Aneurysm of the Membranous Ventricular Septum in Transposition of the Great Arteries

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SUMMARY In patients with transposition of the great arteries (TGA), both the D- and L-forms, an aneurysm of the membranous ventricular septum (AMS) produces subpulmonic stenosis due to the higher right ventricular pressure which forces the aneurysm to protrude into the left ventricular, i.e., subpulmonic, outflow tract.

The clinical signs and symptoms, hemodynamic findings as well as surgical results were analyzed in eight patients with TGA and AMS.

The presence of an AMS should be suspected from hemodynamic data consisting of a combination of elevated right ventricular pressure, gradient across the left ventricular outflow tract and presence of a small ventricular septal defect with or without pulmonary artery hypertension. The AMS can be demonstrated by a right ventricular injection in the lateral view.

The anomaly needs to be corrected at the time of the Mustard procedure. If uncorrected it may lead to postoperative death or progressive obstruction. In the presence of an aneurysm even small ventricular septal defects should be closed by a patch and the aneurysm should be excised.

SUBPULMONIC STENOSIS in transposition of the great arteries (TGA) may be produced by an aneurysm of the membranous septum (AMS). The AMS can be recognized by cardiac catheterization and angiography and should be repaired at the time of the Mustard procedure.

This paper reports our experience with this type of subpulmonic stenosis. We discuss the diagnosis, hemodynamics, natural history and surgical results in eight patients with TGA and AMS.

Material

Between 1967 and 1974, 116 children with D-TGA were operated upon by the Mustard procedure at the Buffalo Children's Hospital. Among these we identified AMS in eight cases (7%). All eight children had cardiac catheterization and angiography prior to the Mustard procedure and three patients had a follow-up study. The diagnosis of AMS was made preoperatively in five children.

Five children with AMS were operated upon under surface-induced deep hypothermia.

Results

The clinical data are summarized in table 1.

The male-female sex ratio was 3:5 compared to a 3:1 predominance of males in the total group of children with TGA.

 Cyanosis at rest was marked in two patients (cases 1 and 6), both of whom had large aneurysms. Patient 1 had no ventricular septal defect (VSD) but a large atrial septal defect (ASD). Patient 6 had a small VSD, a small ASD and a small patent ductus arteriosus (PDA). Moderate or mild cyanosis was present in the other six.

Seven patients had systolic murmurs of grade III or louder. Patient 5 had no murmur prior to surgery and she was the only one without a pressure gradient across the left ventricular outflow tract. Each of the five patients with a pansystolic murmur had a VSD.

Each of the eight patients had right ventricular hypertrophy and none had left ventricular hypertrophy by electrocardiogram. The R wave in lead V1 exceeded 0.5 mm in only one patient.

Echocardiograms were obtained in three patients and each showed one or more of the following features compatible with subpulmonic obstruction: prolonged apposition of the mitral valve with the ventricular septum, early closure and systolic flutter of the pulmonary valve, abnormal systolic anterior movement of the anterior mitral leaflet.

The roentgenographic findings were uncharacteristic when compared to the average patient with D-TGA with or without VSD. The cardiothoracic ratio varied from 53% to 69% and pulmonary plethora was moderate in most.

The past history was negative for congestive heart failure in four, all with rather significant left ventricular outflow obstruction.

The hemodynamic and angiographic data at cardiac catheterization prior to the Mustard procedure and in three patients after the repair are given in table 2.

Aortic oxygen saturation ranged from 25% to 83%. However, only two patients had a systemic arterial oxygen saturation below 65% (patients 1 and 6). Pulmonary-to-systemic flow ratio as calculated from oxygen saturation data ranged from 1.3:1 to 4.4:1 and did not correlate with the degree of subpulmonic obstruction nor with the pulmonary artery pressure.

Left ventricular pressure ranged from 50 to 100 mm Hg, i.e., was elevated in all. In three patients (patients 5, 6 and 7), however, the increase was due to elevation of pulmonary artery pressure rather than to the subpulmonic obstruction since the gradient in these measured 20 mm Hg or less. In five patients the left ventricular pressure was elevated due to outflow obstruction, with gradients between 25 and 65 mm Hg.

