Original Article

Discrete Subvalvular Aortic Stenosis: Severity of Aortic Regurgitation and Rate of Recurrence at Midterm Follow-Up after Surgery

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Received 29 July 2008 ; Accepted 16 September 2008

Abstract

Background: *Discrete subaortic stenosis (DSS) is a progressive condition. Controversy still rumbles on as to whether the subaortic membrane causes aortic regurgitation (AR) and whether membrane resection reduces AR severity. We investigated the association between the left ventricular outflow tract peak gradient (LVOT-PG) and AR severity preoperatively and changes in AR severity and obstruction recurrence after surgery in DSS patients.*

Methods: *Twenty patients were evaluated before and after surgery for DSS (mean follow-up time: 13.60±9.61 months). The patients were evaluated via transthoracic echocardiography and transesophageal echocardiography, if necessary. The cut-off point for surgery was LVOT-PG ≥50 mmHg or the presence of progressive AR.*

Results: *The mean age of the patients was 28.55±15.23 years, and 35% of them were male. LVOT-PG decreased from a mean of 80.83±42.72 mmHg preoperatively to 19.14±14.03 mmHg postoperatively and to 25.47±16.10 at follow-up. AR was identified in 15 (75%) patients preoperatively: mild in 8 (40%) and moderate in 7 (35%). The postoperative change in AR severity was insignificant. The correlation between preoperative LVOT-PG and the incidence and severity of preoperative AR was not significant. AR severity had no correlation with age. Membrane recurrence occurred in 25% of the patients.*

Conclusion: *Our results indicated no relationship between AR severity and LVOT-PG and the patient's age. Patient selection for surgery can, therefore, be carried out on the basis of LVOT-PG or AR severity separately. Subaortic resection may reduce AR severity in some patients, but this reduction is not significant. Future studies are required to elucidate whether or not the presence of the AR is an indication for surgery.*

J Teh Univ Heart Ctr 4 (2008) 219-224

Keywords: *Discrete subaortic stenosis • Aortic valve insufficiency • Follow-up studies • Surgery*

Introduction

Discrete subaortic stenosis (DSS) is more common in childhood, but it is also a relatively frequent heart disease in adult patients with a prevalence of 6.5% of all adult congenital heart diseases.[1]

Fixed subaortic stenosis is responsible for up to 20% of the left ventricular outflow tract (LVOT) obstructions requiring intervention.[2] It generally occurs in one of two forms.The less common form is secondary to circumferential fibromuscular tunnels, but it more commonly results from a discrete membrane immediately below the aortic valve.[3]

Traditionally, it has been difficult to differentiate DSS from the valvular aortic stenosis; today, however, we may reliably determine a subaortic membrane masked by a hypertrophied and prominent ventricular septum using

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transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) techniques.[4,5]

The treatment and prognosis of subaortic stenosis have been studied sufficiently in children, whereas there is a dearth of data in the adult patient population.[1] It has been reported that aortic regurgitation (AR) secondary to DSS is common, is usually hemodynamically insignificant in childhood, tends to be progressive, and is related to the degree of LVOT obstruction.[2,6,7] Some investigators have stressed that in contrast to children, adult patients with fixed subaortic stenosis have a high incidence of significant AR.[2] There is a paucity of information to support the benefits of an early surgical intervention on the basis of LVOT gradient in patients with a progressive obstruction.

The main purpose of this study was to determine whether AR severity is related to the severity of LVOT obstruction and the patient's age and whether the severity of AR decreases after the surgical resection of the membrane.

Methods

All the patients with DSS who underwent surgical treatment between March 2002 and January 2006 at Tehran Heart Center, a hospital dedicated to heart diseases in the capital of Iran, Tehran, were evaluated prospectively. The study was approved by the institutional ethics committee. DSS was diagnosed when a fixed subvalvular obstruction causing a subaortic flow acceleration was detected by TTE (Figure1) and TEE, if necessary. TTE was performed for all the patients preoperatively, postoperatively, and at follow-up. Data on the patients' age, sex, previous cardiac surgery, coexisting cardiac anomalies (if any), symptoms, electrocardiography, echocardiography, and cardiac catheterization (if needed) were collected.

