Fixed Subaortic Stenosis in Childhood

Medical and Surgical Course in 90 Patients

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Summary

This study evaluated diagnostic criteria, clinical observations, surgical indications and complications in 90 cases of fixed subvalvular aortic stenosis. Echocardiography, catheter angiography and surgical findings indicated that 60 (66.6%) patients had subvalvular membranous stenosis, 20 (22.2%) patients had fibromuscular stenosis, 4 (4.4%) patients had membranous and fibromuscular stenosis, and 6 (6.6%) had a tunnel-type obstruction. Forty-two patients had additional cardiac anomalies, such as PDA, valvular aortic stenosis and VSD. Fifty patients were treated surgically. Thirty-six patients were evaluated by two-dimensional and Doppler echocardiography at post-surgical intervals ranging from 7 days to 11 years (mean 3.6 years). There were pathological echo findings in 12 patients. The prognosis of this anomaly depends upon early diagnosis and treatment. The results of this study support the importance of the two-dimensional and Doppler echocardiography in early diagnosis and follow-up of the surgical results in treatment of this anomaly.

Key Words:
Fixed subaortic stenosis Discrete subaortic stenosis Two-dimensional echocardiography Doppler echocardiography

FIXED subaortic stenosis is an uncommon congenital cardiac anomaly in which the left ventricular outflow tract is narrowed by either a fibrous membrane, a muscular ridge or a fibromuscular tunnel, either singly or in various combinations.[1] The most common form of fixed subaortic stenosis is the subaortic membrane type. The obstruction and hemodynamic im-

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pairment are only two problems associated with this anomaly: its complications include aortic regurgitation, infective endocarditis and possible development of an inherently associated muscular obstruction.[1)-6)] This report reviews the diagnostic criteria, clinical observations, surgical indications and complications in a series of 90 cases of subvalvular aortic stenosis. The aim of this study is to discuss the characteristics of subaortic stenosis seen in childhood, to emphasize surgical indications and to determine the role of echocardiography in the correct diagnosis and follow-up.

Materials and Methods

Ninety patients with fixed subaortic stenosis were evaluated at the Hacettepe Children's Hospital between 1984 and 1990. Their ages ranged from 1 year to 24 years (mean 9.4). There were 32 females and 58 males. The diagnosis was made according to the patient's history, physical examination, telecardiography, electrocardiographic findings, echocardiography and cardiac catheterization. Two-dimensional, M-mode, pulse and continuous wave Doppler echocardiographic examinations were performed using a Toshiba model SSH-60A with 2.5–3.75–5 MHz transducers. Transducers were positioned in the left parasternal long-axis and left apical long-axis views to determine the presence and type of obstruction. When the transducer was at the jugular notch, subxyphoidal or left parasternal location, the stenotic area was localized with pulse Doppler echocardiography. Continuous wave Doppler methods were used to identify the transaortic pressure gradient. In addition, the diastolic flow pattern of aortic insufficiency was noted.

During cardiac catheterization, the gradient was measured between the left ventricular apex and subvalvular area. Left ventricular angiography and arteriography were performed. Right side catheterization was conducted in some patients.

The previously accepted criteria for surgical intervention were initially the existence of subvalvular stenosis in echocardiography or angiocardiography and the existence of a severe or progressive transaortic gradient. More recently, a transaortic gradient measurement higher than 25 mmHg has also been added as a third criterion. Membrane resection, myotomy, myectomy or ventriculoplasty have been performed with open cardiac surgery techniques. Seventy patients were scheduled for surgery, but only 50 operations were performed. Thirty-six patients were followed with two-dimensional and Doppler echocardiography after surgery to test for the existence of recurrent or residual obstruction.

Results

The distribution of cases according to their age groups is shown in Table I. Forty-five cases had symptoms. These were fatigue and palpitation in 28, chest pain in 10 and syncope in 7 patients. Forty-two cases were shown to have cardiomegaly on telecardiography, 3 had dilatation of the ascending aorta, while the other 45 had normal telecardiographic findings. Subvalvular membranous obstruction was detected in two siblings from the same family.

