Use of Superior Vena Cava-Right Pulmonary Artery Anastomosis in Congenital Heart Disease with Decreased Pulmonary Blood Flow

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SUMMARY
Clinical, hemodynamic, and angiocardiographic findings on eight long-term (more than 3 years) survivors of anastomosis of the superior vena cava (SVC) to the right pulmonary artery (RPA) are described. All patients had severe cyanotic congenital heart disease with decreased pulmonary blood flow. Postoperative cardiac catheterization and angiocardiography defined the physiology of the shunt. SVC pressure was elevated after the creation of the SVC-RPA shunt. This was without obvious clinical effect in six patients. Mild SVC syndrome developed in the immediate postoperative period but was associated with overall clinical improvement in two cases. A late, and more severe SVC syndrome associated with clinical deterioration developed in three patients with severe pulmonic stenosis as part of their cardiac malformation. Clinical improvement and relief of SVC syndrome followed systemic-pulmonary artery shunt or open heart repair of the underlying cardiac malformation, leaving the SVC-RPA anastomosis intact. Clinical improvement following open heart repair of tetralogy of Fallot has persisted in spite of severe postoperative pulmonic valve insufficiency and elevated mean pressure in the left pulmonary artery. This is believed to be a result of the SVC-RPA shunt, which reduced the venous return into the right heart.

Additional Indexing Words:
Glenn procedure Pulmonic stenosis Superior vena caval syndrome
Tetralogy of Fallot

CIRCULATORY bypass of the right side of the heart by anastomosis of the superior vena cava (SVC) to the distal portion of the right pulmonary artery (RPA) has been shown to be compatible with long-term survival and freedom from postoperative congestive heart failure. This communication describes the clinical, hemodynamic, and angiocardiographic results in eight long-term survivors of this procedure. Its purpose is to discuss and emphasize the following: (1) certain etiologic factors and suggested management of a "late" SVC syndrome, (2) the development of unilateral pulmonary hypertension, (3) formation of collateral veins, and (4) role of collateral flow in SVC syndrome.

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(4) the possibility that SVC-RPA anastomosis may offer additional relief to patients with residual postoperative defects.

Methods

The eight patients who comprise our study group underwent SVC-RPA anastomosis when they were between 2 months and 11 years of age. Six of these patients were 1 year of age or older. All had severe cyanotic congenital heart disease with decreased pulmonary blood flow (table 1). The azygous vein was ligated at the time of the shunting procedure in all patients. The right pulmonary artery was divided near the pericardium and the distal end was anastomosed to the superior vena cava, either end-to-end or end-to-side. Three patients underwent additional surgery. One (case 8) had open heart repair for tetralogy of Fallot 2½ years after the SVC-RPA anastomosis. Blalock-Taussig anastomoses were performed 3 years after venous shunting in other two patients (cases 3 and 5).

The diagnoses were established in each patient by preoperative cardiac catheterization and angiocardiography carried out under light premedication. Patients were restudied 2 months to 1½ years following SVC-RPA anastomosis (tables 1 and 2). Two of the children (cases 5 and 8) had an additional study 2½ and 3 years after venous shunting. In three patients (cases 2, 3, and 8) indicator-dilution curves were obtained; the injection site was varied from above to below the anastomotic site. These eight patients have been followed for 3% to 6 years after undergoing SVC-RPA anastomosis.

Results

Clinical Aspects (Table 1)

All patients sustained marked clinical improvement following operation. Exercise tolerance was greatly increased. Although clubbing remained, cyanosis was diminished. There were no instances of postoperative congestive heart failure or chylothorax. Results at postoperative cardiac catheterization confirmed the impression of clinical improvement. Comparing postoperative data to preoperative data, hematocrit decreases of 13 to 38% (fig. 1a and b; table 1) correlated with absolute increases in arterial oxygen saturation of 3 to 68% (mean, 27%), and absolute increases in inferior vena cava oxygen saturations of 6 to 33% (mean, 18%). In one patient (case 8) the preoperative hematocrit level reflected a severe degree of iron deficiency anemia, and

<table>
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<th>Table 1</th>
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<tr>
<td>Clinical Experience With SVC-RPA Shunt*</td>
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<tr>
<td><strong>Case</strong></td>
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*See text for discussion.

Abbreviations: TA = truncus arteriosus; PS = pulmonic stenosis; VSD = ventricular septal defect; SV = single ventricle; CTGV = complete transposition of great vessels; PA = pulmonary atresia; DV = dextroversion; ASD = atrial septal defect.
Table 2

Pressure Relationships After SVC-RPA Shunt

<table>
<thead>
<tr>
<th>Case</th>
<th>Mean SVC pressure (mm Hg)</th>
<th>Pressure difference (mm Hg)</th>
<th>Across Anastomosis</th>
<th>Mean RPA-LA*</th>
<th>Mean RPA-IVC*</th>
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* RPA = right pulmonary artery; LA = left atrium; IVC = inferior vena cava.
† Recatheterized 3 years after operation. Mean SVC pressure was 13 mm Hg 1½ years after operation.
‡ Right atrium.
§ Recatheterized 2½ years after operation.

the clinical improvement was manifested by the substantial increase in arterial oxygen saturation.

