Subaortic fibrous ridge and ventricular septal defect: role of septal malalignment

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ABSTRACT The purpose of this study was to test the hypothesis that the presence of a subaortic ridge associated with a ventricular septal defect (VSD) is related to a malaligned ventricular septum caused by anterior or posterior deviation of the infundibular septum with or without obstructive lesions of the aortic arch. Thirty-two of 295 patients in whom a diagnosis of VSD was made by two-dimensional echocardiography and who were studied from June 1983 to April 1985 presented with a subaortic shelf. Every patient (p < .00001) had a malalignment type of defect; the defect was produced by anterior deviation of the outlet septum (without compromise of the right ventricular outflow tract) in 28 and by posterior deviation of the infundibular septum in four. The prevalence of a subaortic shelf in the malalignment VSD group was 82% (32/39). Among the 28 patients with a subaortic ridge and anterior deviation of the outlet septum only three had aortic coarctation, but all four patients with subaortic stenosis and posterior infundibular malalignment had obstructive lesions of the aortic arch — coarctation in three and interruption of the aortic arch in one (p < .001). We conclude that a malalignment type of VSD may be a consistent feature in patients with VSD and associated discrete subaortic stenosis. We also noted a high prevalence of subaortic ridge in the presence of a malalignment VSD and therefore speculate that there may be a common morphogenesis for malalignment VSD, subaortic shelf, and obstructive lesions of the aortic arch.

Circulation 75, No. 6, 1124-1129, 1987.

THE EASY recognition and characterization of a “discrete” or fixed subaortic ridge by two-dimensional echocardiography now provides a method for the prospective study of the coexistence of ventricular septal defects (VSDs) and subaortic shelf.1-10

In the presence of a VSD, a subaortic stenosis may be clinically silent. Also, a subaortic shelf may not cause any gradient between the left ventricle and the aorta, but the obstruction can progress if the VSD is surgically or spontaneously closed.11-13

The association of subaortic stenosis with VSDs has often been reported in the setting of aortic coarctation or interruption.14 Furthermore, the morphologic type of VSD that is virtually always present in association with aortic coarctation or interruption is that of malalignment between the trabecular and infundibular septum (without right ventricular outflow tract obstruction), with aortic overriding, or as a result of posterior deviation of the outlet septum.15,16 On the other hand, a malalignment VSD can also occur in the absence of coarctation or interruption of the aortic arch.

This study was carried out to test the hypothesis that the presence of a subaortic ridge associated with a VSD is related to a malaligned ventricular septum caused by anterior or posterior deviation of the infundibular septum with or without obstructive lesions of the aortic arch.

Methods

Between June 1983 and April 1985, two-dimensional echocardiographic diagnosis of VSD was done prospectively in 295 patients. Thirty-two patients in which a subaortic shelf was detected formed the study group (table 1). Fifteen were boys and 17 girls, aged 1 month to 14 years (mean: 3 ± 12 years ± 3 8/12 years, median: 1 9/12 years).

Two-dimensional echocardiographic examinations were performed with ATL MK-300 LX or ALOKA SSD-720 equipment, with 3, 3.5, or 5 MHz transducers. The studies were recorded on videotape and were independently reviewed by at least two of the authors. The sequential analysis principle was used, basing the echocardiographic approach on the determination of the situs, the cardiac and aortic arch positions, the type and mode of atrioventricular connection, the ventriculoarterial connections, and associated defects. Standard subcostal, para-sternal, apical, and suprasternal views were used and, when necessary, other views were searched as well. The VSD was morphologically characterized as perimembranous, muscular, or subarterial according to the components of its borders and the

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Received Nov. 7, 1985; revision accepted Feb. 12, 1987.
extension to trabecular, inlet, or outlet portions of the ventricular septum. The presence of septal malalignment was established when the outlet septum was deviated anteriorly or posteriorly (figures 1 to 3). Care was taken not to produce a false echocardiographic appearance of aortic overriding as a result of a too cephalad position of the transducer. The subaortic shelf or ridge was recognized when an echo-bright localized protrusion into the left ventricular outflow tract extending from the superior or part of the ventricular septum was seen below or above the VSD that formed a fibrous or fibromuscular crescent, with or without an accompanying "mitral" insertion (figure 2). In 13 cases cardiac catheterization and angiography were performed during the study, allowing comparison of data. Six patients were operated on in this period, providing the possibility of surgical correlation. An autopsy specimen from a nonoperated child who died of sepsis was studied.

