Aneurysm of the Membranous Septum with Interventricular Septal Defect Producing Right Ventricular Outflow Obstruction


Aneurysm of the membranous portion of the interventricular septum is an uncommon congenital cardiac malformation rarely diagnosed during life. To our knowledge the literature contains four previous cases. Two of these were confirmed at surgery, the other two were visualized by angiography alone. This is the report of an unusual case in which the aneurysm produced hemodynamically significant right ventricular outflow tract obstruction. The aneurysm and an associated interventricular septal defect were both corrected at surgery. One similar case, also surgically corrected, has been reported in the Swedish literature.

Case Report

A 12-year-old white girl was initially seen at Walter Reed General Hospital on March 3, 1961, because of a heart murmur noted since birth. The past history indicated an uneventful pregnancy and delivery. She had frequent respiratory infections between the ages of 3 and 5. Subsequently, she was asymptomatic with normal growth and development and without cyanosis or congestive failure. There was no history of bacterial endocarditis, rheumatic fever, or cardiac arrhythmias. Positive physical findings were confined to the heart. There was a right ventricular lift; a systolic thrill at the third left intercostal space; a grade-IV/VI systolic murmur with an ejection quality heard best in the same area and well transmitted to the pulmonic area; and a widely but physiologically split S2 with a soft pulmonic component.

The blood count and urinalysis were normal. Ear oximetry values were 98 per cent before and 99 per cent after exercise. Chest x-ray with barium in the esophagus was interpreted as within normal limits. The electrocardiogram was normal with a frontal QRS axis +90°. A right heart catheterization had been performed at another hospital at the age of 5. At that time oxygen step-up was noted at the ventricular level with an estimated pulmonary to systemic flow of 1:5:1. A gradient of approximately 20 mm. Hg was recorded across the outflow tract of the right ventricle (table 1). Right heart catheterization was repeated at Walter Reed General Hospital on April 4, 1961. The findings were essentially unchanged from the previous catheterization (table 1, fig. 1). Elective repair of the interventricular septal defect and presumed infundibular pulmonic stenosis was advised.

Operation

Operation was performed on June 12, 1962, with use of the Mayo-Gibbon pump-oxygenator. Inspection of the heart revealed no evidence of an infundibular chamber and only a coarse sys-

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Table 1

<table>
<thead>
<tr>
<th>Age (Preoperative)</th>
<th>Right atrium Pressure</th>
<th>Low right ventricle Pressure</th>
<th>High right ventricle Pressure</th>
<th>Pulmonary artery Pressure</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>O2</td>
<td>O2</td>
<td>O2</td>
<td>O2</td>
</tr>
<tr>
<td>5</td>
<td>6 71%</td>
<td>50 73.5%</td>
<td>35 78.4%</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>7 79%</td>
<td>48 87%</td>
<td>35 90%</td>
<td></td>
</tr>
<tr>
<td>(Postoperative)</td>
<td>6 72%</td>
<td>20 73%</td>
<td>16 72.5%</td>
<td></td>
</tr>
</tbody>
</table>

From the Cardiovascular and Thoracic Surgery Services, Walter Reed General Hospital, Washington, D. C.
Figure 1

Preoperative and postoperative pressure tracings recorded at catheterization. Note that paper speed and standardization differ in the two records.

tolic thrill was palpable over the outflow tract of the right ventricle. This thrill radiated into the main pulmonary artery. An oblique right ventriculotomy was made. Exploration of the interior of the ventricle revealed a 1-cm. defect in the center of the membranous septum. The defect was associated with an aneurysmal dilatation of the remaining portion of the membranous septum. There was no evidence of valvular pulmonary stenosis or of fibromuscular obstruction in the infundibular region. It was noted, however, that during ventricular systole the membranous septal remnant bulged into the outflow tract of the right ventricle and produced a partial functional obstruction. Repair was accomplished by direct closure of the septal defect with imbrication of the aneurysmal remnant of the membranous septum in the suture line. Six interrupted sutures were required to close the septal defect adequately. The associated imbrication technic appeared to obliterate completely the septal aneurysm and to relieve the obstruction (Fig. 2). Following closure of the ventriculotomy and termination of cardiopulmonary bypass, no residual thrill was palpable over the right ventricle or main pulmonary artery. Chest closure was accomplished with temporary tube drainage. The patient's postoperative course was entirely uneventful.

Evaluation, 1 year following surgery, showed no functional cardiac abnormalities. A grade-I/VI systolic ejection murmur was present in the pulmonic area. $S_2$ was widely and persistently split. The electrocardiogram demonstrated com-
complete right bundle-branch block, which had been present since surgery. The chest x-ray was normal. Right heart catheterization performed on January 5, 1964, showed no intracardiac shunt either by oxygen step-up or by ascorbic acid platinum electrode studies. A small gradient of approximately 4 mm. Hg in the right ventricular outflow tract was judged as clinically insignificant (table 1, fig. 1).

Discussion

It is generally accepted that aneurysm of the membranous septum is congenital rather than acquired. Mall has suggested that the defect results from displacement of the aorta to the right and the muscular ventricular septum to the left. This malalignment between the aorta and muscular septum leads to a relatively horizontal membranous septum that is inherently weak and predisposes to aneurysm formation and possible perforation. The above theory was generally accepted by Lev and Saphir with the modification that congenital aneurysm of the membranous septum is a mild form of transposition.