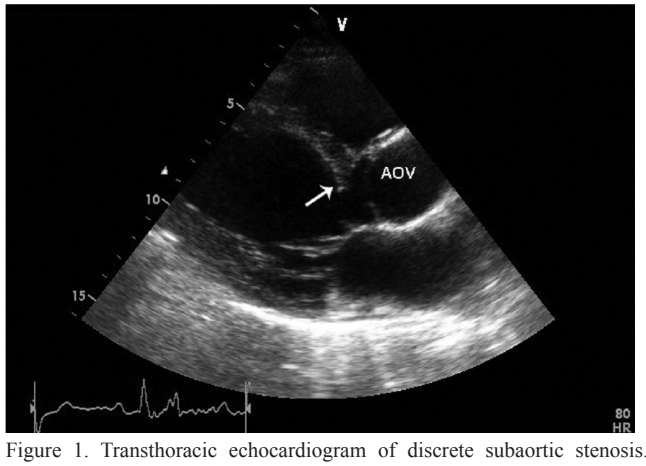

Figure 1. Transthoracic echocardiogram of discrete subaortic stenosis. There is a membrane below aortic valve which is attached to septum. There is another attachment to anterior mitral leaflet (arrow)

TTE (either the apical, parasternal, or suprasternal approach) was utilized to assess the left ventricular outflow tract peak gradient (LVOT-PG), presence of membrane, and degree of AR. If there was any doubt about the presence of a membrane, TEE was performed. Angiography and catheterization were applied for those over the age of 35 years. AR was evaluated according to the appearance of the regurgitation jet by Doppler; and it was categorized as mild, moderate, and severe (Table 1) in accordance with the American Society of Echocardiography Guidelines.[8]

Table 1. Quantitative evaluation of aortic regurgitation severity (American Society of Echocardiography Guidelines)

Parameter	Mild	Severe
Jet width/LVOT	<25%	≥65%
Vena contracta (cm)	<0.3	>0.6
Pressure half-time (ms)	>500	<200
Regurgitant volume (ml/beat)	<30	≥60
Regurgitant fraction (%)	<30	≥50
Regurgitant orifice area (cm²)	<0.1	≥0.3

LVOT, Left ventricular outflow tract

Indications for surgery tend to vary; nonetheless, they normally include LVOT-PG≥50 mmHg, echocardiographic or angiographic evidence of progressive AR,[2] and coexisting cardiac lesions that require surgery. All the operations were performed by the expert cardiac surgeons at the department of cardiac surgery in Tehran Heart Center. In line with techniques usually favored for subaortic resection, an oblique aortotomy was made and extended into the non-coronary sinus. The aortic valve was thereafter retracted carefully to reveal the subaortic membrane. Both the obstructing discrete membrane and the adjacent hypertrophied muscle (when indicated) were subsequently excised circumferentially as completely as possible. All the operations were carried out with great care to avoid injury to the conduction tissue between the right and non-coronary aortic cusps and the anterior leaflet of the mitral valve.

At least one TTE examination was performed for all the patients during hospitalization after surgery. The patients underwent second TTE at the first visit after being discharged. The changes in LVOT-PG and AR severity as well as the recurrence of the membrane were evaluated in both postoperative and follow-up examinations. The recurrence was determined in terms of the presence of the membrane during the follow-up.

The numerical variables were presented as mean±SD, and the categorized variables were summarized by percentages. The continuous variables were compared using the Student's t-test or the nonparametric Mann-Whitney U test whenever the data did not appear to have normal distributions. A significance level was considered ≤ 0.05.

The changes in the continuous variables before surgery, after surgery, and at follow-up were compared through the

repeated measures ANOVA, followed by the Wilcoxon Signed Rank test (considering the Bonferroni method to adjust the significance level of α). The correlations between age and AR severity were analyzed using the Spearman correlation coefficient. The Friedman test and Kendall tau rank correlation test for trend were also used in further analyses.

