Silent Unilateral Pulmonary Venous Obstruction*

Occurrence after Surgical Correction of Transposition of the Great Arteries

James E. Lock, M.D.; Russell V. Lucas, Jr., M.D.; Kurt Amplatz, M.D.; and F. Blanton Bessinger, Jr., M.D.

An 11-year-old girl was found to have completely obstructed left pulmonary veins eight years following corrective surgery for transposition of the great arteries. The patient was acyanotic and asymptomatic. Retrograde flow of arterial blood from the affected left lung accounted for an angiographic appearance that mimicked occlusion of the left pulmonary artery and resulted from a failure of systemic venous development. Pulmonary venous anatomy could only be demonstrated by pulmonary arterial wedge angio graphic studies. This experience emphasizes that complete unilateral pulmonary venous obstruction may occur in an asymptomatic patient and underlines the importance of investigating pulmonary venous anatomy in any patient with gross inequality of the distribution of pulmonary blood flow.

Unilateral pulmonary venous obstruction in childhood is usually congenital in origin. The advent of Mustard's operation for correction of transposition of the great arteries has created a new class of patients who may develop unilateral pulmonary venous obstruction. Pulmonary infiltrates, hemoptysis, and pulmonary hypertension are generally present in both congenital and acquired cases.1,2

We are reporting the findings in a child who developed complete left-sided pulmonary venous obstruction following Mustard's operation. She has the unusual features of a symptom-free course and retrograde flow of oxygenated blood from the affected lung. Utilizing the hemodynamic and angiocardiographic data, we have speculated on possible physiologic mechanisms and have discussed the practical implications for other children undergoing Mustard's operation.

CASE REPORT

An 11-year-old girl had had cyanosis in infancy; when she was two years of age, signs of congestive heart failure were noted. The diagnosis of d-transposition of the great arteries with an atrial septal defect, an intact ventricular septum, and modest elevation of the left ventricular pressure (50/0-8 mm Hg), was made. An angiogram demonstrated anterograde flow to both left and right pulmonary arteries (Fig 1A).

At the age of 30 months, the patient underwent Mustard's operation, and after surgery, she was acyanotic and asymptomatic. When she was six years of age, a routine catheterization...
ization revealed findings consistent with a good surgical result. The pulmonary artery was not catheterized; however, a left ventriculogram (Fig 1B) demonstrated no anterograde flow to the left pulmonary artery.

A second postoperative catheterization was done when the patient was 11 years old. On examination, she was well-developed, acyanotic, and in no distress. The chest was clear bilaterally to auscultation and percussion. Heart sounds were normal; a grade 3/6 harsh systolic ejection murmur was loudest at the upper left sternal border. Diastole was clear. The electrocardiogram demonstrated right ventricular hypertrophy. The chest x-ray film demonstrated increased vascular markings in the right lung, mild left pleural thickening, and relative hypertrophy of the right hemithorax, but no scoliosis.

The hemodynamic data are presented in Table 1. Of note are the normal pressures in the right, left, and main pulmonary arteries and the elevated left pulmonary wedge pressure (mean of 24 mm Hg, as opposed to the right pulmonary wedge pressure of 5 mm Hg). The finding of fully saturated blood throughout the left pulmonary artery indicated retrograde flow of arterial blood from the left lung. Aortographic studies demonstrated that this oxygenated blood originated from enlarged left intercostal arteries and other systemic arteries. Selective cinearteriographic studies confirmed the presence of a normal left pulmonary arterial tree and demonstrated retrograde flow of all contrast material from the site of injection into the right pulmonary artery.

![Figure 1. Left ventriculograms. A (left). Prior to surgery, there is anterograde flow to both right and left pulmonary arteries. B (right). Following surgery, only right pulmonary artery becomes opaque.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21002/)

**Table 1—Hemodynamic Data**

<table>
<thead>
<tr>
<th>Vessel</th>
<th>Oxygen Saturation, percent</th>
<th>Pressure, mm Hg*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>66</td>
<td>5 (mean)</td>
</tr>
<tr>
<td>Baffle (systemic venous atrium)</td>
<td>67</td>
<td>5 (mean)</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>67</td>
<td>46/0-9</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>70</td>
<td>20/10 (16)</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>70</td>
<td>20/10 (16)</td>
</tr>
<tr>
<td>Right pulmonary arterial wedge</td>
<td>93</td>
<td>v = 10 (5)</td>
</tr>
<tr>
<td>Left pulmonary artery</td>
<td>96</td>
<td>20/10 (16)</td>
</tr>
<tr>
<td>Left pulmonary arterial wedge</td>
<td>97</td>
<td>24 (mean)</td>
</tr>
<tr>
<td>Right pulmonary vein</td>
<td>97</td>
<td>v = 12 (8)</td>
</tr>
<tr>
<td>Pulmonary venous atrium</td>
<td>96</td>
<td>v = 10 (7)</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>95</td>
<td>120/0-10</td>
</tr>
<tr>
<td>Aorta</td>
<td>95</td>
<td>120/80</td>
</tr>
</tbody>
</table>

*Values within parentheses are mean pressures.

