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Short- and mid-term results of resection of discrete subaortic stenosis in children

Çocuklarda diskret subaortik darlık rezeksiyonunun kısa ve orta dönem sonuçları

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ABSTRACT

Background: In this study, we report our short- and mid-term surgical results in patients with discrete subaortic stenosis.

Methods: A total of 72 patients (43 males, 29 females; mean age 4.6±4.5 years; range, 6 months to 11 years) who were surgically treated for discrete subaortic stenosis in our clinic between September 2000 and January 2012 were retrospectively analyzed. Limited myectomy in addition to the transaortic excision of the stenotic structure was performed in all patients. Left ventricular outflow tract gradient and aortic valve regurgitation were evaluated by echocardiography following the procedure.

Results: The mean follow-up was 4.9±2.7 (range, 1 to 10) years. No mortality occurred during the early and late postoperative period. Heart block which necessitated the placement of a permanent pacemaker developed in one pediatric patient. The mean preoperative left ventricle systolic gradient which was measured as 48.4±13.7 mmHg (range, 32 to 70 mmHg), diminished to 8.6±5.6 mmHg (range, 4 to 15 mmHg) in the postoperative period. Reoperation was necessary due to recurrent stenosis in two patients (2.7%) during follow-up.

Conclusion: Our study results show that limited myectomy in addition to membranectomy results in good mid-term results without increasing morbidity and mortality.

Keywords: Aortic regurgitation; discrete subaortic stenosis; pediatric heart surgery; subaortic membrane.

ÖZ

Amaç: Bu çalışmada, diskret subaortik darlık olan hastalarda kısa ve orta dönem cerrahi sonuçlarımız sunuldu.

Çalışma planı: Eylül 2000 - Ocak 2012 tarihleri arasında kliniğimizde diskret subaortik darlık nedeniyle cerrahi tedavi uygulanan toplam 72 hasta (43 erkek, 29 kadın; ort. yaş 4.6±4.5 yıl; dağılım, 6 ay - 11 yıl) retrospektif olarak incelendi. Tüm hastalara transaortik stenotik yapı eksizyonuna ek olarak sınırlı miyektomi uygulandı. İşlem sonrası sol ventrikül çıkım yolu gradyanı ve aort kapak yetmezliği ekokardiyografi ile değerlendirildi.

Bulgular: Ortalama takip süresi 4.9±2.7 (dağılım 1-10) yıl idi. Ameliyat sonrası erken ve geç dönemde mortalite gözlenmedi. Bir pediatrik hastada kalıcı kalp pili takılmasını gerektiren kalp bloku gelişti. Sol ventrikül çıkım yolu ortalama sistolik gradyanı ameliyat öncesi 48.4±13.7 mmHg'den (dağılım 32-70 mmHg), ameliyat sonrası dönemde 8.6±5.6 mmHg'ye (dağılım 4-15 mmHg) geriledi. Takip sırasında iki hastada (%2.7) tekrarlayan darlık nedeniyle tekrar ameliyat gerekti.

Sonuç: Çalışma bulgularımız, membranektomiye ek olarak sınırlı miyektominin mortalite ve morbiditeyi artırmadan iyi orta dönem sonuçları olduğunu göstermektedir.

Anahtar sözcükler: Aort yetmezliği; diskret subaortik darlık; pediatrik kalp cerrahisi; subaortik membran.

Discrete subaortic stenosis, which is defined as a lesion causing stenosis in the inferior part of the aortic valve and the left ventricle outflow tract (LVOT), is considered as an acquired disease.[1] It is rarely diagnosed antenatally or during infancy.[2] It often manifests in the first decade of life with features of progressive LVOT obstruction.[1,2]

Discrete subaortic stenosis may lead to left ventricular hypertrophy and regurgitation, as well as arrhythmias, aortic valve regurgitation, and endocarditis.[3,4] Although surgical treatment is recognized as an effective method of treatment, the rates of recurrence of stenosis and requirement of reoperation have been reported to be up to 50% with varying figures according to the results from two studies[5,6]

In this study, we report our short and mid-term surgical results in patients with discrete subaortic stenosis.

