Original Article

Subaortic Stenosis Associated with Perimembranous Ventricular Septal Defect. Clinical Follow-Up of 36 Patients

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Objective

To study the clinical pattern of subaortic stenosis associated with perimembranous ventricular septal defect.

Methods

From January 1979 to June 2000, 36 children with perimembranous ventricular septal defect and fixed subaortic stenosis were followed-up regarding anatomic characteristics, evolvement, and clinical events.

Results

Age at diagnosis of subaortic stenosis ranged from 6 months to 170 months, and it was less than 1 year in only 2 children. Regarding sex, the distribution was 2:1 with a greater predominance of males. Ventricular septal defect was small in 61.0% of cases, medium in 30.56%, and large in 8.40%; the size of the septal defect decreased during follow-up in 30.56% (11 cases). In all patients, subaortic stenosis was membranous and fixed. During follow-up, 23 patients experienced evolvement of the stenosis. Surgical treatment was performed in 21 cases, and one patient underwent surgery for restenosis. Infectious endocarditis occurred in 2 patients; one of the patients died.

Conclusion

Subaortic stenosis occurs in the natural history of ventricular septal defect usually after the first year of life, and it is progressive and requires surgery in most cases.

Key words

ventricular septal defect, subaortic stenosis, congenital heart disease, endocarditis

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Subaortic stenosis may occur as a complication of the natural evolvement of several congenital heart diseases; however, its etiology is still unclear[1,2]. Ventricular septal defect may develop into subaortic stenosis in 20% of cases[2-4]. The subaortic stenosis developed in these cases is frequently membranous and fixed, and it may occur when the diameter of the ventricular septal defect decreases, after spontaneous closure[5-7], or after surgical correction[8,9].

In this study, we assessed the clinical evolvement, the echocardiographic, cineangiographic, and surgical characteristics of children with perimembranous ventricular septal defect who developed subaortic stenosis during follow-up.

Methods

From January 1979 to June 2000, 36 children with perimembranous ventricular septal defect who developed fixed subaortic stenosis were followed-up. Age ranged from 6 to 170 months; 12 children were females, and 24 were males. Children with a diagnosis of subaortic stenosis were included in the study.

Cases of subaortic stenosis associated with other types of septal defect and other heart diseases that might interfere with the clinical evolvement and the indication of surgical correction were excluded.

Data about clinical evolvement of this group were retrospectively collected. With the necessary revisions, the following data from bidimensional Doppler echocardiograms and/or surgery and/or cineangiography were assessed: identification of the anatomical type[10] and size[11] of the ventricular septal defect, anatomic characteristics of the fixed subaortic stenosis[12], evolvement assessment of the left ventricular outflow pressure gradient; symptomatology; events occurring during clinical evolvement: infectious endocarditis, surgical treatment, duration of follow-up, and death.

Doppler echocardiography was recorded during previous examinations, by using bidimensional examinations, pulsed Doppler, and continuous wave Doppler. Maximum and medium transvalvular aortic gradient were determined using Bernoulli's simplified equation.

Statistical analysis was performed using ANOVA - One Way - tables with comparison between the average demonstrated in a numerical scale, the chi-square test or Fisher's exact test (when indicated), Fisher's exact test to determine the level of statistical significance of the differences observed, using the Kruskal-Wallis test whenever Bartlett's test was significant. The level of statistical significance adopted was 5%.

Results

Table I presents the distribution of ages at diagnosis and subaortic stenosis surgery.

Diagnosis of subaortic stenosis by echocardiogram was performed in 29 (80.6%) children and by cineangiography in 7 (19.4%). Males were predominant over females, accounting for 66.67% (24) and 33.33% [12] of the cases, respectively. In 5 patients (13.90%), the diagnosis of subaortic stenosis was simultaneous with that of ventricular septal defect, with all 5 patients over 2 years old. None of the diagnoses of subaortic stenosis occurred after surgical closure of the septal defect, and no reports of familial incidence occurred in the cases studied.

Table II describes the sizes of ventricular septal defects in the sample.

We observed a trend toward development of subaortic stenosis following the decrease in size of the ventricular septal defect in 11 (30.56%) children. Figure 1 demonstrates serial echocardiograms of the same children, with the appearance of subaortic stenosis by 6 months.

The pattern of the ventricular septal defect during follow-up is described in table III.

We identified other associated heart diseases in 6 patients (16.67%), described in table IV.