In two patients (cases 5 and 8), a mild SVC syndrome was present from the time of operation. The signs of SVC syndrome included edema and dilated venous channels, involving the upper trunk, head, and neck. However, there was considerable overall improvement in clinical condition with decrease in cyanosis and hematocrit and increase in exercise tolerance and arterial oxygen saturation (table 1). The symptoms of mild SVC syndrome were easily controlled by upright positioning of the body. One and one-half and two years following operation the signs of SVC obstruction in these two patients became more severe. This late form of SVC syndrome was also associated with increasing cyanosis, elevated cerebrospinal fluid pressure, and decreased exercise tolerance. The hematocrit rose to the preoperative level (fig. 1b). Headache was a prominent complaint and was unrelieved by position or aspirin. In one of these patients (case 5) head growth was out of proportion to that of the rest of the body but was within the upper limits of normal for age. Cardiac catheterization and angiocardiography in these two patients established shunt patency and absence of constriction at the anastomotic site. There was a moderate increase of the collateral circulation between the superior and inferior vena cava. Left atrial pressure was normal in the one instance in which it was determined. This patient (case 5) underwent a successful left Blalock-Taussig anastomosis; another (case 8)

**Figure 1**

Hematocrit values in patients with SVC-RPA shunt. (a) In five patients who did not develop late SVC syndrome. (b) In three patients who developed late SVC syndrome.
had open heart repair for tetralogy of Fallot. The SVC-RPA shunt was left intact in both cases.

A third patient (case 3), unlike patients 5 and 8, did not show signs of mild SVC syndrome in the early postoperative period. However, 20 months after SVC-RPA anastomosis, he also developed a late form of SVC syndrome associated with clinical deterioration. He underwent a successful left Blalock-Taussig anastomosis with the SVC-RPA shunt left intact.

In each of these three cases (cases 3, 5, and 8), marked clinical improvement and prompt disappearance of the signs of superior vena caval obstruction followed additional shunt surgery or successful open heart surgery. Exercise tolerance increased, and hematocrit and cyanosis were diminished. Severe headaches have not occurred, and there have been no episodes of upper body or facial edema.

One year after open heart repair of tetralogy of Fallot (case 8), recatheterization revealed severe pulmonic valve insufficiency, elevated mean pressure in the left pulmonary artery, and systemic systolic pressures in the right ventricle and left pulmonary artery. Left ventricular end-diastolic pressure was normal. Angiocardiography and dye curves confirmed the presence of a small residual right-to-left shunt (14% of systemic blood flow) at the ventricular level. Recatheterization performed 2 years after open heart repair revealed that the right heart and pulmonary arterial pressures had decreased. There was a small left-to-right shunt at the ventricular level, but there was no longer any right-to-left shunt.

Pressure Relationships (Table 2)

Postoperatively, the mean pressure in the superior vena cava varied from 8 to 13 mm Hg. In the two patients studied while suffering an increase in the magnitude of their superior vena cava syndrome (cases 5 and 8), mean SVC pressures were 12 and 18 mm Hg. (Pressures were probably not higher because expansion of the collateral circulation between the superior and inferior vena cava served as a safety valve for the changing hemodynamics of the SVC-RPA shunt; see below.)

There was a small pressure difference (2 mm Hg) across the anastomotic site in only one instance (case 3). The pressure differences across the right lung (RPA to left atrium) varied from 3 to 9 mm Hg, reflecting the head of pressure propelling blood across the pulmonary capillary bed. The pressure difference between the upper and lower body venous beds (RPA-IVC) varied from 3 to 11 mm Hg with the patient in the supine position.

In one patient (case 3) high oxygen saturations (83%) were recorded from the RPA even though the catheter was not in the wedge position. Catheter withdrawal behind the SVC-RPA junction showed a decrease in oxygen saturation (66%). This arterIALIZATION of blood in the RPA proximal to the pulmonary capillary bed is thought to be the result of bronchial collateral circulation.

Dye Curves

Indicator-dilution curves performed by injecting indicator (indocyanine green) into the RPA distal to the anastomosis were different from those in which the injection site was proximal to the anastomosis. Injections into the SVC proximal to the anastomosis were associated with a longer appearance time, a longer buildup time, and a decrease in peak deflection when compared to injections distal to the anastomosis. The increases in appearance time were small (1 to 3.5 sec). This is not comparable to the marked prolongation in right lung circulation time as previously reported. The fact that there is a difference in appearance or circulation time in injecting above or below the anastomosis may represent drainage of blood from the superior vena cava through venous collaterals.

