Published in final edited form as:

Am J Cardiol. 2009 June 15; 103(12): 1770–1773. doi:10.1016/j.amjcard.2009.02.062.

Effectiveness of Balloon Valvuloplasty for Palliation of Mitral Stenosis After Repair of Atrioventricular Canal Defects

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Closure of a mitral valve (MV) cleft, small left-sided cardiac structures, and ventricular imbalance all may contribute to mitral stenosis (MS) after repair of atrioventricular canal (AVC) defects. MV replacement is the traditional therapy but carries high risk in young children. The utility of balloon mitral valvuloplasty (BMV) in postoperative MS is not established and may offer alternative therapy or palliation. Since 1996, 10 patients with repaired AVC defects have undergone BMV at a median age of 2.5 years (range 8 months to 14 years), a median of 2 years after AVC repair. At catheterization, the median value of mean MS gradients was 16 mm Hg (range 12 to 22) and was reduced by 34% after BMV. Before BMV, there was mild mitral regurgitation in 9 of 10 patients, which increased to severe in 1 patient. All patients were alive at follow-up (median 5.4 years). Repeat BMV was performed in 4 patients, 10 weeks to 18 months after initial BMV. One patient underwent surgical valvuloplasty; 3 underwent MV replacement 2, 3, and 28 months after BMV. In the 6 patients (60%) with a native MV at most recent follow-up (median 3.2 years), the mean Doppler MS gradient was 9 mm Hg, the median weight had doubled, and weight percentile had increased significantly. In conclusion, BMV provides relief of MS in most patients with repaired AVC defects; marked increases in mitral regurgitation are uncommon. Because BMV can incompletely relieve obstruction and increase mitral regurgitation, it will not be definitive in most patients but will usually delay MV replacement to accommodate a larger prosthesis.

With a reported incidence of approximately 2% to 6%, clinically important mitral valve (MV) stenosis is a rare but significant complication after the repair of complete and partial atrioventricular canal (AVC) defects.^{1, 2, 3, 4 and 5} Anatomic and technical factors may contribute to this outcome,^{5, 6, 7, 8, 9, 10, 11, 12 and 13} and clinical management is often challenging. The traditional therapy in such cases is MV replacement (MVR).⁴ Although results appear to be improving, MVR in children carries a high risk for morbidity and mortality, particularly in infants and young children, and subsequent redo MVR is nearly inevitable.^{14 and 15} In patients with congenital and rheumatic mitral stenosis (MS), balloon

mitral valvuloplasty (BMV) offers an alternative treatment that can serve as long-term therapy or at least delay MVR until the patient has grown and can accommodate a larger MV prosthesis.^{16 and 17} However, the utility of BMV for postoperative MS in children after the repair of partial or complete AVC defects has not been reported. Since 1996, 10 children have undergone BMV for severe MS after the repair of complete AVC defects or closure of the cleft MV associated with partial AVC defects at our institution. In this report, we describe our experience with this cohort.

Methods and Results

From 1996 to 2007, 10 patients with previously repaired complete (n = 6), partial (n = 3), or transitional (n = 1) AVC defects underwent BMV for MS at a median age of 2.5 years (range 8 months to 14 years). The entry criteria for BMV were not defined precisely but were similar to those we have reported for congenital MS.¹⁸ The median duration between the original AVC repair and BMV was 2 years (range 4.4 months to 14 years). The median weight at BMV was 10 kg (range 5 to 62), and the median weight percentile for age was 6th (range <3rd to >97th). In all patients, the MV cleft was closed either at the time of initial AVC repair (n = 9) or subsequently (n = 1). Two patients underwent repair of aortic coarctation 3 months before repair of partial AVC, 1 of whom also underwent 2 separate procedures for relief of subaortic stenosis, 3 and 5 years after initial AVC repair, the second of which was in conjunction with surgical mitral valvuloplasty. Isolated surgical mitral valvuloplasty had been performed in 1 patient.