The size of the AMS found at surgery correlated with the severity of obstruction in the five patients in whom pulmonary artery pressure was low or normal. A small aneurysm produced a gradient of 42 mm Hg in patient 4 while three large aneurysms (cases 1, 2 and 8) led to gradients of 51, 55 and 65 mm Hg, respectively.

In the three patients with pulmonary artery hypertension (cases 5, 6 and 7) the gradient across the left ventricular outflow tract was small or absent in spite of an AMS.

The AMS was demonstrated by angiography in five
patients. In the three others the angiograms were inadequate to recognize the AMS.

All except one patient (case 1) had a VSD, found by angiography or at surgery. The size of the VSD found at surgery was moderate in two patients, small (5 mm) in three patients and probe patent in two. The patient without a VSD at surgery had a small VSD early in life as proven by catheter passage. The VSD had closed by the time of the Mustard correction at the age of one year and ten months.

Comparing the catheterization studies in early life (available in all patients) with the presurgical studies, we found no evidence that the VSD had decreased in size in the seven patients with a defect found at surgery.

Three patients (cases 1, 4 and 6) had a postsurgical catheterization study. An obstructing AMS was identified in patient 1 whose VSD had closed spontaneously. The pressure gradient across the left ventricular outflow tract was progressive and resection of the AMS may be required in the future. In patient 4 the AMS had been surgically retracted and sutured into the VSD; however, the follow-up study showed that the aneurysm had reoccurred with protrusion into the left ventricle and a progressive gradient had developed across the outflow tract.

Patient 6 had a similar repair of a large AMS. A repeat study with left and right ventricular angiograms did not demonstrate an AMS.

Table 3 lists the chronological surgical data. Patients 4 to 8 were operated upon utilizing deep hypothermia and circulatory arrest. In patient 1, the AMS had not been identified by angiography preoperatively and thus was not repaired at operation.

Our experience with patient 2 made us aware of the entity of AMS in TGA. The angiogram showed the aneurysm (fig. 1) but at surgery it could not be seen from the right ventricle because it protruded entirely into the left ventricle and the VSD was rather slit-like. The VSD was patched and the Mustard correction carried out. The patient died three days after surgery after repetitive episodes of decrease in cardiac output while a loud systolic ejection murmur was heard. The presence of an obstructing subpulmonary aneurysm was postulated and identified at postmortem examination (fig. 2).

In patients 4, 6 and 8 the AMS was demonstrated by angiography and its presence confirmed at surgery. In patients 3, 5 and 7 the AMS was found at surgery through careful probing and inspection of the VSD. Patients 3 and 7 had suture closure of the ventricular septal defect, while the AMS was retracted and its tissue imbricated in the suture closure. When patient 4 had a follow-up catheterization two years after surgery it became evident that in spite of this type of correction the AMS still protruded into the left ventricle producing subpulmonary obstruction of moderate degree (see table 2). To avoid recurrence of the AMS in patient 8 the VSD was closed with a Dacron patch.

### Table 1. Clinical Data on Eight Patients with TGA and AMS

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Cyanosis</th>
<th>Hb</th>
<th>L5B murmur</th>
<th>ECG</th>
<th>X-ray</th>
<th>History of CHF</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>Marked</td>
<td>24.5</td>
<td>Ejection systolic grade III</td>
<td>RAE, RVH</td>
<td>53% moderate</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>Moderate</td>
<td>19.3</td>
<td>Pansystolic grade IV</td>
<td>RVH</td>
<td>62% moderate</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>Mild</td>
<td>17.1</td>
<td>Ejection systolic grade III</td>
<td>RAE, RVH</td>
<td>69% moderate</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>Moderate</td>
<td>15.7</td>
<td>Pansystolic grade IV</td>
<td>RAE, RVH</td>
<td>62% moderate</td>
<td>0</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>Moderate</td>
<td>20.0</td>
<td>No murmur</td>
<td>RAE, RVH</td>
<td>59% moderate</td>
<td>NK</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>Marked</td>
<td>16.8</td>
<td>Pansystolic grade III</td>
<td>RVH</td>
<td>66% mild</td>
<td>Yes</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>Mild</td>
<td>16.8</td>
<td>Pansystolic grade III</td>
<td>RVH</td>
<td>67% marked</td>
<td>Yes</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>Mild</td>
<td>19.4</td>
<td>Pansystolic grade III</td>
<td>RVH</td>
<td>62% moderate</td>
<td>Yes</td>
</tr>
</tbody>
</table>

Abbreviations: M = male; F = female; Hb = hemoglobin; L5B = left sternal border; RAE = right atrial enlargement; RVH = right ventricular hypertrophy; CTR = cardiothoracic ratio; IPBF = increase pulmonary blood flow; CHF = congestive heart failure; NK = not known.

