The role of enucleation with or without septal myectomy for discrete subaortic stenosis

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Objective: Substantial controversy persists regarding the need and efficacy of a routine myectomy in the treatment of discrete subaortic stenosis. Although some believe myectomy more effectively relieves subaortic narrowing, this is uncertain, and complications, including heart block and aortic valve injury, are concerns. The aims of the study were as follows: (1) to analyze the role of enucleation for relief of subaortic stenosis and the risk factors associated with recurrence and reoperation and (2) to delineate the characteristics of the patients who might benefit from enucleation alone.

Methods: From January 1990 through May 2007, 221 patients with subaortic stenosis underwent biventricular repair. Of those, 106 patients had discrete subaortic stenosis. The preoperative peak left ventricular outflow tract gradient, as determined by means of transthoracic echocardiographic analysis, was 67.3 ± 29 mm Hg. Forty patients had previous operations for other intracardiac anomalies. Mean age at repair was 7 years. Sixty-one patients underwent isolated enucleation, and 45 patients underwent concomitant myectomy. Patients with recurrent subaortic stenosis whose first operation was performed elsewhere were excluded from analysis.

Results: There was 1 early death and 1 late death. The postoperative peak left ventricular outflow gradient decreased to 12.5 ± 12.9 mm Hg ($P < .001$). No patient had development of heart block or required a pacemaker. A recurrent gradient of greater than 30 mm Hg was found in 26 (27%) patients, and 8 (7.5%) patients had reoperations. Actuarial freedom from reoperation rates at 5, 10, and 15 years were $94.7\% \pm 1.8\%$, $89.6\% \pm 3.5\%$, and $84.8\% \pm 4.9\%$, respectively. Of those patients who had not undergone a previous cardiac operation, there were no significant differences in the rates of recurrence (28% vs 27%) or reoperation (4.7% vs 4.4%) between the enucleation group and the concomitant myectomy group. For the patients who had a previous cardiac operation, the concomitant myectomy group had a significantly lower rate of recurrence (44% for enucleation vs 13% for enucleation plus myectomy, $P = .031$).

Conclusions: For those patients undergoing primary operations for discrete subaortic stenosis, routine myectomy does not offer superior relief of left ventricular outflow tract obstruction; enucleation alone provides good results in this selected population. However, in those patients with associated cardiac anomalies, concomitant additional myectomy is recommended.

Discrete subaortic stenosis is generally considered a progressive disease that can be isolated¹ or associated with other heart defects.^{2,3} The results of different surgical techniques have been described, with no clear benefit regarding the need and efficacy of a routine myectomy. Although some believe myectomy more effectively relieves subaortic narrowing,^{4,6} this is uncertain, and complications, including heart block and aortic valve injury, are concerns. The aims of the study were as follows: (1) to analyze the role of enucleation for relief of subaortic stenosis and the risk factors associated with recurrence and reoperation and (2) to delineate the characteristics of the patients who might benefit from enucleation alone.

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MATERIALS AND METHODS

This study received approval by the Institutional Review Board of Columbia University. We retrospectively reviewed the cardiac surgery database and catheter intervention database at the Morgan Stanley Children's Hospital of New York for those patients with the diagnosis of discrete subaortic stenosis who underwent resection of subaortic stenosis between January 1990 and May 2007. Discrete subaortic stenosis was defined as a discrete subaortic fibrous membrane or a fibromuscular ridge beneath the aortic valve. Patients with severe valvular aortic stenosis, a tunnel form of subaortic stenosis, and hypertrophic obstructive cardiomyopathy were excluded from the study. Patients with recurrent subaortic stenosis whose first operation was performed elsewhere were also excluded.