Table V demonstrates the subaortic gradient at diagnosis and surgery in 23 children (63.80%). We have observed a progressive increase in the left ventricular outflow pressure gradient during follow-up (mean, 20.90 ±6.81 mmHg), whereas in 5 children (13.89%) significant progression of the gradient (>5 mmHg) did not occur in the follow-up period of 36 and 127 months (mean, 81.60 ± 45.60). In 13 patients (36.11%), significant progression of the gradient occurred during follow-up, and the patients underwent surgery.

None of the patients had symptoms related to the development of subaortic stenosis.

The characteristics of evolvement and treatment are found in table VI.

In 15 cases (41.67%), children were followed-up only through clinical treatment for a period of 12 to 130 months (mean, 58.38 ± 40.17). Surgical treatment was performed in 21 children (58.33%), and surgery was indicated when the subaortic gradient was higher than 40 mmHg except for a patient with endocarditis who had a 35 mmHg gradient and underwent. In 8 children (22.22%), the surgery was indicated immediately after the diagnosis because they had a subaortic gradient above 40 mmHg. The 21 children (58.33%) undergoing surgery were followed-up for a mean time of 23.41 ± 34.34 months, with one case of reoperation for subaortic stenosis recurrence during that period.

Two patients (5.56%) had infectious endocarditis at 48 and 60 months of age, respectively, and underwent surgery. One of the patients evolved to death in the immediate postoperative period.

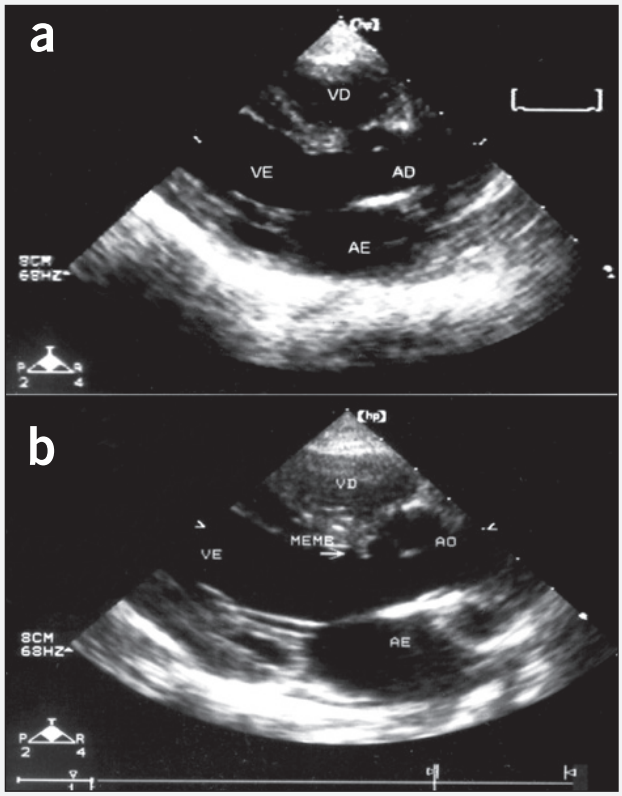

Fig. 1 - Echocardiographic images of left ventricular outflow tract of a child with a perimembranous septal defect, before (a) and after (b) the diagnosis of fixed subaortic stenosis. Difference in age of the child between the 1st and 2nd echocardiogram was 6 months (b).

Table I - Age at diagnosis and surgery of fixed subaortic stenosis with ventricular septal defect

Variable	Parameters
Age at diagnosis (months) Mean ± SD (minimum - maximum) n =	55.58±42.16(6-170) 36
Age at surgery (months) Mean ± SD (minimum - maximum) n =	81.40±46.53(14-170) 21

n - number of cases; SD - Standard deviation.

Table II - Size of the ventricular septal defect in 36 children with ventricular septal defect and fixed subaortic stenosis

Characteristic studied Size of VSD	% (number of cases)
small	61.11 (22)
medium	30.56 (11)
large	8.33 (3)
Total	100 (36)

Table III - Behavior of ventricular septal defect in 36 children with ventricular septal defect and fixed subaortic stenosis

Behavior	% (number of cases)
Spontaneous closure	2.78 (1)
Decreasing in size	30.56 (11)
Operated	58.33 (21)
Unaltered	8.33 (3)
Total	100 (36)

Table IV - Associated heart diseases in 36 children with ventricular septal defect and fixed subaortic stenosis

Heart disease	% (number of cases)
Aortic failure	8.33 (3)
Right ventricle abnormal band	2.78 (1)
Bicuspid aortic valve	2.78 (1)
Aortic coarctation	2.78 (1)
Total	16.67 (6)

Table V - Fixed subaortic stenosis gradient at diagnosis and surgery in 36 children with ventricular septal defect

Variable	Parameters
Gradient at diagnoses (mmHg)	38.86 ± 22.50 (10-98)
Mean ± SD (minimum – maximum) n =	36
Gradient at surgery (mmHg)	60.31 ± 15.13 (35-98)
Mean ± SD (minimum – maximum) n =	21

n - number of cases; SD - Standard deviation.