### Table 2. Hemodynamic Data on Eight Patients with TGA and AMS Obtained at Preoperative Catheterization Study and at Postoperative Studies in Patients 1, 4 and 6

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Saturation %</th>
<th>Flow ratio</th>
<th>Pressure</th>
<th>LV-PA gradient</th>
<th>Angiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Pre</td>
<td>10/12</td>
<td>44/83</td>
<td>1.8:1</td>
<td>85/50</td>
<td>55/60/3</td>
</tr>
<tr>
<td></td>
<td>Post</td>
<td>7/12</td>
<td>99/68</td>
<td>1.0:1</td>
<td>116/70</td>
<td>80/12</td>
</tr>
<tr>
<td>2</td>
<td>Pre</td>
<td>2/12</td>
<td>72/90</td>
<td>2.7:1</td>
<td>112/64</td>
<td>100/9</td>
</tr>
<tr>
<td>3</td>
<td>Pre</td>
<td>0/12</td>
<td>83/92</td>
<td>4.4:1</td>
<td>125/50</td>
<td>50/7</td>
</tr>
<tr>
<td>4</td>
<td>Pre</td>
<td>8/12</td>
<td>67/88</td>
<td>2.6:1</td>
<td>82/40</td>
<td>62/4</td>
</tr>
<tr>
<td></td>
<td>Post</td>
<td>2/12</td>
<td>92/67</td>
<td>1.0:1</td>
<td>106/68</td>
<td>75/5</td>
</tr>
<tr>
<td>5</td>
<td>Pre</td>
<td>2/12</td>
<td>74/80</td>
<td>2.4:1</td>
<td>85/50</td>
<td>50/5</td>
</tr>
<tr>
<td>6</td>
<td>Pre</td>
<td>3/12</td>
<td>25/85</td>
<td>1.3:1</td>
<td>90/50</td>
<td>75/10</td>
</tr>
<tr>
<td></td>
<td>Post</td>
<td>1/12</td>
<td>95/56</td>
<td>1.0:1</td>
<td>85/55</td>
<td>40/0</td>
</tr>
<tr>
<td>7</td>
<td>Pre</td>
<td>5/12</td>
<td>82/89</td>
<td>3.5:1</td>
<td>100/35</td>
<td>80/14</td>
</tr>
<tr>
<td>8</td>
<td>Pre</td>
<td>2/12</td>
<td>71/84</td>
<td>3.5:1</td>
<td>80/50</td>
<td>90/10</td>
</tr>
</tbody>
</table>

Abbreviations: Ao = aorta; PA = pulmonary artery; LV = left ventricle; RV = right ventricle; VSD = ventricular septal defect; PDA = patent ductus arteriosus; AMS = aneurysm of membranous septum, TGA = transposition of great arteries.
Table 3. Surgical Data on Eight Patients with TGA and AMS