The median age at the time of the operation was 5.4 ± 4.9 years (range, 0–20 years). The left ventricular outflow tract (LVOT) pressure gradient was estimated by means of continuous-wave Doppler scanning. The peak gradient across the LVOT was 67.3 ± 29 mm Hg (range, 20–154 mm Hg). Complex discrete subaortic stenosis was defined as discrete subaortic stenosis with a previous intracardiac operation. Forty (37.7%) patients had previous intracardiac operations, and 8 patients had previous repair for coarctation of the aorta. Aortic valve regurgitation was present in 31 patients. It was mild in 24 patients and moderate in 7 patients (Table 1).

Operation

Operations were performed with cardiopulmonary bypass, mild hypothermia (32°C – 34°C), left ventricular venting, aortic crossclamping, and use of cold antegrade blood cardioplegia. After a transverse aortotomy extending

Abbreviations and Acronyms

CI = confidence interval
LVOT = left ventricular outflow tract

into the noncoronary sinus of the aortic valve, transaortic valve membrane excision, myectomy, or both was performed through the aortic valve with a combination of blunt and sharp dissection. An effort was made to remove all fibrous tissue, including that encroaching on the aortic and mitral valves. The additional myectomy is up to the discretion of the surgeon; however, our practice is not to routinely perform a myectomy. After the membrane resection, the LVOT was observed through the aortic valve, and if it was considered to be equal to or larger in area when compared with the aortic annulus, additional myectomy was not performed (61 [58%] patients).

Follow-up

Reviews of clinical records and contacts with the referring cardiologists served for collection of hospital and follow-up data. Echocardiographic reports were available for all patients. Mean follow-up was 6.9 ± 5.2 years.

Indication for Reoperation

Indications for reoperation were (1) a peak LVOT gradient of greater than 50 mm Hg and (2) left ventricular hypertrophy. For the patients undergoing multiple reoperations, we have been more conservative.

Statistical Analysis

Continuous variables were expressed as the mean $\pm$ standard deviation. The univariate association between risk factors and postoperative outcome was assessed by using either the χ^2 test for dichotomous risk factors or a *t* test for continuous risk factors. Logistic regression analysis of the recurrence and reoperation was used to evaluate the risk factors in a multivariate manner. The Kaplan–Meier method was used to estimate the freedom from reoperation. Analyses were carried out with JMP for Windows (SAS Institute, Inc, Cary, NC).

RESULTS

Survival

There was 1 hospital death (operative mortality, 0.9%) and 1 late death. One hospital death occurred for the patient

TABLE 1. Preoperative characteristics of the patients

Male/female sex	61/45
Age (y)	7.2 $\pm$ 4.9 (0–20)
Preoperative peak LVOT gradient (mm Hg)	67.3 $\pm$ 29 (20–154)
Preoperative AI	31
Mild	24
Moderate	7
Previous operation	
Intracardiac operation	40
CoA/VSD	4
IAA/VSD	7
Complete AVSD	14
Incomplete AVSD	5
VSD	3
DORV	2
TOF	5
Nonintracardiac operation	8
CoA	8

LVOT, Left ventricular outflow tract; AI, aortic insufficiency; CoA, coarctation of the aorta; VSD, ventricular septal defect; IAA, interrupted aortic arch; AVSD, complete atrioventricular septal defect; DORV, double-outlet right ventricle; TOF, tetralogy of Fallot.

who underwent resection of subaortic stenosis and mitral valve replacement after repair of an incomplete atrioventricular septal defect. One late death was for the patient who underwent subaortic resection after repair of an incomplete atrioventricular septal defect. Nine years after the subaortic resection, the patient presented with severe mitral regurgitation and severe aortic regurgitation with congestive heart failure. The patient did not respond to maximal medical therapy and died 5 days after admission.

Recurrence/Reoperation

All patients underwent transthoracic echocardiographic analysis at discharge. The postoperative peak gradient across the LVOT decreased to 12.5 ± 12.9 mm Hg ($P < .01$) at discharge. However, the peak gradient increased to 22.5 ± 22

FIGURE 1. Results of the patients with isolated and complex discrete subaortic stenosis.