Angiocardiography

Superior vena cavography was performed in seven of the eight patients in our study. Our evaluation included a study of the site of SVC-RPA anastomosis, the pulmonic arteries, the capillary phase of the angiocardiogram, status of the pulmonary veins, and type and degree of collateralization proximal to the SVC-RPA...
Figure 2
Superior vena cavogram (SVC) following SVC-RPA anastomosis in case 8. (a) Anterior view and (b) lateral view. The anterior segment of the right upper lobe and the entire middle lobe are not perfused. (b) The collateralization between the superior and inferior vena caval systems via the internal mammary (IM) and pericardiophrenic (PC) veins is well demonstrated. The superior vena cava (SVC) proximal to the site of anastomosis is dilated.

anastomosis. These examinations were performed with the patients supine.

Only one of these seven patients showed opacification of all three major segments comprising the right upper lobe. In three, the entire right upper lobe did not opacify. In the other three, absence of opacification was confined to the anterior segment of the right upper lobe. The right middle lobe was more commonly opacified than the right upper lobe. Four of the seven patients exhibited filling of all branches to the right middle lobe while in three the right middle lobe did not opacify. All five major segments of the right lower lobe opacified in each patient examined.

Collateral venous channels of varying degrees had developed in five of the seven cases studied. The right pericardiophrenic and internal mammary veins acted as collateral venous channels proximal to the SVC-RPA anastomosis in three patients while the pericardiophrenic vein was the sole collateral vessel in the remaining two. There was no correlation between degree and extent of collateralization and degree of perfusion of the right upper and middle lobes. Representative angiograms are shown in figures 2 and 3.

No patient had angiocardiographic evidence of anatomic narrowing of the SVC-RPA anastomosis. Moreover, intrapulmonary clots or emboli were not observed in any case. The pulmonary veins connected normally to the left atrium in each case. All patients with SVC-RPA anastomosis showed a decrease in velocity of flow through the right lung compared to that of normal patients whose pulmonary arteries were perfused by the right ventricle.

Discussion
Mortality after anastomosis of the distal right pulmonary artery (RPA) to the superior vena cava (SVC) most frequently occurs in infants less than 6 months of age during the immediate postoperative period. In our experience with 15 of these procedures, there were eight long-term survivors (more than 3

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years). One of the 15 patients died from intraoperative hemorrhage. Five other deaths occurred in the immediate postoperative period. Four of the patients who died were less than 7 months of age. There was one late death 16 months after surgery.

Our experience with eight long-term survivors of SVC-RPA anastomosis has been reviewed. Postoperative changes in hematocrit and arterial and inferior vena caval oxygen saturations confirmed the impression of clinical improvement. Since there were no instances of postoperative congestive heart failure, the improvement in arterial oxygenation was accomplished without unduly increasing the work of the heart.

Among patients undergoing superior vena cava-right pulmonary artery (SVC-RPA) anastomosis, previous experience1, 2, 5, 6, has shown the immediate development of some degree of superior vena caval hypertension. This occurs because superior vena caval flow encounters increased resistance after being diverted directly into the pulmonary bed. If the magnitude of the pulmonary vascular resistance is low, and left heart obstruction is absent, superior vena caval hypertension may be without obvious clinical effect (cases 1 to 4, 6 and 7). In the presence of significantly increased vascular resistance, there may be an immediate severe superior vena caval syndrome with physiologic obstruction of flow through the shunt and a fatal termination. The latter problem is seen most frequently in infants less than 6 months of age.

Between these two ends of the clinical spectrum, a mild SVC syndrome may develop early in the postoperative period as in two of our eight patients (cases 5 and 8). Significant to these cases with mild, early SVC syndrome was the gradual overall improvement in clinical condition similar to that of patients without early SVC syndrome.

Between 1½ and 2 years after SVC-RPA anastomosis, signs of SVC syndrome in three patients became markedly increased (cases 3, 5, and 8). This late form of SVC syndrome was associated with increasing cyanosis, a rising hematocrit (fig. 1b), severe headaches, and decreased exercise tolerance. All three

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**Figure 3**

Superior vena cavaogram (SVC) following SVC-RPA anastomosis in case 4. (a) Anterior and (b) lateral view. There is opacification of the entire right middle and lower lobes. The right upper lobe shows decreased flow to the posterior segment and absence of opacification of the anterior and apical segments.
patients had severe pulmonic stenosis as part of their cardiac malformation.

