Continuous Murmur Following Mustard Operation for Transposition of the Great Arteries
A Sign of Pulmonary Venous Obstruction

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SUMMARY Three patients developed severe pulmonary venous obstruction following Mustard operation for transposition of the great arteries. Each patient had a soft continuous murmur with distinct diastolic accentuation at the low left sternal border or xiphoid area. Simultaneous recording of intracardiac sound and pressure in one patient showed that the murmur originated at the site of obstruction in the surgically-constructed pulmonary venous atrium. Selective cineangiograms demonstrated the baffle to be the cause of the obstruction. A continuous murmur in a patient following Mustard operation may suggest significant pulmonary venous obstruction.

PULMONARY VENOUS OBSTRUCTION is a relatively rare complication of the Mustard procedure for transposition of the great arteries (TGA). From 1964 through 1975, 83 Mustard operations were performed at the Children’s Hospital of Pittsburgh. Three of 58 survivors were found to have severe pulmonary venous obstruction, confirmed by cardiac catheterization and angiocardiographic studies. In each patient a soft continuous murmur was observed over the left lower sternal border or xiphoid area. A continuous murmur had not previously been reported to appear after the Mustard operation. This finding should suggest the presence of pulmonary venous obstruction.

Case Reports

Case 1

This patient was seen for the first time at Children’s Hospital of Pittsburgh at 34 days of age with cyanosis and grunting respirations of two weeks duration. Cardiac catheterization and angiography confirmed the diagnosis of transposition of the great arteries (TGA) with an intact ventricular septum. Following balloon atrial septostomy, systemic arterial saturation rose to 80% from 34%. Repeat cardiac catheterization at five months and again at two years of age demonstrated normal pulmonary artery pressure and pulmonary vascular resistance. At 3 3/12 years of age she underwent a Mustard operation using cardiopulmonary bypass with moderate hypothermia. Pericardium was used for the baffle. After this baffle had been placed, a small central portion of redundant baffle was reefed with a running horizontal mattress suture.

The immediate postoperative course was uncomplicated and the patient was discharged two weeks after operation in excellent condition. Ten months after surgery exertional dyspnea, fatigability, and mild cyanosis appeared. Thirteen months postoperatively a Grade II/VI soft continuous murmur was noted over the left lower sternal border. The murmur had distinct diastolic accentuation. S2 was single and loud. Heart rate was 110 beats/min and respiratory rate was 24/min. There was no evidence of cardiac decompensation. The chest roentgenogram showed minimal cardiomegaly with prominent right hilar vascular markings but no pulmonary venous congestion was noted (fig. 1A).

Cardiac catheterization demonstrated suprasystemic pulmonary artery pressure and markedly elevated pulmonary artery wedge pressure (table I). The pulmonary veins were entered in a retrograde fashion from the femoral artery via the aorta, right ventricle, tricuspid valve and pulmonary venous atrium. Mean pressure in a pulmonary vein was 40 mm Hg. Pullback pressure tracing from the pulmonary vein to the right ventricle showed an abrupt pressure drop between the proximal and distal pulmonary venous atrium (fig. 2). On angiocardiography there was an "hour glass" configuration of the pulmonary venous atrium (fig. 3B). There was neither baffle leakage nor obstruction to superior or inferior vena cava return.

At repeat operation, the baffle had narrowed the diameter of the central portion of the pulmonary venous atrium to 4 mm, producing severe obstruction between the posterior and anterior portion of this atrium (fig. 4B). The posterolateral wall of the pulmonary venous atrium was incised traversely; the incision extended between the right upper and lower pulmonary veins and an elliptical patch of Gore-Tex® was inserted to enlarge the posterior atrial wall (fig. 4C). Immediately following operation the pulmonary arterial mean pressure decreased to 30 mm Hg while the systemic pressure measured 110/60 mm Hg. Pressure in the pulmonary venous atrium was 6 to 8 mm Hg. The postoperative course was uneventful and the previously noted continuous murmur was no longer audible. At follow-up examination 11 months postoperatively the patient is well and free of symptoms.

Case 2

This patient was referred to Children’s Hospital of Pittsburgh at ten days of age because of cyanosis and

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Supported in part by a grant from the Beaver County (Pennsylvania) Heart Association.

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Received April 5, 1976; revision accepted May 14, 1976.

*Expanded polytetrafluoroethylene (PTFE) — Wiligore and Associates, Flagstaff, Arizona.

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tachypnea. Cardiac catheterization confirmed the diagnosis of TGA with an intact ventricular septum and an adequate balloon atrial septostomy was performed. Repeat cardiac catheterization at one year of age demonstrated low pulmonary artery pressure. Shortly thereafter a Mustard operation was performed under profound hypothermia with circulatory arrest and limited cardiopulmonary bypass. The baffle was constructed of pericardium. At the termination of the procedure the patch seemed to be bulging excessively and a reefing suture was placed in the baffle near the inferior vena cava.