PATIENTS AND METHODS

A total of 72 patients (43 males, 29 females; mean age 4.6±4.5 years; range, 6 months to 11 years) who were surgically treated for discrete subaortic stenosis in the Pediatric Cardiovascular Surgery Clinic between September 2000 and January 2012 were retrospectively analyzed. The patients with the diagnosis of tunnel-type subaortic stenosis were excluded, as the surgical approach and long-term results of this condition may vary. The data of the patients were recorded. The study was conducted in accordance with the directions of the institutional ethics committee and with the principles of the Declaration of Helsinki.

Subvalvular aortic stenosis was defined as a lesion with a discrete fibrous, membranous, or fibro-muscular structure under the aortic valve in the LVOT in consistent with the literature data.[7] The diagnosis of discrete subaortic stenosis was performed by transthoracic echocardiography (TTE). Residual gradient and aortic valve competence were assessed by intraoperative transesophageal echocardiography (TEE) after cardiopulmonary bypass (CPB). All patients were followed on a regular basis by TTE to assess recurrent subaortic stenosis and aortic valve competence.

Doppler flow mapping and continuous wave Doppler echocardiography techniques were used to calculate the maximal flow rate in the LVOT. The maximum flow gradient was calculated according to the simplified Bernoulli formula. Aortic regurgitation was graded as trivial/mild/moderate/severe according to the backflow jet as confirmed by color Doppler ultrasound at aortic valve level.[8]

The surgical indication was a mean LVOT gradient of ≥30 mmHg in the presence of normal ejection fraction in patients who were diagnosed with discrete subaortic stenosis. In patients with a mean pressure gradient of <30 mmHg, surgical indications were the presence of concomitant cardiac abnormality necessitating surgical repair, the presence of progressive aortic regurgitation, development of left ventricle systolic dysfunction or development of left ventricle hypertrophy. On the other hand, indication for reoperation was an outflow tract gradient of >40 mmHg with or without progressive aortic valve regurgitation and left ventricular hypertrophic changes.

The mediastinum was accessed through a median sternotomy in all operations. The operation was performed under moderate hypothermic CPB with standard cannulation and using antegrade blood cardioplegia. Following transverse aortotomy, the lesion causing stenosis was outlined under the aortic leaflets. The tissue extensions of the membranous or fibromuscular structure located in the LVOT under the aortic annulus and protruding to the aortic valvulae or mitral valve were completely resected. A special care was paid to preserve the ventricular septum, valvular structures, and transmission tissues.

Statistical analysis

Statistical analysis was performed using the SPSS for Windows version 16.0 software program (SPSS Inc., Chicago, IL, USA). Data were expressed in mean ± standard deviation. In addition, paired mean results were analyzed using the Student's t-test. A p value of <0.05 was considered statistically significant.

RESULTS

The mean weight and body surface area of the patients were 11.1±2.3 kg (range, 7 to 25) and 0.8±0.15 m^2 (range, 0.5 to 1.1), respectively. The mean preoperative LVOT gradient was 48.4±13.7 mmHg (range, 32 to 70 mmHg). Aortic regurgitation was trivial in 20 patients, mild in 28 patients, and moderate in seven patients (55/72; 76.3%). Forty-six patients (63.8%) were asymptomatic. On the other hand, symptomatic patients had complaints of palpitation, dyspnea, chest pain, or dizziness on exertion. Concomitant congenital cardiac disease was present in 28 patients (38.8%). Demographic and clinical characteristics of the patients are presented in Table 1.