![Figure 2. Catheter enters pulmonary venous atrium via defect in baffle. Contrast material injected under pressure fills right pulmonary veins; left border of atrium appears smooth.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21002/)
The literature of congenital stenosis of individual pulmonary veins has recently been reviewed. All reported children with significant unilateral pulmonary venous obstruction were symptomatic; most had had recurrent pulmonary infiltrates, hemoptysis, and pulmonary hypertension in infancy or early childhood. Nasrallah et al described a child with unilateral pulmonary venous atresia, recurrent infiltrates, and hemoptysis, who improved following pneumonectomy. They noted that injection of contrast material distal to a balloon-tipped catheter wedged in the pulmonary artery allowed visualization of systemic venous drainage from the affected lung.

The present case is an exceptional one in that complete obstruction was not associated with symptoms of any type. The plain chest x-ray films and angiograms when the patient was six years old indicated absent flow to the left lung, but since the patient was asymptomatic, the diagnosis of left pulmonary arterial occlusion was made. It was not until that artery was catheterized and wedge angiographic studies were performed that the correct diagnosis was made.

Two features of this case warrant further consideration. First, despite complete obstruction of the pulmonary venous drainage from the left lung, collateral systemic venous or lymphatic channels did not develop. In dogs, pulmonary venous occlusion is followed by hemoptysis and engorgement with fluids. Later clinical improvement occurs with the development of increased capacity of bronchial and transpleural veins and lymphatic vessels. Although the development of bronchial veins might be prevented by obstruction in the superior limb of the baffle, this complication was not present in this patient. The findings of retrograde flow of arterial blood in the left pulmonary artery has been described and suggests that the right lung, with a mean wedge pressure of 5 mm Hg, can provide a circuit with very low resistance for drainage of the blood supply to the left lung.

The persistence of low resistance in the right lung may be due to a gradual onset of obstruction. Trusler and Mustard have described gradual contraction of the baffle and development of adhesions following Mustard’s operation. A slow onset of obstruction might allow the contralateral lung to adapt, thus preventing the development of pulmonary hypertension and bronchial venous collateral vessels. Also, the onset of venous obstruction relatively late in development, unlike cases of congenital stenosis, may find a contralateral lung more capable of accepting increased flow. Ferencz and Dammann have noted anatomic data suggesting that the fetal vasoconstrictor response to pulmonary venous obstruction is more intense than the adult response.

Secondly, the development of enlarged intercostal arteries to the left lung cannot be explained by the

Figure 3. End-hole balloon-type catheter enters left lower pulmonary artery (LLPA) via left ventricle. Balloon is inflated and wedges catheter, thus preventing drainage of contrast material into right pulmonary artery. Injection of contrast material causes opacification of left pulmonary vein (LPV), as well as systemic pleuropericardial arteries (PPA). On late frames, left pulmonary artery is faintly opaque.

These data implied severe or total obstruction of the left pulmonary venous drainage. At a follow-up catheterization undertaken to define the anatomy of the left pulmonary veins, the previous hemodynamic findings were confirmed. The pulmonary venous atrium was entered via a defect in the baffle, and an angiogram demonstrated transient retrograde flow of contrast material into the right pulmonary veins. The left pulmonary veins did not become opaque (Fig 2).

A No. 7 Swan-Ganz end-hole catheter was advanced into the distal left lower lobar pulmonary artery. The balloon was inflated, and 10 ml of contrast material (60 percent diatrizoate sodium) was slowly injected distal to the occluding balloon. The angiograms obtained were recorded on 16-mm film and biplane cut-film (Fig 3). These films demonstrated filling of capillaries and subsequent opacification of the lower lobar vein, which communicated with a large left pulmonary vein. This large vein was seen on cineangiograms to move with the heart during contractions but did not empty into the left atrium. After several seconds, intercostal and pleuropericardial vessels, corresponding exactly to the arteries seen on aortograms, became faintly opaque. There was no opacification of bronchial or systemic veins. The contrast material cleared slowly, with late opacification of the left main pulmonary artery. There was no residual stain, and the patient remained asymptomatic.

