Development of Pulmonary Arteriovenous Shunt after Superior Vena Cava-Right Pulmonary Artery (Glenn) Anastomosis

Report of Four Cases

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SUMMARY Four patients with cyanotic congenital heart disease who had previously undergone superior vena cava-right pulmonary artery (Glenn) anastomosis developed pulmonary arteriovenous malformations that resulted in significant intrapulmonary right-to-left shunting. This abnormality was documented by selective angiography, oximetry, and contrast echocardiography. It may be a major cause of late clinical deterioration in patients treated with the Glenn anastomosis.

THE CONCEPT OF ANASTOMOSING a large systemic vein to the right pulmonary artery to increase pulmonary blood flow in patients with cyanotic heart disease was proposed by Carlton et al.1 in 1951. In 1954, Glenn and Patiño2 described a modification of this anastomosis by suturing the superior vena cava (SVC) directly to the right pulmonary artery (RPA). Theoretical hemodynamic advantages of this method of palliation, compared with operations anastomosing a systemic artery to pulmonary artery (Blalock-Taussig, Potts shunt), were increased pulmonary blood flow accomplished by an absolute reduction in intracardiac right-to-left shunt and avoidance of excessive pulmonary blood flow.3 However, reports soon began to appear describing a high mortality among infants and frequent complications and early deterioration in children.4-6 These complications included superior vena cava syndrome,7 hemithorax,4 chylothorax,4, 8 venous collateralization around the anastomotic site,4, 5, 6, 8 pulmonary artery thrombosis,6, 6, 8 pulmonary infarction,9 and abnormal perfusion patterns.10, 11

This report describes yet another, not widely recognized, complication of the SVC-RPA anastomosis in four patients in whom abnormal pulmonary arteriovenous (AV) communications developed. The laboratory methods employed in documenting these abnormalities and their clinical significance are presented.

Report of Cases

Case 1

The patient was first evaluated at the Mayo Clinic when she was seven years of age. A heart murmur had been noted at birth, and cyanosis was apparent by six months of age. Cardiac catheterization at seven months revealed tricuspid atresia and pulmonary stenosis. Because of the cyanosis, a SVC-RPA anastomosis was performed at seven months of age. She did well for four years, after which time the cyanosis again worsened. Phlebotomies were required for polycythemia, and hypoxia-like episodes occurred on several occasions. A second catheterization was performed to assess the degree of pulmonary obstruction and the function of the SVC-RPA anastomosis. Angiography revealed a small, restrictive ventricular septal defect in addition to the pulmonary stenosis and atretic tricuspid valve. Contrast injection into the innominate vein showed the SVC-RPA anastomosis to be widely patent. Several small pericardio-phrenic collaterals were present. The terminal arterial vessels of the right lower lobe were grossly enlarged, and early pulmonary venous return into the right lower pulmonary veins was evident, suggesting abnormal AV shunting.

At seven years of age, a left Blalock-Taussig shunt was performed, and the venous collaterals arising from the innominate vein were divided. The postoperative course was uncomplicated; a continuous murmur throughout the left chest indicated patency of the systemic pulmonary arterial shunt.

Increasing cyanosis and polycythemia again became evident, however, six months after this second operation. The patient's exercise capacity became severely limited over the next two years. A third catheterization was performed, at age nine, to evaluate the patency of the Blalock-Taussig shunt and to reassess the arterial malformation in the right lung.

Normal oxygen saturations were obtained from the left lower pulmonary veins with 100% oxygen (table 1). In contrast, blood sampled from the right lower pulmonary veins was significantly desaturated while the patient breathed 100% oxygen. The PO2 of blood in the right pulmonary veins proved to be only 8 mm Hg greater than that in the right pulmonary artery, indicating nearly complete shunting, without oxygenation, of blood through that portion of the lung. Angiography indicated progressive angiomatoid malformation in the right lower lobe with rapid pulmonary venous filling from that portion of the lung (fig. 1).