Perioperative TEE following resection revealed no residual gradient and impairment in the aortic valve competence and the discrete membrane was seen to

be completely excised. No mortality in the early or late postoperative period was encountered. The mean gradient in the early postoperative period before discharge was found to be 8.6±5.6 mmHg (range, 4 to 15 mmHg). In the early postoperative period, the left branch block was seen in eight patients, while bifascicular block and atrioventricular (AV) complete block were observed in one patient. Transvenous permanent pacemaker implantation was performed in one patient with AV complete block. This patient was a three-year-old girl weighing 13.4 kg.

The mean follow-up was 4.9±2.7 (range, 1 to 10) years. The mean gradient at the end of the follow-up period was 12.9±6.2 mmHg (range, 7 to 23 mmHg). Mid-term follow-up results are summarized in Table 1. In the short and mid postoperative period, there was a significant reduction in the LVOT (p<0.001). In addition, the reduction in aortic regurgitation was statistically significant in the postoperative period in all patients (p<0.001).

Three patients (4.1%) required reoperation. Indications for reoperations were recurrent subaortic stenosis in two male patients aged three and five, respectively (an outflow tract gradient of 40 and 45 mmHg, respectively). Severe aortic valve stenosis was found in a nine-year-old boy with the diagnosis of bicuspid aortic valve. The patients with recurrent subaortic stenosis had a residual gradient of 32 and 35 mmHg, respectively; however, no progression or symptom was seen in the early follow-up. These patients become symptomatic and surgically re-treated in the second and third year of follow-up, respectively. Myectomy was also added to the fibromuscular tissue resection procedure in these patients. During the postoperative period, the outflow tract obstruction was measured as 8 and 12 mmHg, respectively. The patients

Table 1. Demographic and clinical characteristics of patients

	n	%	Mean±SD	Range
Gender				
Male	43	59.7		
Female	29	40.3		
Mean age (years)			4.6±4.5	6 months to 11 years
Preoperative LVOT gradient (mmHg)			48.4±13.7	24 to 70
Associated cardiac anomaly	28	38.8		
Ventricular septal defect	22	30.5		
Aortic valve stenosis (Bicuspid)	1	1.4		
Mitral valve prolapse	5	6.9		
Length of stay of ICU (hours)			22.5±6.5	14 to 30
Length of stay of Hospital (days)			5.1±1.2	1 to 7
Complications				
Left bundle branch block	8	11.1		
Bifascicular block	1	1.3		
Complete atrioventricular block	1	1.3		
Aortic valve stenosis	1	1.3		
Recurrent stenosis	2	2.6		
LVOT gradient before discharge (mmHg)			8.6±5.6	4 to 15
Follow-up (years)			4.9±2.7	1 to 10
Preoperative aortic regurgitation				
Trivial	20			
Mild	28			
Moderate	7			
Severe	–			
Early postoperative aortic regurgitation				
Trivial	12			
Mild	8			
Moderate	3			
Severe	–			
LVOT gradient at mid-term follow-up (mmHg)			12.9±6.2	7 to 23

SD: Standard deviation; LVOT: Left ventricular outflow tract; ICU: Intensive care unit.

were discharged after an uneventful postoperative period. The postoperative follow-up examination demonstrated that the gradient disappeared. The patient who was followed for concomitant bicuspid aortic valve required reoperation due to aortic valve stenosis in the fourth year of follow-up. The patient underwent mechanical bileaflet aortic valve replacement and was discharged with medical treatment.

DISCUSSION

Despite gaining a better understanding, the pathogenesis of discrete subaortic stenosis is still controversial. Genetic, congenital, inflammatory, and acquired causes are usually held responsible for the development of this entity.[4,9] Although the idea of the acquired nature of the disease has gained popularity in recent years, since the disease is not present at birth but it develops and progresses afterwards, the presence of some familiar cases has suggested that it may also have a genetic etiology.[5]

The LVOT gradient is a surgical indication; however, many patients are asymptomatic.[9] Although the most common symptom is decreased exercise tolerance in symptomatic patients, angina pectoris and syncope may be also encountered.[9] In the present study, the rate of asymptomatic patients was 63.8% (n=46) and the most common symptom in symptomatic patients was exercise intolerance (14 patients, 19.4%). In addition, four patients suffered from syncope, seven patients from palpitation, and one patient from chest pain.