Electrocardiographic examination detected left ventricular hypertrophy in 44 cases, right ventricular hypertrophy in 11 cases (with additional cardiac anomalies in 8), and biventricular hypertrophy in 6 patients (all had additional cardiac anomalies). ST-T segmental changes were found in 15 patients, left ventricular hypertrophy and ST-T changes in 3 and Wolf-Parkinson-White syndrome in 2 patients. The remaining 9 patients had normal ECG findings. Forty-two patients had additional cardiac anomalies (Table II), such as PDA, valvular aortic stenosis and VSD.

Table I. Age Distribution of Patients with Fixed Subaortic Stenosis

Age	Number	%
Less than 5	11	12.2
6–10	44	49.0
11–15	30	33.3
16–21	4	4.4
21–24	1	1.0

Table II. Additional Cardiac Anomalies in Patients with Fixed Subaortic Stenosis

Type of lesion	Number
PDA	9
VSD	8
Valvular aortic stenosis	8
Coarctation of aorta	5
Congenital mitral stenosis	5
Bicuspid aortic valve	2
Mitral insufficiency	2
Atrioventricular septal defect	2
Pulmonary stenosis	2
TOF	1
Supravalvular aortic stenosis	1
Primum atrial septal defect	1

VSD=ventricular septal defect; PDA=patent ductus arteriosus; TOF=tetralogy of Fallot.

The M-mode evaluation was made during a two-dimensional examination. There was early systolic closure and fluttering of the aortic leaflets in 20 patients and focal septal hypertrophy in 25 patients. The transaortic gradient was measured as 14–130 mmHg with continuous Doppler echocardiography before operation. In 10 unoperated patients, the gradient had increased 12–50 mmHg (mean 34 mmHg), 6 months–3 years after the first Doppler echocardiographic study.

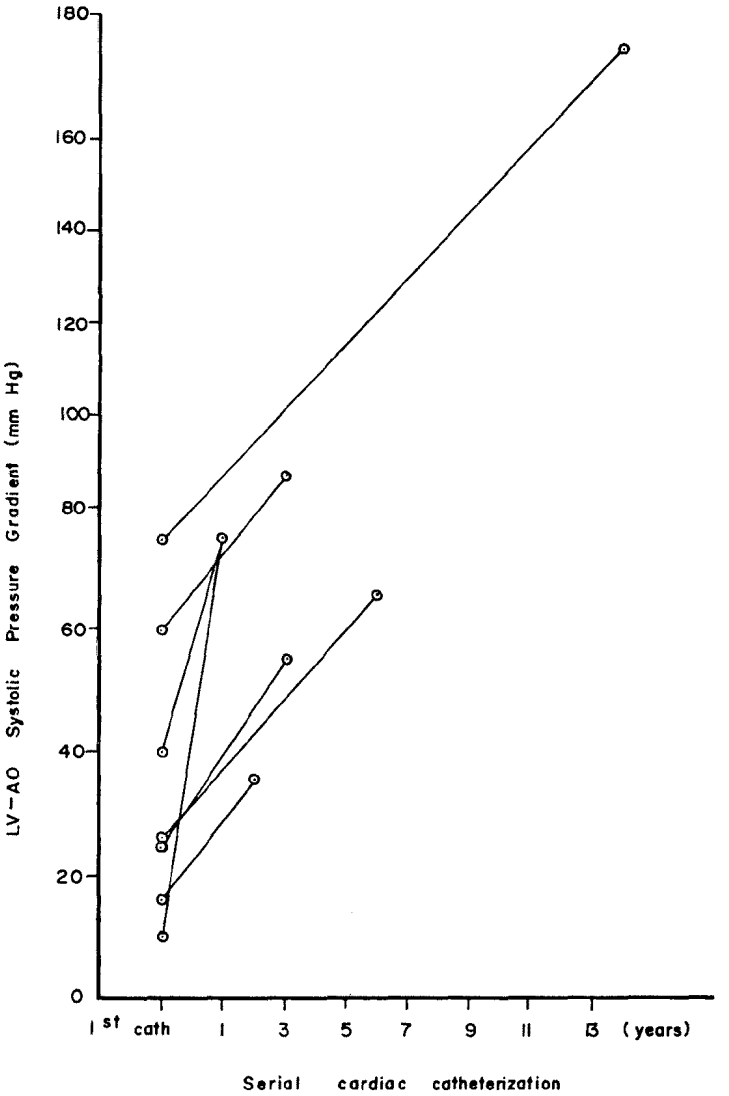

Fig. 1. Increasing left ventricular to aortic pressure gradients documented by serial cardiac catheterization.