Reparative surgery was attempted by a modified Fontan procedure.12 The SVC-RPA anastomosis was left intact. The patient died on the twelfth postoperative day from pulmonary insufficiency and low cardiac output. Microscopic examination revealed an increased number of arterioles and veins in the right lower lobe. The walls of the veins were distended and thickened.

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Table 1. Laboratory Findings

<table>
<thead>
<tr>
<th>Case</th>
<th>Superior vena cava</th>
<th>Pulmonary vein Right</th>
<th>Pulmonary vein Left</th>
<th>Femoral artery</th>
<th>Hemoglobin (g/dl)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>77 (52)*</td>
<td>82 (59.6)</td>
<td>100 (47.5)</td>
<td>94 (82)</td>
<td>20.8</td>
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<tr>
<td>3</td>
<td>75</td>
<td>100 (163)</td>
<td>100 (402)</td>
<td>91 (63)</td>
<td>17.7</td>
</tr>
<tr>
<td>4</td>
<td>61 (39)</td>
<td>68 (35.8)</td>
<td>100 (418)</td>
<td>82 (38.1)</td>
<td>21.2</td>
</tr>
</tbody>
</table>

*The numbers in parentheses are the PO₂ values in mm Hg.

Case 2

This patient was first seen at the Mayo Clinic at 12 years of age. She had been cyanotic since shortly after birth, and catheterization performed at three months of age had revealed pulmonary atresia, ventricular septal defect (VSD), and a patent ductus arteriosus (PDA) supplying confluent right and left pulmonary arteries. A SVC-RPA anastomosis was done at that time to increase pulmonary blood flow. Improvement was noted for four years, after which time the cyanosis gradually worsened. A second catheterization was performed at five years of age and showed several small collaterals around the anastomotic site and diminished flow into the left lung secondary to narrowing of the patent ductus.

By the age of 12, severe cyanosis was present and the patient had significant physical limitations. A third catheterization revealed the peripheral arterial saturation to be 74% and hemoglobin concentration 19.4 g/dl. Although the ductus arteriosus was stenotic at its insertion into the pulmonary artery, the left and main pulmonary arteries were near normal in size. Injection of contrast medium into the right superior vena cava demonstrated wide patency of the anastomosis. Preferential flow was into the right lower lobe, and angiomatoid malformation of the terminal arterial branches was present (fig. 2). The pulmonary veins from this area were rapidly opacified, suggesting AV shunting. Surgical correction was performed by patch closure of the VSD, ligation of the PDA, and insertion of a Dacron conduit (Hancock Laboratories) from the right ventricle to the left pulmonary artery. The SVC-RPA anastomosis was left intact. Because of persistent cyanosis, another catheterization was performed three weeks after operation. The presence of intracardiac right-to-left shunting was excluded by standard indicator-dilution curves as well as by angiography. Cyanosis was attributed to the persistent right-to-left intrapulmonary shunt in the right lung. The patient is asymptomatic two years after reparative surgery but remains mildly cyanotic.

Case 3

The patient was known to have congenital heart disease immediately after birth, when cyanosis and a heart murmur were detected. He began having hypercyanotic spells at seven months of age while hospitalized for varicella. Left hemiparesis followed one of these hypoxic episodes. He was first seen at the Mayo Clinic at 11 months of age. A clinical diagnosis of tricuspid atresia and pulmonary stenosis was made. Cardiac catheterization confirmed the diagnosis and a SVC-RPA anastomosis was performed to increase pulmonary blood flow. Arterial saturation increased from 63 to 93% after operation. For five years the patient remained only mildly cyanotic and exhibited normal exercise tolerance. Symptoms accelerated during his sixth year of life such that he could walk only one block without dyspnea and severe cyanosis.

At seven years of age a second catheterization was performed. The arterial saturation while breathing room air was 74% (PO₂ = 46 mm Hg), and the hemoglobin concentration was 17.7 g/dl. Tricuspid atresia and normally related great vessels were apparent by angiography. The ventricular septal defect appeared restrictive. An innominate
vein angiogram revealed that the SVC-RPA anastomosis was widely patent. Although preferential flow into the right lower lobe was seen, the appearance of the peripheral arteries was normal (fig. 3). Venous return from the right pulmonary veins appeared normal. Simultaneous sampling from the right and left lower pulmonary veins while breathing 100% oxygen demonstrated a markedly decreased PO_{2} in the right pulmonary vein compared with the left (table 1). Contrast echocardiography confirmed the presence of right pulmonary AV fistula.

