Subpulmonary Obstruction in Congenitally Corrected Transposition of the Great Arteries due to Ventricular Membranous Septal Aneurysms

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SUMMARY The clinical, hemodynamic, and angiographic observations, as well as the surgical approach used for repair in three patients with congenitally corrected transposition of the great arteries and ventricular membranous septal aneurysms, are presented. In two of the three patients the membranous septal aneurysm caused subpulmonary obstruction, with 94 and 125 mm Hg systolic gradients. In each patient the aneurysm was demonstrated by angiography, which also showed differences in size and shape with cardiac systole and diastole.

VENTRICULAR MEMBRANOUS SEPTAL ANEURYSMS are commonly found in small, uncomplicated ventricular septal defects. Over the last decade, numerous reports have described the clinical, anatomic, hemodynamic, and angiographic findings associated with these lesions and their possible complications.

In patients with normal ventricular development, atrioventricular (A-V) concordance and normally related great arteries, the aneurysms are often described as incidental findings following angiocardiographic studies and only rarely cause right ventricular outflow obstruction. Ventricular membranous septal aneurysms in patients with transposition of the great arteries (TGA) with and without A-V concordance and ventricular septal defects have been reported rarely. When present, however, these aneurysms usually caused significant subpulmonary obstruction.

Review of the previously described reports indicates that patients with congenitally corrected transposition often display various forms of pulmonary outflow obstruction and when a ventricular membranous septal aneurysm exists, a significant subpulmonary obstruction is present in most patients. The unique anatomic relationship between the pulmonary artery and a ventricular membranous septal aneurysm in patients with transposition of the great arteries with and without atrioventricular discordance explains why subpulmonary obstruction sometimes develops.

The purpose of this report is to present the clinical, hemodynamic, and angiographic findings of three patients with congenitally corrected transposition of the great arteries and ventricular membranous septal aneurysms recently operated upon at the Columbia-Presbyterian Medical Center. In two of the three patients the ventricular membranous septal aneurysm caused significant subpulmonary obstruction. The unique relationship between the ventricular septal defect and the pulmonary valve in these cases makes it likely that ventricular membranous septal aneurysms will result in significant subpulmonary obstruction. We believe that these findings are of special significance at this time when an increasing number of children with various forms of transposition of the great arteries are being referred for surgical correction.

For the purpose of this presentation the authors define congenitally corrected transposition as the congenital cardiac anomaly resulting from atrioventricular discordance and transposition of the great arteries such that both great arteries arise from the inappropriate ventricle.
Clinical Observations

Table 1 summarizes the clinical, electrocardiographic, and roentgenographic findings in the three patients with congenitally corrected transposition seen in our institution. All three patients had situs solitus. Two patients were asymptomatic and one patient had severe left-sided A-V valve insufficiency and congestive heart failure. Two patients had tiny ventricular septal defects and in one patient the ventricular membranous septal aneurysm was completely intact.

The hemodynamic data are presented in Table 2. The femoral arterial oxygen saturation ranged between 95–97% and in none of the cases was there evidence of an intracardiac left-to-right shunt by oxygen saturation analysis. In two patients the anatomical left ventricular pressure (subpulmonic right-sided ventricle) exceeded systemic ventricular pressure, with a subvalvar peak systolic gradient of 94 and 125 mm Hg. There was no pressure gradient recorded at the subvalvar region in the other child.

In each of the patients the diagnosis of ventricular membranous septal aneurysm was made by angiocardiography. Figure 1 shows anteroposterior and lateral views of a selective ventricular (anatomic left ventricle) angiocardiogram in diastole and in systole in the patient with a subpulmonarystolic gradient of 94 mm Hg. A filling defect is clearly seen at the subpulmonary region, indicating the presence of the ventricular membranous septal aneurysm.

Serial biplane films and cineangiocardiograms revealed a distinct change in the systolic and diastolic location and appearance of the filling defect. With ventricular systole the aneurysm decreased in size, became flattened, and moved upward toward the pulmonary orifice (fig. 1A, 1C).

Figure 2A shows an anatomical left ventricular injection in the 14-year-old patient with congenitally corrected transposition and dextrocardia. The filling defect in the subpulmonary region (fig. 2A) is due to the ventricular membranous septal aneurysm. In figure 2B the aneurysm is clearly seen filled with contrast material.