Table VI - Events in the evolvement of ventricular septal defect and fixed subaortic stenosis in 36 children

Event	% (n)
Clinical follow-up	41.67 (15)
Surgery	58.33 (21)
Reoperation	2.78 (1)
Bacterial endocarditis	5.56 (2)
Death	2.78 (1)

n - number of cases.

Discussion

Diagnosis of subaortic stenosis occurred after the first year of life in 94.40% of cases, with a predominance of males in a 2:1 ratio, in agreement with other observations [13-18].

We have not identified a family history in the anamneses in our study group. Petsas and cols [19]. reported 4 cases of different anatomic types of subaortic stenosis in one family.

Seven children underwent cardiac catheterization before the use of echocardiography.

The first reports on subaortic stenosis in patients with ventricular septal defect and left ventricular pressure outflow previously reported as normal through cineangiography occurred in the 70s and the 80s. Some authors attributed these findings to the inaccuracy of the method stating that cineangiography–the procedure routinely performed at that time–was not suitable for the diagnosis of subaortic stenosis [20].

The advent of echocardiography, a safe and noninvasive method of follow-up of patients with ventricular septal defect, enableld us to understand that the obstruction may not be present in the first year of life in children, but arises generally when the ventricular septal defect shows signs of a decrease in size and spontaneous closure [21-23]. We have observed its appearance before one year of age in only 2 patients.

We have clearly identified the tendency of subaortic stenosis to occur especially in perimembranous ventricular septal defects with favorable characteristics for spontaneous closure, because most of them were small and had formation of subtricuspid tissue in their borders. The mechanism of spontaneous closure of the ventricular septal defect was regarded by some authors as responsible for the formation of the obstruction in the left ventricular outflow tract [5,7].

Subaortic stenosis did not cause symptoms, even in those patients with up to 90 mmHg of left ventricular outflow tract gradient, and its appearance did not lead to electrocardiographic or laboratory alterations, therefore being an occasional finding in control echocardiograms as in other studies [24-26], although some authors report symptoms in adulthood.

Eight children presented at the first examination with high gradients in left ventricular outflow tract. These late diagnoses may be explained through the absence of symptoms and the lower social level of our patients.

Twenty-three children (63.90%) had subaortic stenosis progression with a mild increase in the gradient over the years, whereas others evolved in a few months to surgical treatment. This finding confirms the evolving characteristic of the disease [27,28], and demonstrates the need for prolonged follow-up of these patients[29,30].

Usually, surgery was indicated when the subaortic gradient was > 40 mmHg. Some physicians indicate surgery immediately after diagnosis, regardless of the gradient, because it is not a benign progressive disease [31,32]. Other physicians indicate it with gradients between 20 to 80 mmHg [33,34]. It is safer to indicate surgery with higher gradients, because many children observed for years did not experience progression of the subaortic stenosis[35,36].

It is interesting to observe that, in our series, surgical indication occurred due to the development of stenosis rather than to clinical repercussions from ventricular septal defect, which was small in most cases, therefore explaining the absence of a correlation between size of ventricular septal defect and surgical indication. We agree that the need for surgery is determined by the subaortic gradient rather than by the size of the ventricular septal defect in most cases [37,38], in several studied series.

Restenosis occurred in one child who was operated on, in the 80s, when surface resection of the stenotic membrane was common. Later, prevention of restenosis was performed through myectomy deeper in the left ventricular outflow tract [39,40].

Two of 36 children (5.50%) had infectious endocarditis, and one child died. The incidence of endocarditis, a frequent complication in this disease, has decreased [41,42]. The low social level of our sample, with poor access to dental treatment, may explain its recent occurrence in one of our children.

In conclusion, we have observed in our sample that the diagnosis of fixed subaortic stenosis occurred in the majority of cases after the first year of life, with a predominance of males in a ratio of 2:1. All patients were asymptomatic, with the diagnosis made by echocardiographic follow-up. In the majority of cases, subaortic stenosis with ventricular septal defect developed together with a small perimembranous septal defect. A progression in the subaortic gradient in 63.89% of cases during clinical follow-up, and this gradient, rather than the size of the ventricular septal defect, was the main determinant of surgical indication. Infectious endocarditis occurred in 2 children, leading to death in one.

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