A Fontan procedure was successfully performed several days after the second catheterization. The SVC-RPA anastomosis was left intact. No cyanosis was noted at the time of dismissal.

Case 4

After an uncomplicated pregnancy and delivery, cyanosis developed at seven weeks of age in this male infant, and a heart murmur was heard. Cardiac catheterization and angiography revealed tricuspid atresia with ventricular septal defect and pulmonary artery stenosis. Cyanotic spells developed by three months of age, and in the fifth month of life a SVC-RPA anastomosis was performed. Approximately eight years later, the patient was again markedly cyanotic and polycythemic (hemoglobin measured 22.3 g/dl), and he was experiencing severe headaches during physical activity. A second catheterization was performed at nine years of age to evaluate the status of the shunt. An injection into the SVC showed that the anastomosis was widely patent. Multiple dilated vessels were seen in the lower lobe of the right lung. Collateralization around the anastomosis was not apparent. A left Blalock-Taussig anastomosis was then performed. Improvement in color and activity occurred and lasted for approximately three years. However, five years after the second operation the hemoglobin concentration had again risen to 22 g/dl.

At age 14, a third cardiac catheterization was undertaken to assess the patency and function of the previous anastomosis. An angiogram performed in the left innominate vein again revealed the shunt to be widely patent. The malformation in the lower lobe of the right lung noted during the second study had expanded markedly (fig. 4). Early filling of the right lower pulmonary vein suggested that the AV shunt had progressed. Selective catheterization of the right lower pulmonary vein disclosed pronounced desaturation in comparison to blood returning from the left pulmonary vein, indicating minimal oxygen uptake in the right lung (table 1). Selective contrast echocardiography was also utilized to detect the presence of intrapulmonary right-to-left shunting.

Echocardiographic Contrast Studies

In cases 3 and 4, standard M-mode echocardiograms were performed at the time of catheterization. Typical features of tricuspid atresia (single demonstrable atrioventricular valve [mitral valve] with large excision, absent tricuspid valve, and small right ventricular chamber) were noted in both patients. Of great interest, however, were contrast echographic studies utilizing injections of indocyanine green dye and saline in a superficial (hand) vein, SVC, or RPA which detected and localized the intrapulmonary right-to-left shunt. The concentration and volume of dye and the technique of injection have been previously reported. Normal, the echo-producing qualities of indocyanine green dye are completely lost with a single transit through the pulmonary capillary bed. Therefore, after injection of indocyanine green dye into an upper extremity vein, SVC, or RPA in a patient with a SVC-RPA anastomosis, any appearance of echoes in the left heart chambers signifies presence of an abnormal right-to-left shunt. In both patients (cases 3 and 4), after hand vein and SVC injection of indocyanine green dye, a cloud of echoes appeared in the left atrium and left ventricle, suggesting an intrapulmonary right-to-left shunt. To ex-
clude the possibility that the dye might be entering the left atrium via collaterals around the SVC-RPA anastomosis, a Swan-Ganz balloon catheter was inflated to occlude the anastomotic site. With the balloon inflated, the early dye echoes were no longer seen. Moreover, selective injections of indocyanine green in the RPA also demonstrated the appearance of a cloud of echoes in the left ventricle, indicating intrapulmonary right-to-left shunting (fig. 5).

**Discussion**

The reason for late shunt failure in patients with a SVC-RPA (Glenn) anastomosis has been the subject of several reports. Samánek et al. described maldistribution of pulmonary blood flow within the shunted lung. With the use of radioisotopes, it was found that most of the blood entering the right lung perfused the lower lobe, and flow to the upper and middle lobes was markedly diminished. These findings were observed in the majority of the 15 patients an average of five years after establishment of the SVC-RPA anastomosis. The abnormal perfusion pattern was attributed to gravitational effects and nonpulsatile blood flow and was demonstrated to be altered by positional changes. Soon after, Boruchow et al. noted these changes angiographically in eight patients but considered the diminished flow within the right upper lobes to be a result of hypoxemia and secondary pulmonary artery vasoconstriction.