The postoperative course was relatively uncomplicated and the patient was discharged 18 days after operation. Three months postoperatively, she was readmitted because of tachypnea, mild cyanosis, and congestive heart failure. Despite intensive medical therapy with digoxin and diuretics she remained in mild congestive heart failure. Cardiac examination showed a soft systolic ejection murmur over the base. S2 was single and loud. There was a Grade II/VI continuous murmur with diastolic accentuation at the mid to lower left sternal border and in the xiphoid area. A chest roentgenogram showed mild cardiomegaly with prominent central hilar vessels as well as increased pulmonary venous markings (fig. 1B). Cardiac catheterization was carried out via the femoral artery because of thrombosis of the distal inferior vena cava and previous cutdowns in the arms. The retrograde arterial catheter was manipulated into the pulmonary venous atrium from the right ventricle. The systemic venous atrium and the left ventricle were then entered through an opening in the baffle suture line. Mean pressure in the proximal pulmonary venous atrium, near the pulmonary veins was 40 mm Hg. Utilizing a micromanometer-tipped catheter, pressures and intracardiac sound were recorded simultaneously (fig. 5). The intracardiac phonocardiogram clearly demonstrated the presence of a continuous murmur with diastolic accentuation in the pulmonary venous atrium. Angiocardiography demonstrated severe obstruction within the pulmonary venous atrium (fig. 3C). There was minimal leakage across the baffle and no obstruction to systemic venous drainage was found.

At reoperation, the patient was found to have severe obstruction similar to that described in case 1 with an orifice only 2 mm in diameter. A Gor-Tex patch was similarly inserted to relieve the obstruction. Despite adequate surgical relief of the pulmonary venous obstruction, as determined

FIGURE 1. Chest roentgenograms of three patients following Mustard operation but before re-operation for pulmonary venous obstruction. A = case 1; B = case 2; C = case 3.

### Table 1. Catheterization Data Before and After Mustard Operation

<table>
<thead>
<tr>
<th>Patient</th>
<th>1</th>
<th>2</th>
<th>3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at pre-op cath</td>
<td>2 yrs</td>
<td>1 yr</td>
<td>2½ yrs</td>
</tr>
<tr>
<td>Pressures in mm Hg</td>
<td>Site</td>
<td></td>
<td></td>
</tr>
<tr>
<td>AO</td>
<td>95/60 (m = 75)</td>
<td>83/46 (m = 66)</td>
<td>95/50 (m = 70)</td>
</tr>
<tr>
<td>LA</td>
<td>m = 4</td>
<td>m = 3</td>
<td>m = 3</td>
</tr>
<tr>
<td>PA</td>
<td>20/5 (m = 11)</td>
<td>20/6 (m = 11)</td>
<td>12/6 (m = 9)</td>
</tr>
<tr>
<td>Age at op.</td>
<td>3-3/12 yr</td>
<td>15 mo</td>
<td>2-10/12 yr</td>
</tr>
<tr>
<td>Weight at op.</td>
<td>17 kg</td>
<td>8 kg</td>
<td>11.5 kg</td>
</tr>
<tr>
<td>Interval following op.</td>
<td>13 mo</td>
<td>3 mo</td>
<td>17 mo</td>
</tr>
<tr>
<td>Pressures in mm Hg</td>
<td>Site</td>
<td></td>
<td></td>
</tr>
<tr>
<td>PA</td>
<td>110/50 (m = 72)</td>
<td>N.E.</td>
<td>40/22 (m = 30)</td>
</tr>
<tr>
<td>PW</td>
<td>m = 45</td>
<td>N.E.</td>
<td>m = 24</td>
</tr>
<tr>
<td>PVA (prox)</td>
<td>m = 40</td>
<td>m = 40</td>
<td>N.E.</td>
</tr>
<tr>
<td>PVA (dist)</td>
<td>m = 5</td>
<td>m = 3</td>
<td>m = 2</td>
</tr>
<tr>
<td>AO</td>
<td>90/58 (m = 72)</td>
<td>110/60 (m = 83)</td>
<td>65/40 (m = 50)</td>
</tr>
</tbody>
</table>

Abbreviations: AO = aorta; LA = left atrium; m = mean pressure; N.E. = not entered; PA = pulmonary artery; PVA = pulmonary venous atrium; PW = pulmonary artery wedge; m = mean.
by intraoperative pressure measurement, the patient could not be weaned from cardiopulmonary bypass and died intraparatively. Permission for a post mortem examination was denied.

Case 3

This patient was noted to have a cardiac murmur and cyanosis at birth. Cardiac catheterization demonstrated transposition of the great arteries with intact ventricular septum and a balloon atrial septostomy was performed successfully. On repeat cardiac catheterization at eight months and again at 2½ years of age he was noted to have mild subpulmonic stenosis (table 1). He underwent a Mustard operation at 2 1/6 years of age under profound hypothermia with circulatory arrest and limited cardiopulmonary bypass. Pericardium was used for the baffle. The pulmonary valve was normal and no subpulmonic obstruction was noted. The postoperative course was uneventful and the patient continued to be asymptomatic from a cardiac standpoint. On examination three months after operation, a soft continuous murmur was heard at the left lower sternal border. S2 was loud and single. No other significant cardiac murmur was present. A chest roentgenogram showed normal cardiac size and normal pulmonary vascular markings (fig. 1C).

