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Fixed Subaortic Stenosis in Infants and Children

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= Abstract =**Fixed subaortic stenosis(FSS) is important because of not only obstruction but also aortic insufficiency. This study was performed to find out the characteristics and combined anomalies of FSS. Echocardiographic and clinical data were reviewed in 31 children who were diagnosed as FSS between March 1985 and February 1991 (age: 2months-12yr7months, M:F = 2:1). Cardiac defects were associated in 26(84%) and the majority of the cases were related to VSD(VSD only: 14 cases, VSD with other anomalies: 7 cases). The type of VSD was perimembranous in 86%(18/21) and subarterial in 14%(3/21). Aortic insufficiency(AI) was present in 13 cases(42%) and the degree of AI was usually mild. The mean pressure gradient between the left ventricle and the aorta was 21mmHg by cardiac catheterization in 27. Operative resection was done in 20; 11 during the correction of major defects, 4 for AI, 5 for significant pressure gradient. The pathologic findings of 13 were mainly fibrosis and hypertrophied myocardium. In conclusion, because of the possible association with other cardiac defects, especially perimembranous VSD and its covert features, it is recommended to pay attention to the coexistence of FSS during the evaluation of cardiac defects.**

Key Words: *Fixed subaortic stenosis, Infants, Children*

INTRODUCTION

The discrete form of subaortic stenosis is defined as well limited narrowing of the left ventricular outflow tract which is usually located at or just below the attachment of the aortic valve. Fixed subaortic stenosis(FSS) contributes 8 to 10 percent of all forms of left ventricular outflow obstruction(Friedman *et al.* 1989). This can occur in isolation or in association with other structural cardiac anomalie and it is known as a rare condition in infancy. It is clinically important because not only of obstruction but also aortic insufficiency, which may result from damage to the valve leaflets by the jet flow of blood through the subaortic valvular orifice. Nowadays we encounter many cases of FSS associated with ventricular septal defect and it is easier to diagnose fixed subaortic stenosis after the widespread use of echocardiography. Cross-sectional echocardiography enables early detection of FSS, so the detection rate is increased. Therefore the characteristics of FSS may have changed from before, so we have tried to find out the characteristics and combined anomalies in this study.

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PATIENTS AND METHODS

Echocardiographic and clinical data were reviewed in 31 children who were diagnosed as FSS between March 1985 and February 1991 at Seoul National University Children's Hospital (0.32% of annually diagnosed CHD in SNUCH). The diagnosis was made mainly by echocardiographic evaluation (Fig. 1). The echocardiograms were recorded on VHS video tape by Ultramark 8, ATL or Aloka CW SSD 880 machine. The parasternal long axis or apical two chamber view of cross sectional echocardiography were used. Cardiac catheterization and angiography were performed in 27 patients (Fig. 2). The pressure gradient between the aorta and the left ventricle was checked. During cardiac angiography, we checked the right anterior oblique, lateral or long axial oblique view for the evaluation of the aortic outflow tract. We analized clinical profile, associated anomalies and incidence of aortic insufficiency (AI) in these patients.

RESULTS

There were 21 boys and 10 girls in the entire group. (male/female ratio 2:1). Ages of first detection ranged from 2 months to 12 years and 7 months, and mean age at diagnosis was 4 years (Table 1, Fig. 3). Six of them were diagnosed as having FSS before 1 year of age and among them, four patients had surgical resection of the subaortic stenosis. One was lost during follow-up and the other had persistent FSS after the correction of the main cardiac defect(VSD and ASD), because it was missed during operation (Table 2). Most cases were detected incidentally during the evaluation of other cardiac anomalies and there was no case which was suspected clinically before echocardiography (Table 3).

Cardiac catheterization and angiography were done in 27 patients. The mean left ventricular to aortic peak systolic pressure gradient was 21mmHg and the pressure gradients ranged from zero to 150mmHg.