FIGURE 2. Actuarial freedom from reoperation for the isolated and complex groups. *CI*, Confidence interval.

mm Hg during the mean follow-up period of 6.9 ± 5.2 years. Twenty-six (27%) patients had a recurrent LVOT peak gradient of greater than 30 mm Hg, and 8 (7.5%) patients underwent reoperation for recurrent subaortic stenosis. Actuarial freedom from reoperation rates at 5, 10, and 15 years were 94.7% (95% confidence interval [CI], 90% to 97%), 89.7% (95% CI, 81% to 95%) and $84.9\% \pm 4.9\%$ (95% CI, 73% to 92%), respectively.

Isolated Discrete Subaortic Stenosis

Of those patients who had not undergone a previous intracardiac operation (66 patients), 3 underwent reoperation (Figure 1). Actuarial freedom from reoperation rates at 5, 10, and 15 years were 96.7% (95% CI, 92% to 99%), 93.5% (95% CI, 82% to 98%), and 90.4% (95% CI, 75% to 97%), respectively (Figure 2). In this group 23 (35%) patients underwent concomitant myectomy. There were no significant differences in the rates of recurrence (23% vs 30%) or reoperation (4.7% vs 4.4%) between the enucleation group and the concomitant myectomy group. Multivariate analysis revealed the existence of aortic coarctation as an independent risk factor for reoperation ($P = .023$) and younger age ($P = .0094$) and higher preoperative LVOT gradient ($P = .0226$) as independent risk factors for recurrence (Table 2).

Complex Discrete Subaortic Stenosis

Among the patients who had a previous intracardiac operation (40 patients), 5 underwent reoperation. Actuarial freedom from reoperation rates at 5, 10, and 15 years were 91.7% (95% CI, 72% to 97%), 84.1% (95% CI, 67% to 93%), and 77.1% (95% CI, 56% to 90%), respectively (Figure 2). In the complex group 22 (55%) patients underwent the concomitant myectomy. In this complex discrete subaortic stenosis group, the patients who underwent concomitant myectomy had a significantly lower rate of recurrence (44% for enucleation vs 13% for enucleation with myectomy, $P = .031$). In the multivariate analysis existence of aortic coarctation or interrupted aortic arch ($P = .0419$)

TABLE 2. Risk factors analysis for recurrence and reoperation for the patients with isolated discrete subaortic stenosis

	<i>P</i> value		Odds ratio
	Univariate	Multivariate	
Recurrence (peak LVOT gradient >30 mm Hg)			
SubAS resection	.674		
without myectomy			
CoA	.121		
Age (younger)	.0118*	.0094*	0.97 (0.94–0.99)
Preoperative LVOT gradient	.0428*	.0226*	1.34 (1.10–1.72)
Preoperative AI	.2655		
Reoperation			
SubAS resection	.910		
without myectomy			
CoA	.0212*	.0233*	19.0 (1.60–447)
Age (younger)	.0618		
Preoperative peak LVOT gradient	.173		
Preoperative AI	.161		

LVOT, Left ventricular outflow tract; SubAS, subaortic stenosis; CoA, coarctation of the aorta; LVOT, left ventricular outflow tract; AI, aortic insufficiency. * $P < .05$.

and enucleation without myectomy ($P = .0416$) were independent risk factors for reoperation in the multivariate analysis, and enucleation without myectomy was an independent risk factor for recurrence in multivariate analysis (Table 3).

Although there was no new complete heart block after the subaortic resection, new left bundle branch block occurred in 5 (11%) patients only in the concomitant myectomy group.

TABLE 3. Risk factors analysis for recurrence and reoperation for the patients with previous intracardiac operation (complex group)

	<i>P</i> value		Odds ratio
	Univariate	Multivariate	
Recurrence (most recent peak LVOT gradient >30 mm Hg)			
SubAS resection	.0309*	.0392*	5.19 (1.16–28.7)
without myectomy			
CoA/IAA	.8842		
Age (younger)	.3787		
Preoperative LVOT gradient	.9899		
Preoperative AI	.755		
Reoperation			
SubAS resection	.0698	.0449*	19.4 (1.65–762)
without myectomy			
CoA/IAA	.0819	.0416*	15.5 (1.44–411)
Age (younger)	.3313		
Preoperative LVOT gradient	.8822		
Preoperative AI	.4341		

LVOT, Left ventricular outflow tract; SubAS, subaortic stenosis; CoA, coarctation of the aorta; IAA, interrupted aortic arch; AI, aortic insufficiency. * $P < .05$.