The occurrence of a late, yet reversible SVC syndrome in patients with pulmonic stenosis may be secondary to a decrease in blood flow to the left lung. We postulate that a relative or absolute increase in the degree of pulmonic stenosis with age is the initiating factor. This in turn leads to increased right-to-left shunting and a concomitant decrease in blood flow through the left lung. In this way, a cycle of decreasing arterial saturation, progressive polycythemia, and increased viscosity of the blood is begun. Since the pressure difference across the right lung (RPA-left atrium) is small, pulmonary vasoconstriction secondary to hypoxemia and changes in hematocrit and blood viscosity would significantly increase resistance to flow and cause an increase in pressure in the superior vena cava. Headache and increasingly severe signs of SVC syndrome follow. Dramatic relief of symptoms occurs when hypoxemia, polycythemia, and increased viscosity of the blood are improved after correction of the cardiac malformation, or the performance of a systemic-left pulmonary artery shunt. A very small pressure difference is again able to easily propel blood through the right lung.

Following open heart repair of tetralogy of Fallot in case 8, the pulmonary vascular resistance in the left lung was increased. Flow through the right lung (SVC-RPA anastomosis), however, was unimpeded. Although lung biopsies are not available, thrombotic lesions and secondary intimal fibrosis involving the distal pulmonary vessels have been described in cases of isolated pulmonic stenosis and tetralogy of Fallot. These vascular lesions are thought to be responsible for postoperative pulmonary hypertension and an unsuccessful attempt at open heart repair of tetralogy of Fallot. We postulate that the flow of blood through the right lung (SVC-RPA shunt) was always of sufficient magnitude so that the tendency to microthrombosis was less likely. In contrast, flow through the left lung gradually decreased because of increasing pulmonic stenosis.

Although the constant pressure difference between the upper and lower body venous beds (3 to 11 mm Hg) would serve as a stimulus to the development of collateral venous channels between the superior and inferior vena cava, the magnitude of the pressure difference and the degree of development of venous collateralization may be more related to changes in the resistance to flow through the right lung. The disappearance of venous collateral channels, a fall in the pressure gradient across the right lung, and a decrease in right lung circulation time with spontaneous remission of superior caval syndrome have been noted. Collateral vein formation has been found to be most striking when the pre-shunt pulmonary vascular resistance has been elevated.

No strictures have been demonstrated at the site of anastomosis. The effectiveness of the shunt has been compromised only when changes in hemodynamics have led to hypoxemia and progressive polycythemia. The presence of collateral venous channels per se have not created obvious clinical problems during the follow-up period being reported.

Angiocardiographic studies demonstrated decreased perfusion of the right upper lobe, and to a lesser extent, the right middle lobe after SVC-RPA anastomosis. This may be an exaggeration of the normal distribution of pulmonary blood flow through various parts of the lung according to the efficiency of aeration of each area. In areas with the lowest blood oxygen saturation a stimulus to constriction of the pulmonary arterioles would exist.

Repair of correctable lesions by open heart surgery may be accomplished in patients with SVC-RPA anastomosis, as previously reported. Our patient (case 8) did remarkably well after correction of tetralogy of Fallot in spite of certain findings which usually indicate otherwise. We believe that this patient’s ability to survive postoperatively and her clinical improvement are a result of the presence of the SVC-RPA shunt. In this way the patient is able to perfuse the right lung by diverting part of the systemic venous return from the right ventricle. The right ventricle is
thereby better able to cope with the volume and pressure overload secondary to severe pulmonic insufficiency and pulmonary vascular hypertension. A previously created SVC-RPA shunt, therefore, may be of considerable hemodynamic benefit, following open heart repair, in the the presence of severe pulmonic insufficiency or a diminished capacity of the left pulmonary vascular bed.

Conceivably, an SVC-RPA anastomosis might be added as an adjunct for a patient undergoing open heart repair when severe residual pulmonic insufficiency is found. In this way, the work of the right side of the heart would be decreased. A decrease in resting cardiac output and right ventricular dilatation have been found 1 to 1½ years after the experimental production of isolated pulmonary valve insufficiency.9 SVC-RPA shunt has been used successfully in the palliative management of infants with tetralogy of Fallot and absence of the pulmonary valve.10 A SVC-RPA shunt would be contraindicated, however, if pulmonary resistance were elevated in the right lung. Determining the distal RPA pressure after proximal occlusion has been suggested as an indirect guide to the degree of pulmonary vascular resistance.2 SVC-RPA anastomosis is not performed unless the distal pulmonary artery pressure is below 10 mm Hg.

Lastly, a protocol for surgical management of the late form of SVC syndrome seems appropriate. The first step is to rule out an anatomic or mechanical cause (strictured or thrombosed anastomotic site). The presence of an associated persistent left superior vena cava should be determined. In the absence of an apparent cause, blood flow to the left lung is increased by a systemic to pulmonary artery shunt if open heart repair of the underlying lesion is not feasible. For patients with correctable lesions, open heart repair is recommended.

References
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