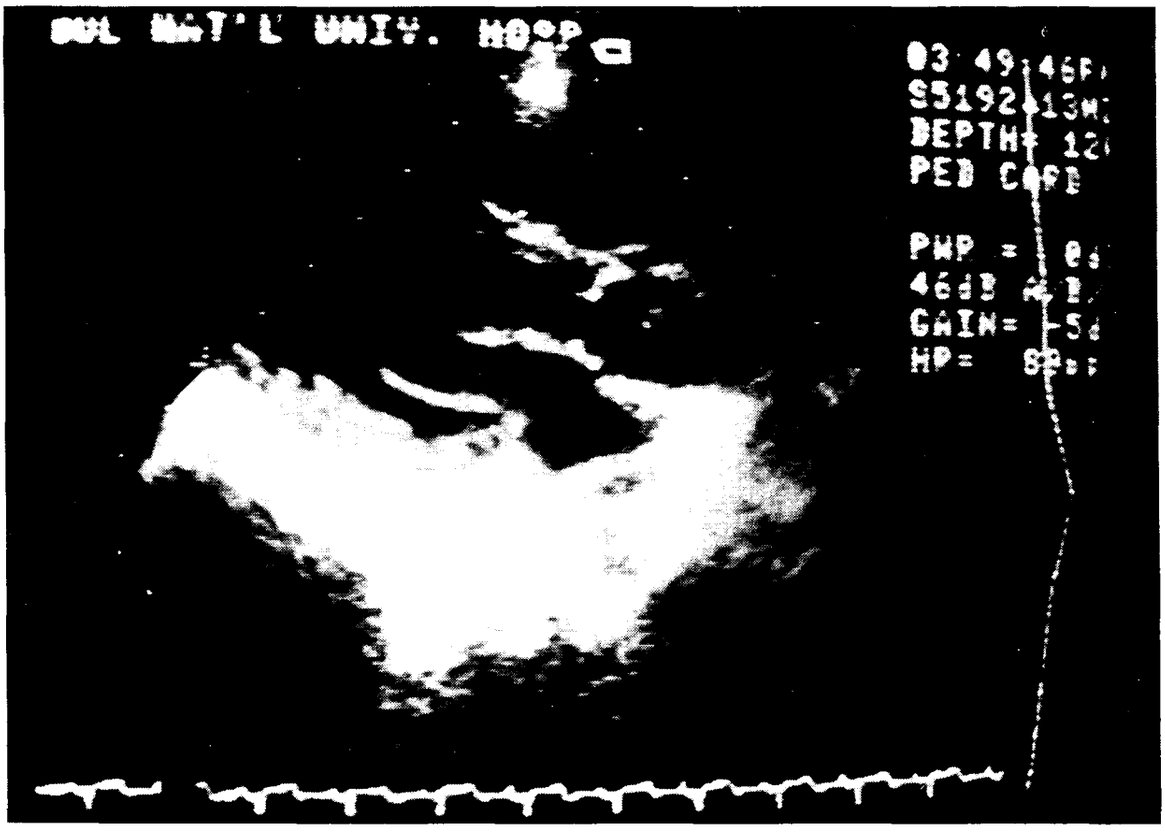

Fig. 1. Echocardiographic findings of a patient who had PDA and subaortic stenosis. This was detected when he was 9 months old.

Fig. 2. Right anterior oblique view of angiography. Subaortic membrane was detected below the aortic valve in the left ventricular outflow tract.

Table 1. Summary of clinical features of 31 cases of FSS

patients	31 cases
Male/Female ratio	2/1
age at diagnosis	4yr (2m-12yr 7m)
mean duration between diagnosis and resection(20 patients)	12 m
mean follow-up duration	28m
mean left ventricular to aortic peak systolic gradient	21mmHg (27 cases)

Twenty-six patients(26/31, 84%) had associated cardiac defects(Table 4). Ventricular septal defect was found in 21 patients(21/26, 81%). Isolated ventricular septal defect was identified in 14 patients(14/26, 54%) and the rest of them had ventricular septal defect

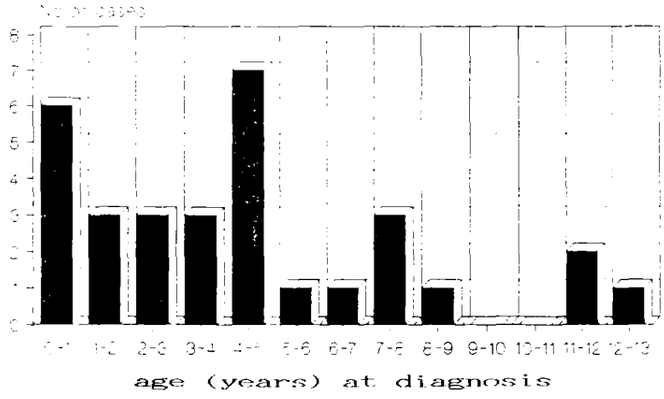

Fig. 3. Age distribution.