Table 1. Patient Data

<table>
<thead>
<tr>
<th>Patient</th>
<th>SM</th>
<th>TB</th>
<th>MC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (yr)</td>
<td>5</td>
<td>8</td>
<td>14</td>
</tr>
<tr>
<td>Sex</td>
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<td>Female</td>
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<tr>
<td>Symptoms</td>
<td>None</td>
<td>None</td>
<td>CHF</td>
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<tr>
<td>Segmental Descrip.</td>
<td>(S,L,L)</td>
<td>(S,L,L)</td>
<td>(S,L,L)</td>
</tr>
<tr>
<td>Cardiac Position</td>
<td>Levo-cardia</td>
<td>Dextro-cardia</td>
<td>Dextro-cardia</td>
</tr>
<tr>
<td></td>
<td>Diastolic</td>
<td>2/6 Ap.</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>Ss</td>
<td>None</td>
<td>Single</td>
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<tr>
<td></td>
<td>Ss</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>ECG</td>
<td>PR (sec)</td>
<td>0.16</td>
<td>0.14</td>
</tr>
<tr>
<td></td>
<td>FAQRS</td>
<td>+90°</td>
<td>+90°</td>
</tr>
<tr>
<td></td>
<td>qV</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td></td>
<td>R/S V1 (mm)</td>
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<td>14/17</td>
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<tr>
<td></td>
<td>R/S V2 (mm)</td>
<td>20/12</td>
<td>3/0</td>
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<tr>
<td>Chest X-ray</td>
<td>C/T Ratio</td>
<td>Increased</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>PVM</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Aortic Arch</td>
<td>Left</td>
<td>Left</td>
<td>Left</td>
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</tbody>
</table>

Abbreviations: LSB = left sternal border; Ap = apical; C/T = cardiothoracic; CHF = congestive heart failure; Ss and Ss = second and third heart sounds; PVM = pulmonary vascular markings; FAQRS = direction of QRS vector in the frontal plane.

Following cardiac catheterization, all three patients underwent open heart surgery for repair of their defects. Surgical correction was carried out by excision of the aneurysm and ventricular septal defect closure in two patients and plication of the aneurysm in one patient. Because of an additional subpulmonary stenosis in one of the former patients, a dacron conduit incorporating a porcine valve (Hancock prosthesis) was used to bypass the obstruction. Sutures were placed in the fibrotic rather than muscular portions of the septum in order to avoid injury to the bundle of His, which crosses in these cases anterior and superior to the ventricular septal defect.14 15

Discussion

Although ventricular membranous septal aneurysms have been described in association with a number of congenital cardiac anomalies, the association with transposition of the great arteries with or without A-V discordance is rather rare.2 6 11 We are aware of several previously published reports of congenitally corrected transposition who had ventricular membranous septal aneurysms.2 6 9 11 Most of the previously reported cases deal with the pathological description of this congenital malformation. Including the present series, available hemodynamic data revealed that the anatomical left ventricular pressure was similar to or exceeded systemic pressure in six of eight cases. These findings indicate that ventricular membranous septal aneurysms in association with congenitally corrected transposition are likely to cause subpulmonary obstruction.

This phenomenon is related in part to the intraventricular anatomy in these patients. With normal ventricular anatomy and viscerotrial concordance, the pulmonary artery is separated from the tricuspid valve by the conus arteriosus, (fig. 3). The membranous ventricular septal defect lying inferior to the conus arteriosus is separated from the pulmonary valve by the crista supraventricularis and is below the infundibulum. In this situation it is unlikely that a membranous ventricular septal aneurysm will cause severe obstruction to the subpulmonary region. In contrast, in patients with congenitally corrected transposition, the pulmonary valve is usually in continuity with the right-sided A-V valve. The ventricular septal defect is adjacent and immediately inferior to the pulmonary valve, and there is no interposed crista supraventricularis (fig. 3). When a ventricular membranous septal aneurysm occurs in this situation it lies virtually within the pulmonary valve orifice and is likely to cause a significant subpulmonary obstruction.

