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, evolution, 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 evolution 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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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 evolution 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.

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

<table>
<thead>
<tr>
<th>Variable</th>
<th>Parameters</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at diagnosis (months)</td>
<td>55.58±42.16(6-170)</td>
</tr>
<tr>
<td>Mean ± SD (minimum - maximum) n</td>
<td>36</td>
</tr>
<tr>
<td>Age at surgery (months)</td>
<td>81.40±46.53(14-170)</td>
</tr>
<tr>
<td>Mean ± SD (minimum - maximum) n</td>
<td>21</td>
</tr>
<tr>
<td>n - number of cases; SD - Standard deviation.</td>
<td></td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Characteristic studied</th>
<th>% (number of cases)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Size of VSD</td>
<td></td>
</tr>
<tr>
<td>small</td>
<td>61.11 (22)</td>
</tr>
<tr>
<td>medium</td>
<td>30.56 (11)</td>
</tr>
<tr>
<td>large</td>
<td>8.33 (3)</td>
</tr>
<tr>
<td>Total</td>
<td>100 (36)</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Behavior</th>
<th>% (number of cases)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spontaneous closure</td>
<td>2.78 (1)</td>
</tr>
<tr>
<td>Decreasing in size</td>
<td>30.56 (11)</td>
</tr>
<tr>
<td>Operated</td>
<td>58.33 (21)</td>
</tr>
<tr>
<td>Unaltered</td>
<td>8.33 (3)</td>
</tr>
<tr>
<td>Total</td>
<td>100 (36)</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Heart disease</th>
<th>% (number of cases)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic failure</td>
<td>8.33 (3)</td>
</tr>
<tr>
<td>Right ventricle abnormal band</td>
<td>2.78 (3)</td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>2.78 (3)</td>
</tr>
<tr>
<td>Aortic coarctation</td>
<td>2.78 (3)</td>
</tr>
<tr>
<td>Total</td>
<td>16.67 (6)</td>
</tr>
</tbody>
</table>
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.

<table>
<thead>
<tr>
<th>Event</th>
<th>% (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical follow-up</td>
<td>41.67 (15)</td>
</tr>
<tr>
<td>Surgery</td>
<td>58.33 (21)</td>
</tr>
<tr>
<td>Reoperation</td>
<td>2.78 (1)</td>
</tr>
<tr>
<td>Bacterial endocarditis</td>
<td>5.56 (2)</td>
</tr>
<tr>
<td>Death</td>
<td>2.78 (1)</td>
</tr>
</tbody>
</table>

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, enabled 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 24-26.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Parameters</th>
</tr>
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<tbody>
<tr>
<td>Gradient at diagnosis (mmHg)</td>
<td>38.96 ± 22.50 (10-98)</td>
</tr>
<tr>
<td>Mean ± SD (minimum – maximum)</td>
<td>n = 36</td>
</tr>
<tr>
<td>Gradient at surgery (mmHg)</td>
<td>60.31 ± 15.13 (35-98)</td>
</tr>
<tr>
<td>Mean ± SD (minimum – maximum)</td>
<td>n = 21</td>
</tr>
</tbody>
</table>

n - number of cases; SD - Standard deviation.

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

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