Table 2. Six cases detected before 1 year of age

Cases	Age	Combined anomaly	Operation
1	2m	VSD + ASD	No
2	3m	VSD	No
3	6m	VSD	Yes
4	6m	CoA + PDA	Yes
5	9m	PDA	Yes
6	10m	VSD + PDA	Yes

Table 3. Mode of detection

Mode	No. of patients
incidental (during the evaluation of other cardiac defects)	26(84%)
heart murmur	4(13%)
dyspnea on exertion	1(3%)

associated cardiac anomalies. . We found 2 cases of ventricular septal defect associated with coarctaton of the aorta and patent ductus arteriosus. In 5 cases, ventricular septal defect was not combined and more information is listed on Table 4.

In the majority of the patients(18/21, 86%), perimembranous type VSD was identified. Among them, 4 patients had an associated aneurysm of the membranous septum by echocardiographic evaluation. The remainder (3/18,14%) had an infundibular type (subarterial) of VSD (Fig. 4).

Surgical resection of the discrete subaortic membrane was performed in 20 patients. The

Table 4. Associated cardiac defects

Associated defects	No. of cases
None	5(16%)
VSD	21(68%)
VSD only	12(39%)
VSD + ASD	2(6%)
VSD + PDA	3(10%)
VSD + CoA + PDA	2(6%)
VSD + DCRV	2(6%)
others	5(16%)
CoA + PDA	2(6%)
PDA	3(10%)

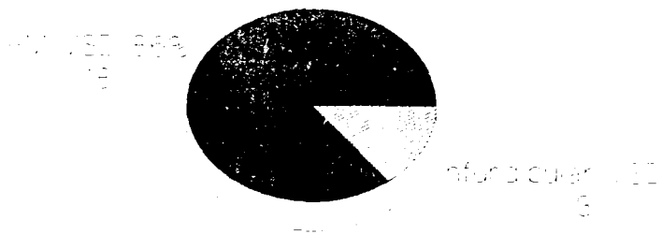

Fig. 4. Type of VSD.

mean duration between diagnosis and resection in 20 patients who received the operation, was 12.9 months. The mean follow-up duration was 28 months. Eleven patients received resection of fixed subaortic membrane during correction of the main cardiac defects. In 4 patients, the operation was done because of a progressive aortic insufficiency and the resection was performed because of significant pressure gradients(mean:64mmHg) in 5 patients. In 2 patients, modified Konno operation (aortoventriculoplasty) was done and one of them showed tunnel like myocardial hypertrophy during operation. Operative indications are summarised in (Fig. 5).

FSS was also detected after the correction of main congenital heart disease in 8 patients. Among them, 4 patients needed reoperation due to the progressive nature of the obstruction.

Aortic insufficiency was detected in 13 cases(42%). Among 10 patients who received operations, aortic insufficiency disappeared after operation in 8 patients (Fig. 6).

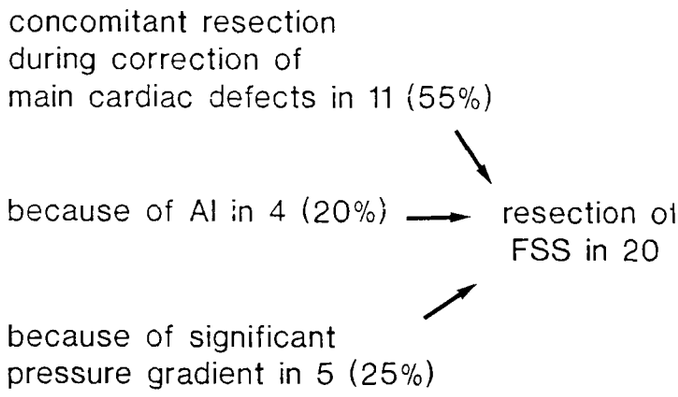

Fig. 5. Operative indication.