It has been previously suggested that the direction toward which the aneurysm develops (e.g., within the right or left

Table 2. Hemodynamic Findings in Three Patients with Congenitally Corrected Transposition and Ventricular Membranous Septal Aneurysm

<table>
<thead>
<tr>
<th>Pt. No.</th>
<th>LV MPA Systolic</th>
<th>MVB MPA FA</th>
<th>Qp/Qs Insufficiency</th>
</tr>
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<tr>
<td>1</td>
<td>114/5</td>
<td>20/6</td>
<td>95/5*</td>
</tr>
<tr>
<td>2</td>
<td>145/15</td>
<td>20/10</td>
<td>120/80 t1 = 0</td>
</tr>
<tr>
<td>3</td>
<td>55/10</td>
<td>55/30</td>
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*Anatomical right ventricular pressure.

Abbreviations: LV = anatomical left ventricle; MPA = main pulmonary artery; MVB = mixed venous blood; Qp/Qs = pulmonary to systemic flow ratio.
FIGURE 1. Simultaneous anteroposterior and lateral views of an anatomical left ventricular angiocardiogram in a patient with congenitally corrected transposition, ventricular septal defect, and a membranous septal aneurysm in diastole (A and B) and in systole (C and D). Note the filling defect (A, arrows) below the pulmonary artery. Also note the differences in size, shape and location of the aneurysm between diastole (arrows, A) and systole (arrow, C).

ventricle) relates to the differences in pressure between the two cardiac chambers. In two of our patients the ventricular membranous septal aneurysm bulged toward the left ventricle and into the subpulmonary region, although the anatomical left ventricular pressure exceeded systemic pressure. It appears likely that the ventricular membranous septal aneurysm originally developed within the anatomical left ventricle, when the pressure was low. As the aneurysms
gradually enlarged, increased systolic gradients developed across the subpulmonary regions. It is reasonable to assume that when the anatomical left ventricular pressure reached or exceeded systemic pressure, the aneurysms were established by virtue of size and their final positions were no longer dependent on ventricular pressure relationships.

Figure 3. Schematic drawing of hearts with normal ventricular anatomy [S.D.S] (on the right) and congenitally corrected transposition [S.L.I.] (on the left) to demonstrate ventricular septal defect to pulmonary artery relationship. Note that in patients with normal cardiac anatomy the septal defect is separated from the pulmonary artery by the conus arteriosus and in most cases by the crista supraventricularis. In contrast, in patients with congenitally corrected transposition the ventricular septal defect is adjacent to the pulmonary valve.
The ventricular membranous septal aneurysms in our patients changed shape and relationship to the pulmonary valve during systole and diastole. In diastole, the filling defects due to the aneurysms are larger and somewhat lower in the anatomical left ventricular chamber. In our two patients in whom the pressure in the anatomical left ventricle exceeded the pressure in the anatomical right ventricle, the obstructing ventricular membranous septal aneurysms became smaller during systole and shifted toward the pulmonary valve. This phenomenon could be seen particularly well with cineangiographic studies.

Patients with congenitally corrected transposition often have various forms of pulmonary outflow obstruction.\(^\text{11}\) Among nine children with congenitally corrected transposition recently operated upon at our institution, seven had some type of pulmonary obstruction; two patients had pulmonary valve stenosis, one had isolated infundibular stenosis, one had pulmonary infundibular and valvar stenosis, one had pulmonary atresia and two had obstructive ventricular membranous septal aneurysms. Four of the nine patients required insertion of a Hancock prosthesis from the anatomical left ventricle to the pulmonary artery in order to bypass the pulmonary obstruction.

In each of the nine patients, the intraventricular conduction system was identified electrophysiologically and was found to be superior and anterior to the septal defects, as previously described.\(^\text{14, 15}\) All nine remained in normal sinus rhythm following completion of surgery. In one patient who required repeat surgery for a residual ventricular septal defect, the precise location of the conduction system was not identified at the second operation and complete heart block ensued.

Improvement in surgical techniques during the past few years and the ability to localize the cardiac conduction system during open heart surgery has allowed an increasing number of patients with congenitally corrected transposition to undergo open heart correction. In such patients it is of utmost importance that a precise anatomic diagnosis is made. It may be expected that with the increase in the number of children with congenitally corrected transposition referred for surgical correction, subpulmonary obstruction secondary to ventricular membranous septal aneurysm will be more frequently encountered.

References

Subpulmonary obstruction in congenitally corrected transposition of the great arteries due to ventricular membranous septal aneurysms.
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