These studies demonstrated complete obstruction of the left pulmonary veins. The left lung is supplied by enlarged intercostal and pleuropericardial arteries and is drained via the left main pulmonary artery into the right pulmonary vascular bed. When this “normal” route of egress was blocked by a balloon-tipped catheter, contrast material was cleared by retrograde flow in systemic arteries and by opacification of hypoplastic pulmonary veins draining, presumably, into the left main pulmonary artery. Systemic veins were not opaque on any films.

Discussion

The literature of congenital stenosis of individual pulmonary veins has recently been reviewed. All reports of unilateral pulmonary venous obstruction were symptomatic; most had had recurrent pulmonary infiltrates, hemoptysis, and pulmonary hypertension in infancy or early childhood. Nasrallah et al. described a child with unilateral pulmonary venous atresia, recurrent infiltrates, and hemoptysis, who improved following pneumonectomy. They noted that injection of contrast material distal to a balloon-tipped catheter wedged in the pulmonary artery allowed visualization of systemic venous drainage from the affected lung.

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Secondly, the development of enlarged intercostal arteries to the left lung cannot be explained by the
recognized causes of bronchial arterial enlargement, namely pulmonary hypertension and pulmonary arterial constriction. Experimental pulmonary venous obstruction alone has not resulted in enlarged bronchial arteries, although Nasrallah et al noted bronchial arterial enlargement in their case. It may be that bronchial arteries enlarge whenever pulmonary blood flow falls.

Unequal distribution of pulmonary blood flow in children with transposition of the great arteries has been a subject of concern to several authors. Muster et al described increased right pulmonary flow in many children with transposition of the great arteries and considered left venous obstruction a possible explanation; however, the distribution of flow was similar before and after surgery, suggesting that venous obstruction by the baffle was not a factor.

Our case has implications for other children surviving Mustard’s operation. First, a lack of clinical symptoms can occur with severe or even complete unilateral pulmonary venous obstruction, despite previous experience. Secondly, although the patient is asymptomatic, the left lung does not participate in the exchange of gases, and this child’s eventual prognosis is not known. Finally, it is likely that the left pulmonary veins have regressed in size over six years, and relief of the obstruction might have been possible had the diagnosis been made at the first postoperative catheterization. We would recommend, therefore, that after Mustard’s operation the distribution of pulmonary blood flow should be evaluated by chest x-ray film and perfusion lung scan and that pulmonary venous anatomy should be investigated, regardless of symptoms, in any patient with gross inequality in the distribution of pulmonary blood flow.

REFERENCES

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Echocardiographic Diagnosis of Atrial Septal Aneurysm in an Infant with Hypoplastic Right Heart Syndrome*

David J. Sahn, M.D.; Hugh D. Allen, M.D.; Robert Anderson, M.D.; and Stanley J. Goldberg, M.D.

Echocardiographic findings are described in a patient with hypoplastic right heart syndrome (pulmonary atresia type with intact ventricular septum and small right ventricular cavity) who had an associated atrial septal aneurysm. An unusual appearance of echoes behind the aorta bulging into the left atrium in diastole on both the M-mode and cross-sectional echo suggested this diagnosis prior to cardiac catheterization. The angiographic findings confirmed the diagnosis of right ventricular hypoplasia, pulmonary atresia and the large atrial septal aneurysm. The infant died after surgery and the atrial septal aneurysm was observed at autopsy. The importance of the diagnosis of the atrial septal aneurysm and its association with restriction of right-to-left atrial shunting prompts this report.

Infants born with significant obstruction to right ventricular inflow or outflow often require patency of the atrial septum in the form of an atrial septal defect or patent foramen ovale for their survival. Thus, emergency therapy for infants with forms of hypoplastic right heart syndrome (pulmonary atresia, tricuspid atresia) includes palliative systemic-pulmonary anastomosis and balloon atrial septostomy or operative septectomy with or without pulmonary valvulotomy. The pathologic findings of aneurysmal dilation of the atrial septum have previously been described. Additionally, the angiographic diagnosis during life of an atrial septal aneurysm in an infant with tricuspid atresia has recently been reported. In this latter report, the association of this finding with a restriction of right-to-left atrial shunting was emphasized. We wish to report the echocardiographic findings in an infant with hypoplastic right heart syndrome (pulmonary venous return. Circulation 18:1046-1056, 1957