Results

A total of 20 patients diagnosed with DSS who underwent membrane resection were identified. The patients' mean age was 28.55±15.23 years (range 5 to 64). The group comprised 7 (35%) males and 13 (65%) females. One of the patients had previously undergone the resection of the subaortic membrane and had to have redo surgery for the recurrence of the membrane with a significant gradient. The mean follow-up time was 13.60±9.61 months. Seventeen (85%) patients were symptomatic: 10 (50%) had dyspnea, 3 (15%) had chest pain, 2 (10) had palpitation, and 2 (10%) had mixed complaints. For the remaining 3 patients (15%), who were without symptoms, diagnosis was made using echocardiography for an evaluation of cardiac murmur. The number of the patients with concomitant congenital heart disease was 3 (15%): ventricular septal defect in 1 (5%) and bicuspid aortic valve in 2 (10%). None of the patients had endocarditis preoperatively. Of the 20 patients in the study group, 5 (25%) had no AR preoperatively, whereas AR was identified in 15 (75%) patients: mild in 8 (40%) and moderate in 7 (35%). None of the patients had severe AR. There was no significant correlation between the patient's age and AR severity (r_s=0.099, P=0.677). The follow-up TTE showed that while 40% of the patients had no AR, 40% had mild and 20% had moderate AR (Figure 2). Of the patients who had moderate AR before surgery; 42.9% remained unchanged, 28.6% had mild AR, and 28.6% had no AR.

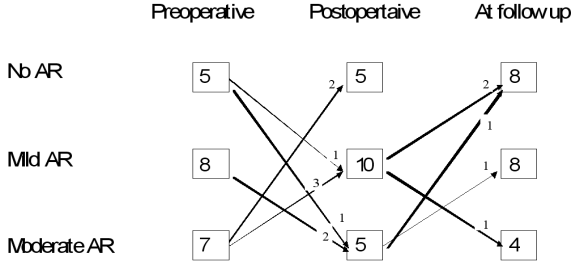

Figure 2. Aortic regurgitation (AR) changes in postoperative and follow-up transthoracic echocardiographies

Among the cases having preoperative mild AR; 75% showed no changes, 12.5% had no AR, and 12.5% progressed to moderate AR in the follow-up evaluations. The patients with no preoperative AR showed no regurgitation of the aortic valve at follow-up. AR severity was reduced after membrane resection; this change, however, was not significant. The correlation between AR severity and the distance of the membrane from the aortic valve was not significant (rs=-0.126, P=0.56).

The preoperative TTE showed a mean LVOT-PG of 80.83±42.72 mmHg. The mean LVOT-PG decreased substantially to 19.14±14.03 postoperatively and to 25.47±16.10 mmHg at follow-up (P<0.001) compared to that of preoperation. Early after surgery, LVOT-PG was less than 10 mmHg in only 30% of the patients, while 70% had still LVOT-PG >10 mmHg. At follow-up, 20% of the patients had LVOT-PG <10 mmHg.

There was no significant correlation between preoperative LVOT-PG and the incidence and severity of preoperative AR, and nor was there any association between preoperative LVOT-PG and the size of the membrane or the distance of the membrane from the aortic valve (r_s=0.270, P=0.24 and r_s=0.185, P=0.39, respectively). Membrane recurrence was detected in 5 (25%) patients at follow-up. The residual gradient in these patients ranged from 5 to 20 mmHg (mean 12.20±7.26 mmHg) in early postoperative and from 16 to 41 mmHg (mean 27.96±10.46 mmHg) in follow-up TTE. At follow-up, the increase in LVOT-PG was significant in the patients with recurrence by comparison with those without it (P=0.03); none of them required reoperation (LVOT-PG <50 mmHg and less than moderate AR). Table 2 depicts a comparison of LVOT-PG postoperatively and at follow-up between the patients with and without recurrence. The severity of LVOT obstruction did not have a significant influence on membrane recurrence after surgery.