Data were analyzed by the chi-square test and Fischer's exact test, with a confidence limit of 99.9% ($\alpha = 0.001$).

Results

The analysis of the relationship between the presence of septal malalignment and the echocardiographic finding of a subaortic fibrous ridge in the setting of VSD showed that of 295 patients with VSD, 32 had a fixed subaortic ridge. All of them were in the group with malalignment VSDs ($X^2 = 235.63$, $p < .00001$). Furthermore, in the present series no patient with VSD and a normal alignment between the trabecular and infundibular septum had a subaortic shelf (table 1).

The majority of defects were perimembranous, with only one case of a doubly committed subarterial defect. The septal malalignment was produced by anterior or deviation of the infundibular septum (without compromise of the right ventricular outflow tract) in 28 of the 32 patients (only three of which had aortic coarctation) and by posterior deviation of the outlet septum in four, all with obstructive lesions of the aortic arch (coarctation in three and interruption in one) (table 2, $p < .0001$).

There was a spontaneous decrease in the diameter of the VSD caused by apposition of accessory tricuspid valve tissue over its margins in two-thirds of the patients with fixed subaortic stenosis; the majority of defects were smaller than the aortic anulus. An aortic ejection systolic murmur was detected in five patients (15.6%), with no other clinical signs or symptoms of subaortic obstruction.

Of 13 patients who also prospectively underwent cardiac catheterization and angiography during the study, five had a definite subaortic shelf (figure 4), three of them with a detectable pressure gradient between the left ventricle and the aorta. One angiogram was dubious and seven failed to unequivocally demonstrate a subaortic ridge. In six patients the presence of the subaortic fibrous crescent was surgically confirmed; the shelf was excised during VSD closure in five. However, in four of these patients the angiogram had been considered "negative" for the existence of a subaortic ridge. In the one autopsy specimen examined, the finding of a fibrous subaortic shelf below the margins of a perimembranous VSD with anterior deviation of the infundibular septum and aortic over-

TABLE 1

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<tr>
<th>Subaortic shelf and VSD in patients with septal malalignment</th>
<th>Malalignment VSD</th>
<th>VSD without malalignment</th>
<th>Total group of VSD</th>
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<tbody>
<tr>
<td>Subaortic shelf</td>
<td>n</td>
<td>%</td>
<td>n</td>
</tr>
<tr>
<td>Present</td>
<td>32</td>
<td>82.0</td>
<td>0</td>
</tr>
<tr>
<td>Absent</td>
<td>7</td>
<td>18.0</td>
<td>256</td>
</tr>
<tr>
<td>Total</td>
<td>39</td>
<td>100.0</td>
<td>256</td>
</tr>
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$X^2 = 235.63$ ($p < .00001$).
riding over the trabecular septum confirmed the echocardiographic findings.

Discussion

The most striking result obtained in this study not previously reported by other authors was the constant presence of septal malalignment (100%, p < .00001) in association with a VSD and a subaortic fibrous ridge.

Even though it is possible that VSD and fixed subaortic stenosis may coexist without septal malalignment, this was not the case in our entire population of 295 patients with VSD. We therefore believe that this feature is not a random finding and that a real correlation exists. It was also shown that a subaortic shelf or crescent was prevalent in our population with malalignment VSD (32 of 39 patients, 82%).

Two-dimensional echocardiography is the best method for the diagnosis of a subaortic ridge in the setting of VSD\textsuperscript{12, 13, 23, 24} and for the detection of septal malalignment.\textsuperscript{16, 25} Thus, this association must be carefully and systematically searched during routine two-dimensional echocardiographic examinations.