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Type</th>
<th>Age</th>
<th>Year</th>
<th>Aneurysm</th>
<th>Assoc. defects</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>7 days</td>
<td>Balloon</td>
<td>1</td>
<td>11/12</td>
<td>Not seen, not removed</td>
<td>ASD 2 cm</td>
<td>Alive, well 9 years, subpulm. obstruction</td>
</tr>
<tr>
<td>2</td>
<td>5/12</td>
<td>Blalock-Taussig</td>
<td>3</td>
<td>9/12</td>
<td>1 cm</td>
<td>Tunnel-like VSD, patched</td>
<td>Died 3 days postop, low cardiac output</td>
</tr>
<tr>
<td>3</td>
<td>3 days</td>
<td>Blalock-Hanlon</td>
<td>4</td>
<td>3/12</td>
<td>Small, imbricated</td>
<td>Probe patent VSD, sutured</td>
<td>Alive, well 9 1/2 years, not restudied</td>
</tr>
<tr>
<td>4</td>
<td>14 days</td>
<td>Balloon</td>
<td>7</td>
<td>12/10</td>
<td>Moderate, imbricated</td>
<td>Large ASD, pinpoint VSD, sutured</td>
<td>Alive, well 5 4/12 years, subpulm. obstruction</td>
</tr>
<tr>
<td>5</td>
<td>1 2/12</td>
<td>—</td>
<td>2</td>
<td>11/12</td>
<td>1 cm, imbricated</td>
<td>ASD 10 mm Small VSD, sutured</td>
<td>Alive, well 6 years, not restudied</td>
</tr>
<tr>
<td>6</td>
<td>3 days</td>
<td>Balloon</td>
<td>3</td>
<td>12/10</td>
<td>Large, imbricated</td>
<td>Probe patent ASD Small VSD (5 mm) sutured</td>
<td>Alive, 2 2/12 years, poor RV function, no subpulm. obstruction</td>
</tr>
<tr>
<td>7</td>
<td>7 days</td>
<td>Balloon</td>
<td>7</td>
<td>12/10</td>
<td>Moderate, imbricated</td>
<td>PDA, ligation Small ASD (5 mm) Small VSD (5 mm) sutured</td>
<td>Alive, well 2 years, not restudied</td>
</tr>
<tr>
<td>8</td>
<td>1/12</td>
<td>Balloon</td>
<td>2</td>
<td>0/12</td>
<td>Large, imbricated</td>
<td>Very large ASD, moderate VSD, patch closure Muscular VSD Small</td>
<td>Alive, well 2 1/2 years, not restudied</td>
</tr>
</tbody>
</table>

Abbreviations: VSD = ventricular septal defect; ASD = atrial septal defect; PDA = patent ductus arteriosus; RV = right ventricle.

Discussion

The membranous part of the interventricular septum may be anomalous in two ways: 1) it may be absent or deficient resulting in a ventricular septal defect, or 2) it may be redundant and produce an aneurysmal pouch. Combinations of deficiency and redundancy frequently exist.

Presence of an AMS in hearts with normally related great arteries usually does not lead to obstruction, except in very large aneurysms. The higher pressure in the left ventricle pushes the aneurysm into the right ventricle where it remains infracristal and distant from the outflow tract. In hearts with transposed great arteries, however, the systemic pressure in the right ventricle pushes the AMS into the left ventricular outflow tract and obstruction occurs readily because of its proximity to the pulmonary valve.

![Figure 1](https://via.placeholder.com/150)

**Figure 1** Right ventricular injection in lateral view showing right ventricle (RV), aorta (AO) originating from RV and membranous septal aneurysm (A) protruding underneath pulmonary valve.

![Figure 2](https://via.placeholder.com/150)

**Figure 2** Autopsy specimen of case 2. View from above through pulmonary artery showing pulmonary valve (PV), membranous septal aneurysm (A) and jet lesion (JL) in pulmonary artery wall.
The electrocardiogram is typical only for TGA. The conspicuous absence of left ventricular hypertrophy as we observed in our cases, was not readily explained in the presence of pressure and/or volume overload of the left ventricle. Previous studies\(^6\)\(^\text{-}10\) of the electrocardiogram in patients with TGA and VSD or pulmonic stenosis or both demonstrated frequent presence of biventricular hypertrophy.

The echocardiogram appears to be useful in the diagnosis of subpulmonic obstruction per se in TGA but is not specific for obstruction due to AMS.

The roentgenographic findings of the chest film are non-contributory.

The AMS can be demonstrated by angiography (figs. 1 and 3). A lateral right ventricular angiogram seems to be the best view and the AMS can be seen throughout systole as it protrudes underneath the pulmonary valve (fig. 1). Frequently it can also be seen in an anteroposterior view of the right ventricle or from a negative filling defect seen in a lateral or oblique view after left ventricular injection.

We encountered two patients with angiographic pictures suggesting small AMS but at surgical exploration (including view through the pulmonary valve) no AMS was found. Later review of the angiograms showed that the pouch seen on right ventricular injection filled for a short period only in late diastole and was compatible with a bulging septal leaflet of the tricuspid valve.

