The Development of Pulmonary Vascular Obstructive Disease After Successful Mustard Operation in Early Infancy

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SUMMARY Pulmonary vascular obstructive disease developed postoperatively in an infant with aortopulmonary transposition and intact ventricular septum who underwent a Mustard operation at 3 months of age. Preoperative catheterization had shown normal pulmonary artery pressures. Four months after surgery, catheterization showed pulmonary artery systolic pressure above the systemic level and a tortuous, attenuated pulmonary vascular tree visualized angiographically. Early corrective surgery may not preclude the development of pulmonary vascular obstructive disease in patients with aortopulmonary transposition.

PULMONARY VASCULAR obstructive disease may develop during infancy in patients with aortopulmonary transposition.1-4 Although it occurs more commonly in infants with an associated ventricular septal defect or patent ductus arteriosus,5,6 pulmonary vascular obstructive disease may occur in infants with an intact ventricular septum and no associated aortopulmonary communication. Controversy still exists about the best time to perform a Mustard operation on patients with aortopulmonary transposition and an intact ventricular septum. Because newer surgical techniques allow operative intervention in very small infants, some individuals have proposed that a Mustard procedure during the first months of life may be an approach to the prevention of pulmonary vascular complications in infants with aortopulmonary transposition.7-11

We are reporting a patient who underwent a Mustard procedure at 3 months of age and who developed subsequently severe pulmonary vascular obstructive disease.

Case Report

JI, a 2.7 kg newborn, was cyanotic at 2 hours of age. He was transferred to the Intensive Care Nursery of the Milton S. Hershey Medical Center.

Physical findings were consistent with aortopulmonary transposition. Arterial blood gases, sampled while the infant breathed 100% oxygen, showed PO₂ = 23 torr, PCO₂ = 24 torr and pH = 7.35. Cardiac catheterization on the first day of life confirmed aortopulmonary transposition with a small patent ductus arteriosus (table 1). Left ventricular systolic pressures were nearly equal to those in the right ventricle and aorta. After balloon atrial septostomy, the aortic oxygen saturation rose from 41% to 72%. The pulmonary arteries were not entered during the catheterization.

The infant was readmitted for catheterization at 2½ months of age because of increasing cyanosis (table 1). The hematocrit at the time of the second catheterization was 55%. The patent ductus arteriosus had closed, and the aortic oxygen saturation was 35% initially, but rose to 68% after a repeat balloon atrial septostomy. Left ventricular systolic pressure was mildly elevated, but well below systolic pressure in the systemic circuit. There was a 15 mm Hg systolic pressure gradient across the left ventricular outflow tract. The pulmonary artery pressure was normal. A left ventricular angiogram showed normal-appearing main and peripheral pulmonary arteries (fig. 1).

After the second catheterization the infant again became increasingly cyanotic. The electrocardiogram (fig. 2) showed right ventricular hypertrophy. Because of the infant’s increasing cyanosis, a Mustard procedure was performed at 11 weeks of age. Pericardium was used to construct the intra-atrial baffle, and the newly formed pulmonary venous chamber was enlarged using a gusset.12

At 5 months of age signs and symptoms of congestive heart failure developed, and digoxin and diuretic therapy were instituted. An electrocardiogram at 6 months of age revealed the interval development of left ventricular hypertrophy (fig. 3).

A repeat catheterization at 7 months of age (table 1) revealed no evidence of intracardiac shunting. Left ventricular and pulmonary arterial systolic pressures were higher than systemic arterial levels. Pulmonary arterial wedge pressures in both the right and left pulmonary arteries were 12 mm Hg, mean. The pulmonary venous atrium was entered by advancing a #5 Berman catheter retrograde from the right ventricle across the tricuspid valve. A 1 mm Hg mean pressure difference existed between the right-sided pulmonary veins and the pulmonary venous atrium. A left ventricular angiogram showed an enlarged main and proximal pulmonary arterial tree, with tortuosity and attenuation of distal pulmonary arteries and relative underopacification of the peripheral lung fields (fig. 4).
At 13 months of age, the infant died suddenly. The postmortem examination showed an intact intra-atrial baffle. There was no evidence of pulmonary venous obstruction, and no intracardiac thromboses were found. Photomicrographs from each of the lung lobes showed severe pulmonary vascular obstructive disease with angitis and intravascular thromboses of varying ages (fig. 5). Some of the thrombi were recanalized.

**Discussion**

Pulmonary vascular obstructive disease has been noted uncommonly after the Mustard procedure. Some authors, therefore, have argued that early corrective surgery may reduce the incidence of pulmonary vascular obstructive disease in patients with aortopulmonary transposition. Furthermore, no one has described an infant who underwent a Mustard procedure before 1 year of age and developed pulmonary vascular obstructive disease after surgery. Newfeld et al. reported two patients without ventricular septal defect or patent ductus arteriosus who developed pulmonary vascular obstructive disease 12 and 16 months postoperatively after undergoing the Mustard procedure at 13 months of age. In each case, pulmonary arterial systolic pressures were approximately one-half systemic arterial levels, in contrast to our patient whose pulmonary artery systolic pressure was above the systemic level. Mair et al. reported the development of pulmonary vascular obstructive disease in two of 54 patients with aortopulmonary transposition and intact ventricular septum who underwent the Mustard procedure at 2 and 2½ years of age. They felt that early corrective surgery might reduce the risk of development of pulmonary vascular disease after surgery. Godman et al. reported that two patients who underwent the Mustard procedure at 18 months and 3½ years of age had elevated pulmonary vascular resistance at cardiac catheterization 4 and 5 years, respectively, after surgery. Both had postoperative baffle leaks, and in neither case was the preoperative pulmonary vascular resistance reported. Newfeld and Rosengardt each reported the appearance of pulmonary vascular obstructive disease in an infant with aortopulmonary transposition and an intact ventricular septum who underwent the Mustard procedure at approximately 12 months of age. In each case pulmonary vascular resistance was normal before surgery.

The etiology of pulmonary vascular obstructive disease in patients with aortopulmonary transposition is not clear. Most of the postulated explanations focus on mechanisms which are present before but not after pulmonary vascular obstructive disease.

![Figure 1](http://circ.ahajournals.org/doi/fig/10.1161/01.CIR.58.1.182)  
**Figure 1.** Left ventricular angiogram at 2½ months of age, before Mustard procedure.
the Mustard procedure. These include hyperviscosity of the blood due to polycythemia, prominent bronchial collateral circulation to the lungs, and increased pulmonary blood flow and pulmonary arterial pressure. The development of pulmonary vascular obstructive disease after a successful Mustard operation is difficult to explain. Hyperreactivity of the pulmonary vascular bed may play a role. Pulmonary venous obstruction also must be considered as a cause of pulmonary arterial hypertension, since it can develop both immediately after surgery and as a late complication. In this case, however, the similarity of right and left pulmonary artery wedge pressures to pressures measured in the right-sided pulmonary veins and the pulmonary venous atrium excludes pulmonary venous obstruction. Newfeld has suggested that pulmonary arterial thromboses may cause pulmonary vascular obstruc-

**Figure 2.** Electrocardiogram before surgery, age 2½ months.

**Figure 3.** Electrocardiogram at 6 months of age, showing interval development of left ventricular hypertrophy.
corrective surgery for aortopulmonary transposition with intact ventricular septum. The course of this patient suggests that a successful Mustard operation performed at an early age may not prevent the development of pulmonary vascular obstructive disease in all patients. Thus, careful postoperative surveillance of infants who have undergone the Mustard procedure is most important.

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