It is still not clear whether this subaortic fibrous shelf is congenital or acquired in origin. It is quite possible that the increased turbulence adjacent to the area of the VSD, enhanced by the presence of septal malalignment, could favor the development of an abnormal fibrous tissue below the aortic valve.\textsuperscript{26-33}

In the subset of patients with malalignment VSD produced by a posteriorly deviated infundibular septum, all four had associated obstructive lesions of the aortic arch (table 2, p < .001). On the other hand, in the patients with malalignment VSD due to anterior deviation of the infundibular septum and a subaortic ridge, the prevalence of obstructive lesions of the aortic arch was low. This finding leads to the attractive speculation that these special types of VSD, which
produce septal malalignment by posterior deviation of the outlet septum, could favor preferential flow from the left ventricle to the pulmonary artery during fetal life, thus contributing to the genesis of obstructive lesions of the aortic arch.\textsuperscript{15, 16} We suggest that the same mechanism could explain the presence of a subaortic ridge in the setting of malalignment VSD.

Despite the high prevalence of small defects in association with discrete subaortic obstruction, it is doubtful that the presence of accessory tricuspid valve tissue (as a mechanism of VSD spontaneous partial closure)\textsuperscript{34-38} is anatomically related to the formation of a subaortic shelf. On the other hand, it has already been postulated that surgical closure of a VSD can stimulate the progression of a previously "silent" subaortic ring.\textsuperscript{11} Accordingly, the spontaneous decrease in the size of a VSD could favor the progression of a fibrous subaortic shelf, whatever mechanism is involved.\textsuperscript{30, 31}

Based on this theory, it is possible that some cases of discrete subaortic stenosis in the adult could be a late clinical presentation of the association of VSD and a subaortic fibrous ridge in which the VSD has closed;


table 2

<table>
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<tr>
<th>Association of malalignment VSD (and subaortic stenosis) with obstructive lesions of the aortic arch</th>
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<tr>
<td></td>
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<tr>
<td>Present</td>
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<tr>
<td>Absent</td>
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<tr>
<td>Total</td>
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\(p < .001\) for comparison of present vs absent.
\textsuperscript{A}Coarctation.
\textsuperscript{B}Three patients with coarctation and one patient with aortic arch interruption.
FIGURE 4. VSD with subaortic shelf. Angiocardiographic appearance of the subaortic ridge below the VSD. AO = aorta; LV = left ventricle; SR = subaortic ridge.

this is an especially strong possibility in the presence of septal malalignment.

The practical implications of the present study are that a subaortic fibrous ridge should be routinely searched in the presence of VSD with septal malalignment and, once detected, this ridge should always be excised during surgery for VSD closure, since it carries a high risk of postoperative left ventricular outflow tract obstruction. If the defect is small and surgery is not indicated, the patient should be closely followed up because of the possibility of progressive subaortic obstruction after spontaneous closure of the defect.

Subsequent studies are necessary to establish a possible common morphogenesis of malalignment VSD, obstructive lesions of the aortic arch, and fixed subaortic stenosis.

We appreciate the cooperation of Dr. Beatriz Leão, Dr. Ivo Nesralla, Dr. Paulo Prates, Dr. Renato Kalil, Dr. João Ricardo Sant’Anna, Dr. Iran Castro, Dr. Nestor Daudt, Dr. Raul R. Rossi, and Dr. José Roberto Goldim. The technical assistance in the preparation of the manuscript was provided by Mrs. Mara R. Fieburg and Ms. Helena M. Rossi, to whom the authors are also grateful.

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PATHOPHYSIOLOGY AND NATURAL HISTORY—CONGENITAL HEART DISEASE

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Subaortic fibrous ridge and ventricular septal defect: role of septal malalignment.
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Circulation. 1987;75:1124-1129
doi: 10.1161/01.CIR.75.6.1124

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