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% ± 1.8%, 89.6% ± 3.5%, and 84.8% ± 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.

CHD

Discrete subaortic stenosis is generally considered a progressive disease that can be isolated[1] 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 ± 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 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.

TABLE 1. Preoperative characteristics of the patients

Male/female sex	61/45
Age (y)	7.2 ± 4.9 (0–20)
Preoperative peak LVOT gradient (mm Hg)	67.3 ± 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.

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

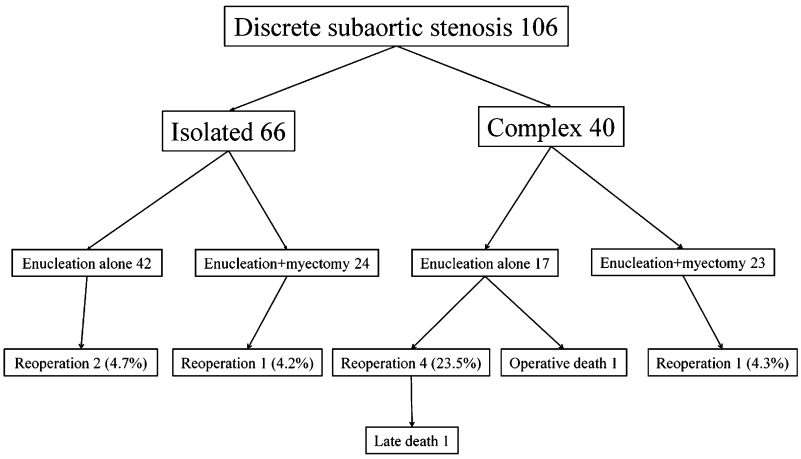

FIGURE 1. Results of the patients with isolated and complex discrete subaortic stenosis.