The presence of AMS should also be suspected whenever left ventricular pressure is elevated and/or a pressure gradient of 20 mm Hg or more exists across the left ventricular outflow tract. In patients with small VSD but elevated pulmonary artery pressure the gradient may be low (patients 5, 6 and 7). The presence of a patent ductus arteriosus may contribute to masking of the gradient.

A VSD is not always present in cases of AMS since they sometimes close spontaneously. In such cases the AMS would have to be visualized at surgery from above through the pulmonary valve. In all patients with TGA and VSD, and especially so with a small VSD, the membranous septum or its residues should be investigated for the presence of an AMS, since it may be easily overlooked lying on the left ventricular side of the septum.

The postoperative death of case 2 as well as the follow-up study on case 1 shows that AMS should be corrected at the time of the Mustard procedure. Simple retraction and suturing into the defect may not be satisfactory as experienced with patient 4. Further, the obstruction may progress in time (patient 1 and 4). Our original policy of direct suture closure of the VSD with AMS was dictated by the presence of thick fibrous tissue surrounding the defect. The aneurysmal tissue seemed also to be adequate for use as a buttress. Experience with surgical repair of VSD with normally related great arteries as well as in TGA showed that such a technique was unsatisfactory. Residual communications and/or recurrence of the aneurysm are seen postoperatively.

Our current policy is to close a VSD with or without aneurysm with a patch. Further repairs should show if it is advisable to excise the aneurysm completely and close the larger defect thus produced with a patch.

Patients with TGA, VSD and subpulmonic stenosis of other etiology (conus anomalies, mitral valve anomalies,
fibrous subpulmonic stenosis) may require a Rastelli procedure and corrective surgery therefore is recommended at a later age. For patients with TGA and subpulmonic obstruction due to AMS the Mustard type of repair can be done early in life.

References

RADIOLOGY

Complete Interruption of the Aortic Arch

2. Characteristic Angiographic Features with Emphasis on Collateral Circulation to the Descending Aorta

RICHARD B. JAFFE, M.D.

SUMMARY The angiograms of 17 patients with aortic arch interruption are reviewed to emphasize the variations in arch interruption and origin of the brachiocephalic vessels, and collateral circulation to the descending aorta. Depending on the anatomical type and subtype of arch interruption, collateral flow to the descending aorta in the presence of a stenotic or closed ductus will be dependent on the development of intercostal collaterals and/or the presence of retrograde flow in all brachiocephalic vessels arising from the descending aorta. Familiarity with the potential pathways for collateral circulation may permit differentiation into types and subtypes on chest radiograph. Patients with Type I interruption may have bilateral rib notching if the right subclavian artery originates normally from the innominate artery, but will have rib notching confined to the left side if the origin of the right subclavian artery is aberrant. Type II or Type III interruption patients will have rib notching confined to the right side if the right subclavian has a normal origin, but no rib notching if the origin of the right subclavian artery is aberrant.

□ COMPLETE INTERRUPTION of the aortic arch is characterized by discontinuity of the arch between the proximal ascending aorta and the distal descending thoracic aorta. The pathologic anatomy of the three different types and three subtypes of aortic interruption, and characteristic radiographic findings in 21 patients have been discussed in Part 1. This section will review the angiograms of 17 of these patients and others reported in the literature to enhance the recognition of the variations in arch interruption and origin of the brachiocephalic vessels, and collateral circulation to the descending thoracic aorta.

The site of arch interruption and origin of the brachiocephalic vessels are best delineated in patients without transposition following left atrial, left ventricular, or ascending aortic angiography. The ascending aorta is characterized by hypoplastic and ascends almost directly vertically into the neck in both frontal and lateral projections (fig. 1A, B). As will be discussed, the origin of the brachiocephalic vessels in relation to the site of arch interruption determines the potential for collateral circulation to the descending thoracic aorta.

The descending thoracic aorta originates from the pulmonary artery through a patent ductus arteriosus. Evans has introduced the term "pulmonary-ductus-descending aorta trunk" to include the pulmonary artery, patent ductus arteriosus, and descending thoracic aorta. In patients with Type II interruption, the left subclavian artery, and in Type III interruption, the left subclavian and left common carotid arteries arise from this trunk. If the right subclavian artery has an aberrant origin it also may be from this location. Typically the main pulmonary artery is moderately to

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