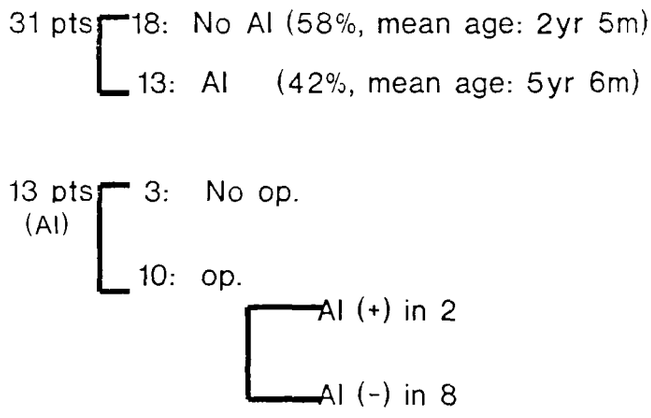

Fig. 6. Aortic insufficiency.

Histologic examination showed thickened endocardium with fibrosis and myxoid degeneration in 13 cases. The myocardium showed myohypertrophy and variation in size of myofiber with interstitial fibrosis (Fig. 7).

DISCUSSION

Fixed subaortic stenosis(FSS) is an uncommon congenital cardiac anomaly, which was first described by Chevers in 1842(Kelly *et al.* 1972). The cause of this anomaly has been explained traditionally as incomplete atrophy of the bulbus cordis(Keith 1924), but Van Praagh *et al.* (1970) has stated that fibrous subaortic stenosis results from maldevelopment of the endocardial cushion tissue of the atrioventricular canal that usually forms the anterior leaflet of the mitral valve. Nowadays the causes of FSS either in isolation or in association with ventricular septal defect, are known to be heterogenous(Vogel *et al.* 1983).

In our study, associated cardiac anomalies

were present in 84% of the cases studied. According to a report of Neufeld *et al.* (1987) 57% of patients with FSS had cardiac defects, which often masked the typical clinical and catheterization features of subaortic stenosis and Vogel *et al.* (1983) also reported associated cardiac defects in 47% of the cases studied. Compared to other reports, our results show that more cases were related to associated cardiac defect and for an explanation for this, we must consider that most FSS in our study was detected during the evaluation of other cardiac defects. Twenty-one out of 26 cases(81%) were combined with VSD. The type of ventricular septal defect was analysed and the majority of cases had perimembranous (type 2) VSD (81% of VSD). In other study(Vogel *et al.* 1983), they also reported that perimembranous ventricular septal defect was most frequently related to FSS. Silent FSS could be unmasked by the closure of the ventricular septal defect and could reduce the flow through the ventricular septal defect. Thus, the size of the ventricular septal defect tends to be underestimated(Vogel *et al.* 1983). In all patients this lesion developed sometime after spontaneous closure or reduction in the size of the ventricular septal defect, suggesting that the subaortic membrane is closely related to the membranous ventricular septum. The subaortic membrane may be derived from endocardial proliferation with fibrosis into the left ventricular outflow tract stimulated by turbulent blood flow through the defect (Pierli *et al.* 1989; Waldman *et al.* 1984). FSS is associated with ventricular septal defect especially with aneurysmal formation of the membranous ventricular septum. We found 4 cases which had infundibular aneurysms.