FIGURE 3. Mosaic plots of the additional myectomy and freedom from reoperation by era for the isolated (A) and complex (B) groups.

Coarctation of the Aorta/Interrupted Aortic Arch

There were 19 patients who underwent previous operations for coarctation of the aorta or interrupted aortic arch. Although it did not reach statistical significance ($P = .120$), the patients who underwent enucleation without myectomy tended to have more reoperations ($n = 3/6$ [50%]) than those who underwent concomitant myectomy ($n = 2/13$ [15.3%]).

Era

Because of the recent arguments regarding the necessity for the additional myectomy in the subaortic stenosis, the ratio of additional myectomy has increased in time in both the isolated (Figure 3, A) and complex (Figure 3, B) groups. In

the isolated group the reoperation rate has not changed with the increased ratio of additional myectomy; however, in the complex group the improvement in the reoperation ratio was observed in accordance with the additional myectomy.

DISCUSSION

Despite the surgical results for the relief of discrete subaortic stenosis being excellent in the short term to midterm, recurrence requiring reoperation is not uncommon and is reported to be between 0%⁴ and 19%.^{1,7,8} Because the subaortic lesion is associated to a certain degree with hypertrophy of the interventricular septum and because of its progressive nature, aggressive myectomy, in addition to the membrane resection, is recommended by several

authors.⁴⁻⁶ However, this has not been confirmed in other studies.⁹⁻¹¹ In our study we divided the patients into 2 groups and found out that additional myectomy benefits the patients who underwent previous intracardiac operations. Morphologic abnormalities in the LVOT might result in altered septal shear stress, which can lead to cell proliferation and recurrent obstruction in the LVOT. Therefore additional myectomy, in addition to relieving the subaortic stenosis, should be attempted to treat the anatomic abnormality causing increased septal shear stress in this complex group. On the other hand, in the patients with isolated discrete subaortic stenosis, we found no statistically significant difference with regard to the risk of recurrence or reoperation. In discrete subaortic stenosis the anterior mitral valve leaflet is often covered by a layer of abnormal fibrous tissue.¹² When this is left behind, the mitral valve stays drawn toward the septum, leaving the LVOT narrow. Peeling off this layer from the mitral valve often widens the LVOT. With our results, we believe that the complete removal of abnormal fibrous tissue from the LVOT, rather than additional myectomy, is important in the prevention of the recurrence of subaortic stenosis.

The existence of coarctation of the aorta or interrupted aortic arch was an independent risk factor for reoperation in both the isolated and complex groups. In this patient cohort, although it did not reach statistical significance, the concomitant myectomy group had higher freedom from reoperation.

Limitations to the study are inherent to the retrospective nature of data retrieval. The size of the aortic annulus and mean LVOT gradient were not recorded in all cases, and therefore their analyses were excluded from this study. The criteria for performing concomitant myectomy were solely based on the surgeon's discretion after the enucleation of the fibromuscular ridge, and therefore there might be a bias toward performing myectomy for more severe obstruction, although it was not shown in our study. Nevertheless, any recommendations for surgical intervention must be considered in light of the recognition that additional myec-

tomy carries a risk, such as complete heart block and perforation of the interventricular septum.^{1,9}

In conclusion, on the basis of our data, routine myectomy does not offer superior relief of LVOT obstruction for those patients undergoing primary operations for discrete subaortic stenosis; enucleation alone provides good results in this selected population. However, in those patients with associated cardiac anomalies, concomitant additional myectomy is recommended.

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