Cardiac catheterization 17 months postoperatively demonstrated moderately elevated pulmonary arterial pressure and a mean pulmonary artery wedge pressure of 24 mm Hg. An angiocardiogram in the lateral view demonstrated an “hour glass” appearance of the pulmonary venous atrium. There was no evidence of systemic venous obstruction or leakage of the baffle. The patient was reoperated upon and was found to have a baffle-induced severe stenosis within the pulmonary venous atrium. A Gor-Tex patch was used to enlarge the pulmonary venous atrium. The postoperative course was uncomplicated and the previously noted continuous murmur over the left lower sternal border disappeared. At follow-up examination eight months after reoperation the patient is asymptomatic and doing well.

Discussion

Obstruction to either superior or inferior vena caval return following Mustard operation1-8 is a widely recognized complication. Obstruction to the pulmonary venous return has been encountered less frequently.9, 13

Various causes of obstruction of the caval system or pulmonary venous drainage have been proposed. Initially shrinking of the pericardium9-13 or postoperative infections8 were implicated. To avoid the problem of supposed shrinkage of the pericardium, Stark et al. used a Dacron baffle but the incidence of caval obstruction increased.9 A similarly high incidence of pulmonary venous obstruction (seven out

Figure 2. Case 1. Pullback pressure tracing from the proximal pulmonary venous atrium (PVA) to the distal PVA. There is an abrupt pressure drop across the baffle. Arrow indicates the site of narrowing of the baffle.

Figure 3. Lateral views of selective pulmonary venous angiocardiograms. A) Angiocardiogram from a patient following the Mustard operation without pulmonary venous obstruction. Notice the wide opening of the baffle. B) Case 1. C) Case 2. Both show an “hour glass” appearance which represents the narrowing at the site of the baffle. Arrows indicate the site of the narrowing.
of 24 survivors) was encountered by Reul et al. in use of a contoured Dacron baffle. Deposition of platelets and other blood constituents in the folds of the Dacron patch was believed to cause further folding and contracture of the baffle. On the other hand, Stafford and McGoon had satisfactory results with an elastic knitted Dacron patch. Currently the use of pericardium is generally preferred. It would appear that the geometry of the baffle and method of placement is of prime importance. The use of a trouser-shaped baffle is reported to have decreased the incidence of baffle obstruction. Trusler has hypothesized that an excessively large baffle tends to adhere to itself or to the atrial septal area and to cause pulmonary venous obstruction (see Discussion, reference 7). Conversely, a small baffle is more likely to narrow one of the venae cavae. As suggested by Kilman et al., the enlargement of the pulmonary venous atrium with a pericardial patch at the time of the Mustard operation may prevent this complication.

Narrowing of the superior or inferior vena cava seems to be well tolerated even if severe, probably because the azygos vein or other collateral venous connections between the superior and inferior vena caval systems help to decompress unilateral vena caval obstruction in either direction. In contrast, significant pulmonary venous obstruction leads to severe pulmonary hypertension because of the lack of collateral channels between the pulmonary veins and the newly constructed pulmonary venous atrium. Patients who died suddenly even before clinical recognition of their problem were found at autopsy to have severe pulmonary venous obstruction. Early detection of pulmonary venous obstruction is of critical importance.

Pulmonary venous obstruction is frequently not easily recognized, particularly in the early postoperative period. Although there may be dyspnea on exertion or easy fatigue, the patient may be asymptomatic. Radiographic examination may not be helpful. Only one of our patients (case 2)
has apparent radiographic evidence of increased pulmonary venous markings although all had high pulmonary venous pressure. The single most helpful clinical finding was the continuous murmur heard in each of our three patients with pulmonary venous obstruction. Following the recognition of its significance in our first patient, the presence of a continuous murmur in the other two children led to a suspicion of pulmonary venous obstruction. The murmur is localized to the left lower sternal border or the xiphoid area. The continuous murmur in each of our patients was soft, medium-pitched, and had distinct diastolic accentuation. The intensity of the murmur did not change with respiration or during a Valsalva maneuver. The intracardiac pressure and sound recordings in one of our patients has shown the murmur to originate at the site of narrowing within the pulmonary venous atrium. Continuous murmurs have been previously reported in a patient with cor triatriatum and in patients with mitral atresia or severe mitral stenosis with a restricting inter-atrial opening. In such patients there is either a large inter or intra-atrial pressure gradient similar to that found in our patients.

Pulmonary venous obstruction from the baffle has been a progressive problem and a late complication. When a continuous murmur is noted on the postoperative follow-up examination, prompt further diagnostic study (cardiac catheterization and angiocardiography) should be performed to rule out the possibility of pulmonary venous obstruction. This may allow early correction and prevent the sudden death caused by pulmonary venous obstruction.

References

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A sign of pulmonary venous obstruction.
S C Park, F H Weiss, R D Siewers, W H Neches, J R Zuberbuhler and C C Lenox

Circulation. 1976;54:684-688
doi: 10.1161/01.CIR.54.4.684

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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the World Wide Web at:
http://circ.ahajournals.org/content/54/4/684

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