Discrete subaortic stenosis can be an isolated condition; however, almost half of patients have concomitant cardiac abnormalities.[9,10] Among these, ventricular septal defect (VSD), coarctation of aorta, atrioventricular septal defects, and aortic valve stenosis are the most commonly seen conditions.[1,11] In the present study, concomitant congenital cardiac disease was detected in 28 patients (38.8%) and the most common condition was VSD in 22 patients (30.5%).

Due to the presence of the membrane which causes stenosis in the subaortic area and turbulence, a gradient develops in the left ventricle outflow tract in addition to a turbulence-related thick and degenerative aortic valve.[3,4] Progressively increasing aortic valve regurgitation due to the presence of stenosis also increases the risk for infective endocarditis.[7,12,13] Therefore, the timing of surgery should be planned in patients with discrete subaortic membrane, considering the gradient in the LVOT and progression of the aortic regurgitation.

The main goal of surgical treatment in patients with discrete subaortic stenosis is to resolve the stenosis and to prevent the development of aortic regurgitation and endocarditis.[9] However, progression of the aortic regurgitation may continue in patients with a high preoperative LVOT gradient.[3,14,15] Although this gradient decreases to hemodynamically insignificant values, patients should be followed for aortic regurgitation and subaortic gradient after surgery.[1] Although the aortic valve functions might be affected by more than one factor during follow-up, we consider that early surgery slows the rate of degeneration of the aortic valve and, thus, decreases the need for valvular replacement.

Furthermore, surgery is currently associated with low mortality and morbidity rates.[3,16] Although the optimal surgical approach is controversial, several aggressive methods such as simple membrane resection,[3,7,17] myomectomy,[10,18] and circumferential myomectomy and mobilization of right-to-left fibrous trigone[11] remain as surgical options. In our study, we performed limited resection of the hypertrophic muscle tissue in addition to the fibrous membrane resection in all cases. The use of intraoperative TEE helps to detect the location and extension of the resection and thus to prevent a residual gradient from remaining. As a result, surgery-related heart block and iatrogenic VSD due to operations can be prevented.

It was reported that the progression of aortic regurgitation decreased and halted in patients with a low residual systolic pressure gradient following surgical intervention.[8,9,19] Similarly, in the present study, the progression of aortic regurgitation halted in patients without subaortic stenotic recurrence postoperatively. Therefore, surgery should be planned before aortic regurgitation develops or when minimal regurgitation starts during follow-up. The tissue which causes stenosis and turbulent flow should be completely resected during the operation.

The rate of recurrent LVOT stenosis after surgery has been reported to vary between 7 to 27% in various studies, although the rate may vary according to the degree of the pathology and the procedural technique.[13,14,18] In our study, two patients (2.7%) required reoperation due to restenosis in the mid-term.

Furthermore, low mid-term recurrence rates in our study can be attributed to the exclusion of patients with complex LVOT stenosis, operation planning with intraoperative TEE, and attempting to excise all the tissue leading to stenosis.

This study has some limitations. Small sample size and retrospective design of the study can be regarded as limitations. In addition, elective echocardiography was unable to be performed in all patients on a regular basis during follow-up, as some of these patients lived in distant provinces of the country. Despite these drawbacks, we still believe that our findings are relevant.

In conclusion, discrete subaortic stenosis is commonly diagnosed incidentally, as it is not frequently present at birth and often develops later. It is asymptomatic in more than half of patients and may cause progressive aortic valve regurgitation and endocarditis due to the abnormal flow type through the left ventricle outflow tract. Since it is a progressive condition and the presence of high outflow tract gradient is considered as a poor prognostic factor, the treatment of the disease should be performed without undue delay. Based on our study results, we recommend limited myectomy in addition to membranectomy to yield good mid-term results without increasing morbidity and mortality.

Declaration of conflicting interests

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