All patients had morphologic abnormalities of the MV leaflets and/or tension apparatus, including short, thick chordae tendineae and obliteration of interchordal spaces in all patients, double-orifice MV in 2, parachute-like MV in 2, and a supralvalvar mitral ring in 1. The median MV annular diameter z scores were 0.1 (range -2.9 to 9.2) in the anterior-posterior dimension and -0.60 (range -2.4 to 4.4) in the lateral dimension.

Individual baseline echocardiographic and hemodynamic data at catheterization are listed in Table 1. Before intervention, the median peak and mean Doppler estimated MV inflow gradients were 25 mm Hg (range 17 to 30) and 14 mm Hg (range 8 to 16), respectively. At catheterization, the median peak and mean left ventricular inflow gradients were 28 mm Hg (range 23 to 30) and 16 mm Hg (range 12 to 22), respectively, and were consistently higher than the Doppler estimates (Figure 1). Mean left atrial pressure ranged from 18 to 36 mm Hg (median 24), and mean pulmonary artery pressure ranged from 30 to 75 mm Hg (median 34). Mild MR was present in 9 of 10 patients (90%).

The technical details of BMV have been described previously^{16 and 19} and were not modified for this particular patient population. The size of the initial dilating balloon was determined by patient size rather than by the diameter of the MV annulus and was also varied by modulation of the inflation pressure.¹⁸ A balloon was successfully passed through the MV and inflated in all patients. A median of 3 (range 2 to 4) different balloon sizes were used, and 4 (range 2 to 7) balloon inflations were performed per patient.

After BMV, the median peak and mean left ventricular inflow gradients were 14 mm Hg (range 12 to 26) and 11 mm Hg (range 6 to 22) (Figure 1), significantly lower than before BMV ($p < 0.007$ and $p = 0.002$). Post-BMV peak and mean left ventricular inflow gradients were reduced by 49% and 34%, respectively. There were minimal gradient reductions in 2 patients; a previously closed cleft was clearly open by 3-dimensional echocardiography in 1 of these patients (Figure 2). In 3 patients, preexisting mild MR increased to moderate ($n = 2$) or severe ($n = 1$).

During follow-up ranging from 8 months to 11 years (median 5.4 years), all patients were alive. Repeat BMV was performed for recurrent or residual MS in 4 patients (3, 3, 9, and 18 months later), 1 of whom required a third dilation 4 years later and 2 years thereafter developed severe biventricular dysfunction, for which cardiac transplantation was performed. In the 4 patients who underwent repeat BMV, median peak and mean left ventricular inflow gradients at the time of second catheterization were 29 mm Hg (range 23 to 34) and 15 mm Hg (range 13 to 28), respectively. The mean left atrial pressure ranged from 20 to 30 mm Hg (median 24). After repeat BMV, the median peak and mean left ventricular inflow gradients were 20 mm Hg (range 15 to 24) and 10 mm Hg (range 6 to 16), 32% and 43% median relative gradient reductions, respectively. All 4 patients had significant reductions in the MS gradient, and none had more than mild MR after repeat BMV. Three of these patients had their native MV on most recent follow-up; the other patient underwent redo BMV and cardiac transplantation, as described previously.

Three patients underwent MVR (2, 3, and 28 months after BMV) for residual or recurrent MS, along with moderate MR in a single case, and had placement of 21-, 25-, and 23-mm prostheses at 4, 6, and 14 years of age, respectively. Of the 2 patients who underwent replacement only a few months after BMV, 1 was found to have a supramitral ring at the time of MVR, possibly explaining BMV failure.¹⁸

At follow-up of 3.2 years (range 8 months to 9 years), among the 6 patients with a native MV (i.e., no MVR or transplantation), the maximum and mean Doppler MS gradients were 18 mm Hg (range 7 to 25) and 9 mm Hg (range 4 to 13), respectively. In this same cohort, the median weight and weight percentile for age had increased from 9.2 kg (range 4.6 to 62) at the time of BMV to 18.3 kg (range 9.8 to 78) at follow-up ($p < 0.001$) and from the 6th (range <3rd to >97th) to the 32nd (range <3rd to 95th) percentile ($p = 0.005$).