Table 2. Comparison of LVOT PG (mmHg) post operatively and at follow up between patients with and without recurrence

	With recurrence	Without recurrence	P value
Postoperative	12.20±7.26	22.99±17.08	NS
At follow up	24.64±17.82	27.96±10.46	NS
Changes of LVOT PG	1.72±10.72	15.76±12.06	0.02

LVOT PG, Left ventricular outflow tract peak gradient; NS, Not significant

Discussion

DSS is a progressive cardiac abnormality which leads to LVOT obstruction by the subvalvular fibromuscular tissue. The main cause of obstruction is still a matter of debate, but there are various theories about the causes of DSS and it seems that the disease is multifactorial. It has been demonstrated that DSS is a pathological complex and that endocardial abnormality is a part of this complex begetting membrane development.[9]

De Vries and colleagues reported AR in 30% of their patients at the time of diagnosis, which increased to 54%

during a mean follow-up interval of 3.7 years.[2] We found AR in 75% of our patients, and there was no significant correlation between the presence or severity of AR and the mean age of the patients.

There has been a great deal of research into the changes in AR severity after surgery; still, the role of surgery and the time of intervention in this regard are unclear. The retrospective nature of the available reports and the heterogeneity of the lesion, together with its unpredictable natural history, are probably responsible for this uncertainty. Several series have reported the progression of AR despite surgery. Be that as it may, some with similar postoperative follow-up evaluations have demonstrated little change, and others have concluded that early surgery decreases the incidence of AR at late follow-up.[2] In addition, much as AR can progress despite a postoperative reduction in LVOT obstruction, the worsening of AR in DSS can be slowed or stopped with an adequate resection of the membrane.[3,9] In our study, AR severity was reduced after surgery in some patients; the reduction, however, was not statistically significant.

Some studies[10] have shown that patients with a lower preoperative systolic gradient have less significant late postoperative regurgitation and that early operation with adequate resection can preserve the integrity of the aortic valve to avoid later valve replacement. By contrast, other series have asserted that surgery does not have a beneficial impact on AR because both the incidence and severity of AR increase after surgery.[1,6]

Some researchers maintain that there is a relationship between the severity of LVOT obstruction and AR in children and adults[1,11,12] and that secondary aortic valve damage can be prevented if surgery is performed when LVOT mean gradient rises to more than 30 mmHg.[10] Our results did not show a significant correlation between LVOT-PG and the severity of AR before surgery. If future studies confirm our findings, there will be no more fear of the progression of AR with an increase in LVOT gradient, and the cut-off point for surgery could, as a result, rise. It is noteworthy that LVOT-PG remained more than 10 mmHg in 70% and 80% of our patients postoperatively and at follow-up, respectively.

The proportion of patients having symptoms varies from series to series. Rayburn and colleagues[3] confirmed that 70% of their cases had no symptoms. Darcin and associates[11] reported that 61.9% of their patients were asymptomatic. The majority of our patients were noted to be symptomatic (85%), the most common symptom being dyspnea (50%). This finding is probably due to the fact that the mean preoperative LVOT-PG in our cases was relatively higher than that in the previous series: 80.83±42.67 mmHg in our patients vs. 63.39±7.63 mmHg in the cases reported by Rayburn et al.[3] and 59.23±35.38 mmHg[11] in the patients of Darcin and his colleagues.

The recurrent LVOT obstruction requiring reoperation is at a rate of 4.8% to 30% in previous studies[11,12] Serraf et al. believed that recurrence and reoperation were significantly influenced by previous repair and by the quality of the initial relief of subaortic stenosis.[13] We observed a 25% recurrence rate, which chimes in with previous findings in the literature.

Conclusion

The findings of this study revealed that an increased LVOT-PG did not influence AR severity significantly. In addition, AR severity had no correlation with age; consequently, patient selection for surgery can be carried out on the basis of LVOT-PG or AR severity separately. The infrequent occurrence of the disease requires longer-term studies and longer follow-up periods with more patients if more light is to be shed on the course of postoperative DSS and a cut-off point for surgery.

Acknowledgement

This study was supported by Tehran Heart Center, affiliated with Tehran University of Medical Sciences. The authors wish to thank all the staff of the Research Department of Tehran Heart Center whoparticipated in gathering and processing the data.

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