Two-dimensional echocardiography was performed on 48 patients, without additional cardiac catheterization. Twenty-five of these patients underwent operations and in 23 cases the diagnosis was confirmed during surgery. Two cases who were evaluated as valvular aortic stenosis by echocardiography were found to have membranous subvalvular aortic stenosis during operations.

Catheter angiography was performed on 42 patients and a 15–190 mmHg gradient was measured between the left ventricular apex and subvalvular area. In 3 patients diagnosed as valvular stenosis by catheter-angiography, a subvalvular membrane resection was made surgically. In 7 patients the gradient increased to 20–100 mmHg (mean 48) during a follow-up period of 1–12 years after the first hemodynamic study (Fig. 1).

The definitive diagnosis was made by two-dimensional echocardiography in 46 patients, by echocardiography and catheterization in 35, by only catheter angiography in 4 and intraoperatively in 5 cases. According to the results of echocardiography, catheter angiography and surgical findings, 60 (66.6%) patients had subvalvular membranous aortic stenosis, 20 (22.2%) patients had a fibromuscular stenosis, 4 (4.4%) displayed both membranous and fibromuscular features and 6 (6.6%) had tunnel type obstructions. Doppler echocardiography and angiography defected aortic insufficiency was detected in 16 patients.

The distribution of the patients according to the surgical methods is shown in Table III. Infective endocarditis was not detected preoperatively. Two patients developed infective endocarditis postoperatively (2.2%).

Table III. Operative Procedures in 50 Patients with Fixed Subaortic Stenosis

Procedure	Number	%
Membrane resection	34	68
Membrane resection and myectomy	8	16
Myotomy and myectomy	7	14
Membrane resection and aortoventriculoplasty	1	2

Procedures in the same operation	Number
Aortic valvotomy	8
VSD closure	5
VSD closure+AVR+MVR	1
MVR	1
Repair aortic coarctation	1
VSD closure and mitral valvotomy	1

AVR=aortic valve replacement; MVR=mitral valve replacement.

Table IV. Preoperative and Postoperative Data in 5 Patients Who Died

Patient	Age (years), Sex	Diagnosis	Preoperative transaortic gradient (mmHg)	Operative procedures	Postoperative exitus time (days)
1	5, M	Bicuspid aortic valve, aortic coarctation, SAS	55	Repair coarctation, aortic valvotomy, membrane resection, myotomy	1
2	7, F	VSD, MS, AI, SAS	85	VSD closure, AVR, MVR, Myectomy	1
3	8, M	Total corrected TOF, residual VSD, SAS	85	VSD closure, membrane resection, myectomy	1
4	11, M	SAS	110	Membrane resection	1
5	11, M	SAS, valvular aortic stenosis, infective endocarditis	110	Membrane resection, myectomy	94

SAS=subvalvular aortic stenosis; MS=mitral stenosis; AI=aortic insufficiency; TOF=tetralogy of Fallot; VSD=ventricular septal defect.

Four patients died during the early postoperative period and one died 3 months after the operation from infective endocarditis (Table IV).

The follow-up period after the operation was between 7 days–11 years (mean 3.6 years). During this period 36 patients were evaluated by two-dimensional and Doppler echocardiography. There were pathological echo findings in 12 cases. According to these studies, 10 patients had focal septal hypertrophy and recurrent narrowing in the subvalvular area, 1 patient had residual membrane, and 1 patient had dynamic obstruction. The mean preoperative transaortic gradient was 81 mmHg (ranged between 47–130 mmHg) in these 12 patients and it was found to be 40 mmHg (ranged between 16–175) postoperatively. The mean age of the patients was 9.3 at the time of operation. Aortic insufficiency was detected in 2 patients with Doppler echocardiography.

Discussion

The terminology for left ventricular outflow tract obstruction (LVOTO) is variable. The syndromes in this study included discrete subaortic stenosis, fibrous subaortic stenosis and fixed subaortic stenosis.[7] Some authors, however, restrict the term discrete to denote the membranous type of stenosis.[1),7] Fixed LVOTO is used in this communication to distinguish these forms of stenosis from the dynamic muscular form of LVOTO.[7]

It has been reported that LVOTO is more common in males than females. Isolated fixed subaortic stenosis appears as a critical lesion after the age of 5 years, most commonly after the age of 10 years.[8),9] In our

study the male to female ratio was 1.8 and the greatest number of the patients were in the 6–10 year old group.