In a recent review of 63 patients palliated by SVC-RPA anastomosis, Mathur and Glenn described five patients who had laboratory and angiographic findings similar to those described in this report. Although evidence of pulmonary AV shunting within the hyperperfused lower lobe was recognized, the authors expressed uncertainty regarding the clinical significance of this abnormal AV communication. In our patients, prominent angiographic alterations and significant desaturation of pulmonary venous blood, in the absence of other significant venous collateralization, indicated that intrapulmonary AV shunt as a late complication of the Glenn anastomosis can be a major factor in the clinical deterioration of these patients. The echocardiographic and oximetric evidence of intrapulmonary shunting without the prominent angiographic changes (case 3) suggests that significant shunting may occur before obvious angiographic abnormalities become apparent.

The use of indocyanine green dye injections combined with echocardiography provided a sensitive means for detecting intrapulmonary right-to-left shunt in these patients. Because the echo-producing quality of indocyanine green dye is normally completely lost with a single transit through the pulmonary capillary bed, appearance of contrast echoes in the left heart after RPA injection must indicate right-to-left intrapulmonary shunting. A peripheral venous or SVC injection may be less specific because large venous collaterals around the anastomotic site to the inferior vena cava could conceivably result in appearance of contrast echoes in the left heart if an intra-atrial right-to-left shunt was also present (that is, tricuspid atresia). None of our patients had any large venous collaterals.

The best way to manage this problem surgically is unknown. The intrapulmonary shunting will continue, and perhaps progress, as long as the anastomosis remains functioning. Reconstruction of the SVC-RPA anastomosis is possible, but the effect of restoring pulsatile blood flow to a lung with AV fistula is not known. Increased pulmonary arterial pressure conceivably could result in increased shunting. On the other hand, regression of the shunting might occur if the anatomic changes were not advanced. For any given patient, the additional risk and operating time required to take down the SVC-RPA anastomosis would need to be weighed against any theoretical benefit. An alternative surgical approach would be to divert the lower right pulmonary venous return to the right atrium. In this situation unsaturated blood returning via the right pulmonary AV fistula would be carried to the left lung, where it could be oxygenated. The approach might be more suitable in patients who, after corrective repair, have normal or only minimally elevated mean right atrial pressures (pulmonary
atresia with VSD and tetralogy of Fallot). Diverting the pulmonary venous return into the right atrium in patients undergoing the Fontan procedure (tricuspid atresia) is less attractive because the right atrial pressure may be sufficiently elevated postoperatively to produce unilateral pulmonary edema. Other alternatives would be right lower-lobe lobectomy or take-down of the superior vena cava-right pulmonary anastomosis and reanastomosis of the superior vena cava to the right atrium with ligation of the right pulmonary artery.

Pulmonary AV shunting after the SVC-RPA anastomosis may be the major cause of late clinical deterioration in patients treated by this method. Since this acquired AV malformation destroys the gas-exchange capabilities in the right lung, we have been led to the conclusion that this procedure is presently the least attractive palliative shunt available for children with cyanotic congenital heart disease.

Addendum

Since acceptance of this manuscript, we have seen two additional patients with tricuspid atresia, ages 12 and 16 years, both of whom had undergone the Glenn anastomosis at the age of 1 year. As in case 3, after a selective angiogram, the right lower-lobe pulmonary arterial branches appeared normal with no evidence of AV malformation. However, presence of abnormal AV shunting in the right lower lobe was demonstrated by the technique of contrast echocardiography. After injection of indocyanine green dye in the superior vena cava and right pulmonary artery, a cloud of echoes appeared in the left atrium and left ventricle. These observations reinforce our impression that abnormal AV shunting may occur in patients with the Glenn anastomosis before obvious angiographic abnormalities are seen.

References

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