the child grows.

**Balloon Valvuloplasty**

For several years, balloon valvuloplasty has been performed on adults with rheumatic mitral stenosis. During balloon inflation, separation of the thickened, fused commissures and leaflets occurs.17 Because CMS is caused by a broad spectrum of anatomic variants, each variant may respond differently to balloon valvuloplasty.

Patients with typical CMS are the best candidates for this procedure. Analogous to rheumatic mitral stenosis, balloon dilation in CMS separates the thickened, fused commissures, providing a larger effective mitral orifice. Four of our patients had typical CMS; all four had an excellent result after DT-DBV.

A parachute mitral valve has limited leaflet mobility because of shortened, thickened chordae and fused papillary muscles. Balloon valvuloplasty may dilate the subannular structures, increasing leaflet mobility. Because the only other alternative was mitral valve replacement, we attempted DT-DBV on three patients; patients 3 and 6 have done very well (patient 4 was discussed previously).

Isolated supravalve mitral membrane is a surgical disease; no benefit would be derived from balloon valvuloplasty. However, if the membrane is not restrictive and is part of a more complex lesion (i.e., Shone’s syndrome), DT-DBV may relieve the other levels of valvular stenosis.

Two separate case reports of balloon valvuloplasty for CMS were published by Kveselis et al18 and Alday et al.19 Recently, Spevak et al20 reported encouraging results on nine patients with CMS who underwent balloon valvuloplasty. All procedures for CMS reported to date involved single-balloon valvuloplasty, and in at least one instance, dilation results were limited by the size of the single-balloon catheter.18

In this series, we performed two separate transseptal punctures and placed one modified exchange wire through each puncture. The balloon catheters were advanced easily through the atrial septum without requiring prior septal dilation because of 1) an adequate septal puncture from the transseptal sheath and dilator, 2) the use of two smaller low-profile balloons, and 3) the use of superstiff exchange wires. The double transseptal, double-balloon technique not only allows a larger effective balloon diameter during dilation but also minimizes left-to-right atrial shunting after valvuloplasty and decreases trauma to the femoral veins.

Alternatively, two exchange wires and balloon catheters could be placed through a single transseptal puncture. This technique is used in adults with rheumatic mitral stenosis.17 Using only a single atrial septal puncture, exchanging catheters for larger-size balloons would be more difficult. Also, a residual atrial septal defect with left-to-right shunt occurs in a number of cases.21

Inoue et al22 designed a special balloon catheter for mitral valvuloplasty in adults. The balloon has a high-profile contour and requires dilation of the atrial septum before insertion. The resultant trauma to the femoral veins and atrial septum makes the DT-DBV approach preferable in children.

**Conclusions**

Congenital mitral stenosis can have profound hemodynamic effects. Surgical options are limited because of the dysplastic nature of the mitral valve components. Double transseptal, double-balloon valvuloplasty was not intended to be a panacea, and these patients may require mitral valve replacement in the future. However, because six of the eight patients were 11 years of age or younger, DT-DBV was undertaken with hopes of 1) reducing symptoms and improving somatic growth, 2) delaying or avoiding mitral valve replacement (allowing further growth of the mitral annulus), and 3) avoiding anticoagulation and its complications.

We were able to significantly reduce the mitral valve gradient and improve their clinical condition with few complications. These results have been encouraging, although the patients still have some degree of mitral obstruction and will require continued follow-up. Double transseptal, double-balloon valvuloplasty can be performed at an age earlier than one would recommend surgical intervention, before the development of arrhythmias, pulmonary vascular obstructive disease, and ventricular dysfunction.

**Acknowledgment**

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