It's progressive nature after spontaneous closure of the ventricular septal defect warrants periodic cardiac evaluation if the patient continues to have a significant heart murmur or abnormal electrocardiographic results(Pierli *et al* 1989). A discrete subaortic shelf situated adjacent to a ventricular septal defect may be "silent" producing minimal, if any, pressure gradient and may pose diagnostic difficulties. Failure to recognize such a shelf and to remove

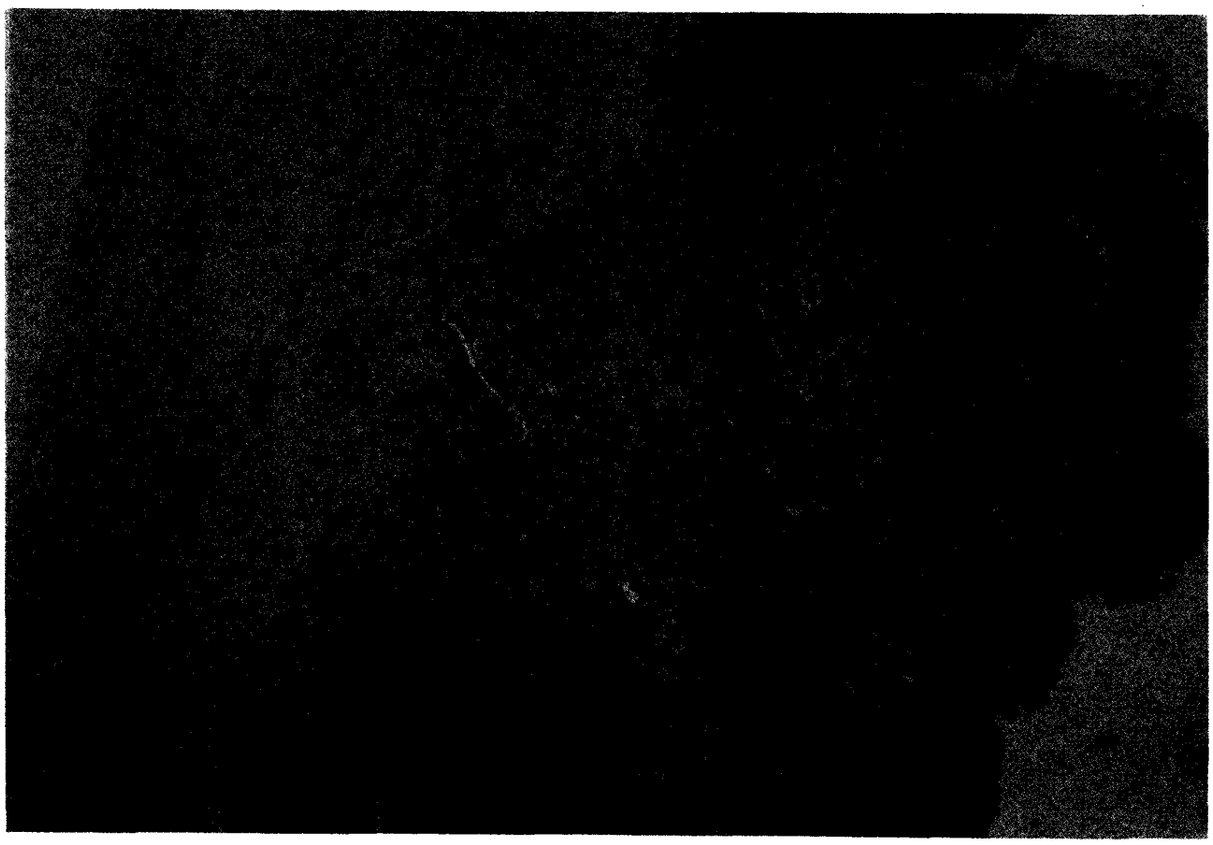

Fig. 7. Pathologic findings of FSS. The endocardium was thickened with fibrotic change and myxoid degeneration. The myocardium showed hypertrophy.

it at the time of surgical closure of the ventricular septal defect, may result in the creation of a severe subaortic obstruction(Waldman *et al* 1984). In our study most cases were detected incidentally during the evaluation of other cardiac anomalies and there was no case which was suspected clinically before echocardiography. There are several possible explanations for these findings of obscured left ventricular outflow tract obstructions. One is that the subaortic stenosis was initially minimal but became progressively more severe over several years. Another possibility is that a large ventricular septal defect proximal to the subaortic stenosis can, by decompressing the left ventricle, minimize the left ventricular outflow tract pressure gradient until the ventricular septal defect is closed. Only after the defect is closed the pressure gradient will be fully revealed(Neufeld *et al.* 1976).