To assess the relationships between age, weight, MV prosthesis size, and the potential palliative effect of BMV, the surgical database at our institution was queried for patients with repaired AVC defects who weighed ≥ 50 kg and underwent MVR. Data were available for 69 patients. As illustrated in Figure 3, patients weighing >10 kg were able to accommodate larger prostheses more consistently than those weighing <10 kg.

Discussion

The optimal treatment for MS after the repair of an AVC defect has not been defined and probably varies according to the severity and substrate of MS, the degree of associated MR, and the size of the patient. Although surgical valvuloplasty and MVR remain the only

therapeutic options for patients with significant coexistent MS and MR, patients with milder degrees of MR may benefit from BMV, particularly to allow somatic growth and the eventual acceptance of a larger prosthesis. This approach is supported by evidence that outcomes after MVR may be worse in the setting of previous AVC repair than for other indications^{20 and 21} and with younger age, smaller valve size, and supra-annular position.^{14, 15 and 21} As demonstrated in this series, BMV results in substantial relief of MS in most patients, without a dramatic increase in MR. Most patients (60%) had their native MVs at median follow-up of 5.4 years, with less MS than before BMV, postponing MVR and the associated mortality and morbidities, which include obligate anticoagulation, heart block and pacemaker implantation, and probable redo MVR.²² Perhaps most important, in the 6 patients who remained with their native MV, median weight and weight percentile increased significantly. Given the association between age, size, and prosthesis size (Figure 3), should these patients ultimately undergo MVR, they will likely receive larger prostheses than if they had undergone MVR at the time of BMV. However, unlike patients who undergo BMV for congenital or rheumatic MS, patients with postoperative MS frequently have significant baseline MR, and although relief of postoperative MS by BMV may derive from opening the sutured cleft, dilation of interchordal spaces, or partial leaflet tears, all may result in increased MR. Given these considerations, BMV may not be a definitive treatment option for all patients with postoperative MS but warrants serious consideration as palliative therapy, with continued careful patient selection and long-term follow-up.

Acknowledgements

This study was supported by the Department of Cardiology John F. Keane Operating Fund and the National Institutes of Health under award number: T32HL007572. The content is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health.

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Figure 1.

(A) Mean left ventricular inflow gradients at time of catheterization were consistently higher than predicted Doppler MS gradients. (B) BMV reduced median peak and mean left ventricular inflow gradients in most patients.

Figure 2.

In 1 patient who had minimal reduction in left ventricular inflow gradient after BMV, comparison of 3-dimensional echocardiography before (A) and after (B) the procedure shows a previously closed MV cleft was clearly open (*arrow* denotes open cleft).

Table 1
Summary of patient demographics and hemodynamic data before and after balloon valvuloplasty

Patient	Age (mos)	Duration Since Repair	Weight (kg)	AVC Type	Status	Echocardiography			Catheterization			Reintervention, Duration After BMV
						Doppler MV Gradient (mm Hg)		Mean	MV Gradient (mm Hg)		% Reduction MV Gradient	
						Maximum	Peak		Mean			
1	8	4.4	6.5	C	Pre	27		15		12	17%	BMV, 3 mos
					Post	9		5		10		
2	9	6.3	4.6	C	Pre	17		8	25	16	44%	
					Post	18		9	15	9		
3	12	5.7	8.1	C	Pre	25		15	30	22	50%	BMV, 9 mos, 4 yrs; OHT, 6 yrs
					Post	21		14	12	11		
4	13	11	10	C	Pre	29		15		14	29%	BMV, 18 mos
					Post	18		10		10		
5	19	14	8.3	C	Pre	30		13	28	15	20%	BMV, 3 mos
					Post	18		11	14	12		
6	43	41	13.4	C	Pre	25		15	29	22	0%	MVR, 2 mos
					Post					22		
7	45	43	9.6	P	Pre	24		13	23	18	39%	MVR, 29 mos
					Post	12		11	12	11		
8	89	33	11.8	T	Pre			10		13	54%	
					Post	11		4		6		
9	142	83	62	P	Pre			10	26	15	7%	
					Post				26	14		
10	167	166	43	P	Pre			16	28	21	52%	MVR, 3 mos

C = complete; OHT = orthotopic heart transplantation; P = partial; T = transitional.