Somerville and co-workers proposed that LVOTO is acquired rather than congenital because obstructive subaortic membranes occur rarely, if ever, in newborns. They suggested that abnormal flow patterns in the left ventricle result in the deposition of fibroelastic material below the aortic valve.[10] However, we detected two siblings with subvalvular membranes and PDA; the incidence of the disorder in the same family has also been previously reported.[3] These findings raise the possibility that congenital and/or genetic factors may contribute to the anomaly.

Associated congenital cardiovascular defects have been reported in 25–55% of patients with fixed subaortic stenosis.[11),12] In our study we detected 45 additional anomalies in 42 patients (46.6%). In 20–25% of patients with fixed subaortic obstruction, the aortic valve was also congenitally abnormal. Furthermore, aortic insufficiency produced turbulant flow and intrinsic involvement of the valve cusps in the fibroelastic membranous process.[8),9] The aortic insufficiency has harmful effects on myocardial ultrastructure and function that may not be reversible after valve replacement. Aortic insufficiency can usually be prevented or stabilized by removing the stenosis.[13] In our study we could show aortic regurgitation in only 16 cases (17.7%). This can be explained by the absence of Doppler echocardiography in our clinic initially, and lack of application of aortography in some hemodynamic trials, since in many cases the murmur of early aortic insufficiency may not be audible clinically. Aortic insufficiency is not related to LVOT gradient.[13]

Infective endocarditis is a serious and frequent complication of subaortic obstruction that is not treated surgically. The incidence ranges from 12–25% of patients in most series.[13] Predisposing factors are aortic insufficiency and a high LVOT gradient. Infective endocarditis developed in only 2 of our patients postoperatively and in none preoperatively. Several series have reported an incidence of postoperative endocarditis ranging from 0–4%.[13] Therefore, subaortic resection probably substantially reduces the risk of infective endocarditis. All patients require lifelong protection against endocarditis before and after surgery because of jet lesions or congenital abnormalities of the aortic valve.[9]

Many authors have reported that cross-sectional and Doppler echocardiography were superior to cardiac catheterization in recognition and characterization of fixed subaortic obstruction.[9),14)–17] Pierli et al demonstrated that two-dimensional echocardiography alone was sufficient to establish a need for surgery.[17] In our study 46 patients were diagnosed by using echo-

cardiography alone. Twenty-five of these patients were treated surgically without preoperative catheterization. The operative results from 23 of these patients supported our previous diagnosis while the other 2 were evaluated as valvular aortic stenosis by echocardiography. Ivert[11] reported the correct diagnosis of subaortic obstruction was not established before surgery in 14% of the cases and that it was not established in 86% until angiography or echocardiography was performed.

Subaortic stenosis is a progressive disease; obstruction increased in almost all reported patients during serial preoperative studies.[1),3),8),9),12),13)] The pressure gradient increased in 7 patients who were recatheterized during the follow-up period and in 10 patients who were reevaluated by Doppler echocardiography. Given the likelihood of both progressive obstruction and aortic insufficiency, the presence of even mild to moderate subaortic stenosis warrants consideration of an elective operation.[3),7)–12),17),18)] Although this idea is generally accepted, few centers consider the presence of high transaortic gradient, left ventricular dysfunction or aortic insufficiency as indications for surgical treatment.[19)] In our study, the older patients with a high gradient were considered for surgical intervention, whereas patients with isolated forms of fixed subaortic stenosis and a transaortic gradient higher than 25 mmHg have only recently been candidates for elective surgery. Thus, the interrelationship between the surgical criteria and recurrent and/or residual obstruction could not be demonstrated clearly in our study.

In a series of 222 surgically treated cases of LVOT obstruction, Ivert et al[11)] reported that the early mortality was 6%, and that only 54% had satisfactory results 6–14 years postoperatively. In our series 4 patients died in the early (8%) and 1 (2%) in the late postoperative period. This high mortality rate is probably due to the surgical interventions made for the other complex cardiac anomalies during membrane resection, a high preoperative gradient and deteriorated left ventricular function and infective endocarditis in 1 patient.

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