Although one cannot exclude the possibility that subaortic stenosis is acquired after surgical closure of the ventricular septal defect, preoperative identification of associated subaortic stenosis suggests that this anomaly is one of congenital lesions(Fisher *et al.* 1982). It is possible that the subaortic membrane is present before the first operation and that it becomes manifest when the interventricular communication is closed at that operation (Shachar *et al.* 1981). There is much controversy nowadays, whether FSS is a congenital or acquired lesion. Although our data is not conclusive the fact that 19% of patients were infant and most of them were below the age of five, can be supportive evidence of congenital lesion. There are several possible explanations for the apparent age-related disparity in the incidence of FSS. One possibility is that FSS leads to early death. Another possible explanation for the age-related disparity in the incidence of FSS is that this lesion itself changes with age, a conceivable, but certainly a rare and fortuitous loss of obstruction might occur secondary to erosion of the obstructing membrane itself by endocarditis(Katz *et al.* 1977).

Complete resection of mild FSS is likely to leave a patient with an anatomically normal aortic valve and minimal prospects for significant recurrent obstruction. Therefore, we suggest that resection of FSS is indicated if a patient requires intracardiac repair of an associated lesion and for gradients above 30mmHg in children below 12 years-old(Wright *et al.* 1983). In this study, among 20 cases which underwent the operation, five patients received resection because of a significant pressure gradient above 30 mmHg.

Our data suggests that aortic insufficiency is a frequent complication in FSS. Aortic insufficiency in FSS has been reported in approximately 50% of other cases(Deutsch *et al.* 1971). We detected AI in 13 cases and after operative resection, AI disappeared in 8 cases. The reason why AI disappeared after operation in a significant number of cases is suggested in two ways. A relatively early operation(mean duration: 28 month), and a mild degree of AI were the factors. According to our data, the mean age of the cases which had AI was older than the cases without AI. So, it is suspected that AI might be progressive or newly developed as patients become older. The progressive nature of this malformation suggests an early surgical treatment in the presence of initial aortic regurgitation, even in patients with low pressure gradients is desirable. This approach by avoiding the progressive damage to the aortic valve due to the blood flow turbulence, significantly reduces the risk of reoperation of aortic valve either through reconstruction or replacement in the late follow-up(Pierli *et al.* 1989).

FSS can be demonstrated by apical long axis view or left parasternal long axis view of 2-D echocardiography. In several of echocardiographic illustrations only the anterior aspect of the membrane is demonstrated. 2D echo is as effective as angiography in demonstrating FSS. Although only a portion of the lesion is found, the experienced examiner should have no difficulties in finding FSS.

Neufeld *et al.* (1987) stated that, because of the likelihood of progression of the left

ventricula outflow tract obstruction in FSS, it is unreasonable to use the same pressure gradient criteria for operation as those used in valvular aortic stenosis(Motro *et al.* 1984). Results of surgical treatment of FSS indicate that most patients show improvement after localized resection and were long term survivors. As follow-up duration increases however, there is evidence that recurrent stenosis becomes a real threat to survivors. Late mortality after a successful initial operation is not associated with progressive aortic regurgitation or endocarditis, but is related to the need for and performance of reoperation. For the repair of tunnel like narrowing of the left ventricular outflow tract, the aortoventriculoplasty method is used nowadays, which is the widening of the aortic valve as well as the subaortic outlfow tract (Konno operation) (Berry *et al.* 1979).

In conclusion Mean age of thirty-one children with FSS (21 boys, 10 girls) at diagnosis was 4 years (2 months - 12 years and 7 months). Cardiac defects were associated in 26 cases (84%). The majority of the cases were related to VSD (VSD only in 14 cases, VSD with other anomalies in 7 cases) and others were associated with PDA, CoA. Perimembranous VSD is more commonly associated (84%) than is infundibular type. The mean LV to aortic systolic pressure gradient was 21 mmHg. AI was detected in 13 cases (42%). Ten of them received operative resection and AI disapperaed in 8 cases. Most of the cases had covert features and underwent the operation during the repair of combined anomalies. FSS was detected after the correction of main congenital heart disease in 8 cases. Among them, four patients needed reoperation due to progression of the obstruction.

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