In 1938 Lev and Saphir's extensive review of the world literature produced reports of 70 cases of aneurysms of the membranous septum. They added two cases of their own. A few sporadic cases have been reported in the literature since then, bringing the total to approximately 85. Rae encountered four cases in 3,000 necropsies and Steinberg, only two in 16,000.

The lesion may occur singly or in combination with other cardiac anomalies, particularly interatrial and interventricular septal

Figure 2

Artist’s conception of operative findings and technic of repair: a, aneurysm of ventricular septum with centrally located defect; b, diagrammatic production of right ventricular outflow obstruction; c, imbrication of the aneurysmal tissue in the septal defect closure; d, final result with closure of the septal defect and obliteration of the aneurysm.
defects. In one instance its association with subaortic stenosis was reported. These aneurysms may protrude superior to the tricuspid valve, inferiorly into the right ventricle, or occasionally inferiorly and anteriorly into the right ventricular outflow tract. With pulmonary stenosis and marked elevation of right ventricular pressure the aneurysm may protrude into the left ventricular outflow tract. The case reported by Leckert and Sternberg actually projected backward and upward behind the ascending aorta between the pulmonary artery and left atrium. Depending upon the size and location of the aneurysm, varying hemodynamic alterations may be produced. In our patient, the aneurysm protruded into the right ventricular outflow tract simulating infundibular pulmonic stenosis. Similar findings were observed in the case reported by Peräsalo et al. Schumacker and Glover reported the surgical correction of an aneurysm of the membranous septum associated with a small interventricular septal defect. Their case, however, did not have obstruction of the pulmonary outflow tract. When the aneurysm protrudes superior to the tricuspid valve, its subsequent rupture might provide the etiologic basis for a left ventricular-right atrial shunt.

Recognition of aneurysm of the membranous septum during life is difficult. Clinical manifestations are uncommon but serious cardiac arrhythmias have been reported. They may result from mechanical stimulation or distortion of structures in this irritable portion of the heart. The case reported by Lekisch was studied because of a cardiac murmur, which was possibly due to tricuspid insufficiency secondary to distortion of the valve by the aneurysm. Other clinical manifestations usually result from associated intracardiac lesions.

It appears that when these aneurysms are encountered fortuitously during open-heart surgery, resection or imbrication of the sac should be accomplished to prevent further enlargement, associated cardiac arrhythmias, or rupture. With the present frequency of open-heart surgery it is suggested that the lesion may be found to be more common than previously reported.

Summary

The second known case of aneurysm of the membranous interventricular septum simulating infundibular pulmonic stenosis associated with an interventricular septal defect is described. Hemodynamic studies before and after successful surgical correction of both lesions are presented. The current literature on the subject is briefly reviewed.

Addendum

Since this paper was submitted for publication an 8-year-old girl demonstrated similar operative findings. At open-heart surgery the ventricular defect was located eccentrically in an aneurysm of the membranous portion of the septum. The defect was closed and the aneurysm corrected by imbrication.

References

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Malpighi and the Capillaries

The answer to the thousand-year-old question had to be there: How does the artery communicate with the vein? No one denied such a connection, and had never done so, even before the circulation of the blood had ever been suspected.

Erasistratus himself had declared that veins and arteries joined up, and Cesalpino, in his usual negligent style, had written remarks about blood vessels which "do not end but rather carry on." But what exactly he had in mind when he wrote this, we do not know.

When Harvey proved the circulation of the blood in so many different and convincing ways, he found himself confronted by this question, too. But he also could only approach the mystery with conjectures. "... Either there is connection between the vessels," he wrote, "or else there must be pores allowing the passage of blood in the flesh and harder tissues." And he went on somewhat helplessly: "So far no one has brought to light anything valid concerning the connections between veins and arteries, and where and how and by what means they are present."

The experiments Harvey thought out for the solution of this problem were the basis for his final views on the subject. In his letter to Biolan written some twelve years previously, he wrote: "Neither in the liver, spleen, lungs, kidneys, nor any other viscus, is such a thing as a connection to be seen, and by boiling I have rendered the tissues of these organs so friable that it could be shaken like dust from the fibres or picked away with a needle, until I could trace every capillary filament distinctly. I can, therefore, boldly affirm that there is neither any anastomosis of the vena portae with the cava, of the arteries with the veins, nor of the capillary ramifications of the biliary ducts, which can be traced through the entire liver, with the veins."

This clearly supported the view that the blood vessels ended up blindly among the tissues where the blood disappeared like spring water lost in sand, to be gathered up again by the veins like underground water . . .

"From all this," he wrote to Borelli, "the question of the union of blood vessels and anastomosis can readily be solved in a perfectly acceptable manner. For if in one case nature wanted the blood inside the vessels and united the ends of the vessels into a network, it is probable that the vessel-ends in other places are also connected by means of their opening into each other."—Tibor Dobry, M.D. Discoverers of Blood Circulation. From Aristotle to the Times of Da Vinci and Harvey. New York, Abelard-Schuman, 1963, pp. 228, 232-233.
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