Circulatory Bypass of the Right Side of the Heart

VI. Shunt between Superior Vena Cava and Distal Right Pulmonary Artery; Report of Clinical Application in Thirty-eight Cases

By William W. L. Glenn, M.D., Nelson K. Ordway, M.D., Norman S. Talner, M.D., and Edward P. Call, Jr., M.D.

IT HAS been more than 10 years since our first successful anastomosis of the side of the superior vena cava to the distal end of the right pulmonary artery in a dog.¹ The dog is still alive, in fact is robust and leading an active life in a domestic environment. In the 10 years following, several hundred experimental anastomoses have been performed. Results of these have been reported recently.² From this experimental work it has become evident that partial bypass with the superior vena cava is preferable to that with the inferior vena cava. Total right heart bypass has never resulted in prolonged survival in animals and probably is not practical in man.

More than 6 years have elapsed since our first successful superior vena cava-pulmonary artery anastomosis in a patient.³ Since operation, the patient, a 7-year-old boy with transposition of the great vessels and pulmonary stenosis, has been free from complaints, except for a mild cyanosis increasing on vigorous exercise, and has been leading a normal life.

The cava-pulmonary artery anastomosis has been used in 38 patients in our clinic. All patients had severe malformation of the right side of the heart for which no established corrective procedure was available. It is the purpose of this present paper to report the postoperative results and show our progress with the clinical application of the procedure, not to offer a definitive evaluation that must await further operative experience and a longer period of follow-up examination.

Methods

Blood was obtained by arterial puncture while the subjects were at rest and in certain patients also during activity, either voluntary or while struggling and crying. The use of 100 per cent oxygen eliminated any possible unsaturation due to ventilatory factors, an essential consideration in those infants who received thiopental anesthesia to ensure a resting state during sampling.

Arterial oxygen was determined directly by a modified Roughton-Scholander technic in the early years of the study, while more recently it has been calculated from arterial P'O₂ utilizing appropriate pH correction factors and oxygen capacity.

The shunt calculations require, in addition to arterial oxygen, pulmonary capillary and mixed systemic venous oxygen contents. Pulmonary capillary oxygen has been calculated as the volume expressing the normal percentage of oxygen capacity plus appropriate dissolved oxygen. Mixed venous oxygen content has been arbitrarily assumed to be 4 ml./100 ml. less than arterial.

Only data from samples removed during the resting state have been used in the calculation of shunts. Net right-to-left shunt was calculated from appropriate mixing equations. Net right-to-left shunt is defined as the fraction of mixed systemic venous blood that regains the systemic (arterial) circulation without passing through the lungs. Inasmuch as oxygen uptake was not determined, net right-to-left shunt has been expressed as a fraction of systemic blood flow.

Indications

Superior vena cava-right pulmonary artery anastomosis has been applied to eight different cardiac conditions characterized by malfunction of the right side of the heart and diminished flow to the lungs. These are listed in Table I and constitute the operative series described below. This operation is applicable to...
any other condition in which there is intra-cardiac mixing and reduced blood flow to the lungs, such as truncus arteriosus with congenital or surgically produced pulmonary stenosis.

Table 1
Indications for Cava-pulmonary Artery Anastomosis

1. Tricuspid atresia
2. Defective development of right ventricle with intact ventricular septum
3. Tetralogy of Fallot (certain cases)
4. Corrected transposition of great vessels with pulmonary stenosis
5. Transposition of the great vessels with ventricular septal defect and pulmonary stenosis
6. Single ventricle with rudimentary outflow to pulmonary artery
7. Origin of both great vessels from right ventricle with pulmonary stenosis
8. Ebstein's anomaly

Technic of Operation

Because the operative procedure has been described in detail elsewhere, it is described here only briefly with emphasis on particular points (fig. 1).

The incision generally employed is a right anterolateral thoracotomy in the fourth interspace with the right side of the patient's body somewhat elevated. Although this incision provides the best exposure of the operative area, a sternal-splitting incision can also be used and allows a wider choice of operation.

The right pulmonary artery is divided close to its point of origin and its distal end anastomosed to the side of the superior vena cava. Every effort is made to incorporate the stump of the divided azygos vein in the anastomosis, and with rare exception it is possible. There is no kinking of an anastomosis made in this manner.

To prepare the cava for anastomosis the segment incorporating the azygos stump is drawn laterally by means of a ligature with long ends surrounding the central stump of the azygos. A curved, right-angled serrated Potts clamp is closed gently on the cava to occlude it partially, with

Figure 1
Technic of cava-pulmonary artery anastomosis.
care taken to avoid twisting or pressing upon the vessel, which might cause a complete blockage of its blood flow. Electroencephalographic monitoring is done continuously to assess the effect of an increase in venous pressure upon the brain; flattening of the brain waves indicates cerebral ischemia and requires the prompt release or readjustment of the partially occluding caval clamp (fig. 2).

With the cava grasped by the Potts clamp, the stump of theazygos vein is cut off nearly flush with the cava and the opening into the cava is enlarged to match in diameter that of the severed distal end of the right pulmonary artery. Back flow from the severed artery is controlled with small, curved, rubber-shod bulldog clamps placed temporarily on the artery’s first two branches; in addition to the clamps, or occasionally instead of them, an over-and-over heavy silk suture is laid around the vessels and held taut enough to control the bleeding.

The joining of the two vessels is accomplished by taking a stay stitch with a 5-0 silk suture at each end of the posterior suture line, tying each one and completing the posterior suture line with one of them. The anterior margins are joined by interrupted everting mattress stitches of 6-0 silk. The widest possible anastomosis is made. When the suture lines are completed, the clamps on the cava and pulmonary artery are released and the cava is ligated just below the anastomosis with a double ligature to prevent recanalization.

To prevent any formation of thrombi, no needle punctures above the waist are made 2 weeks preoperatively to 1 month postoperatively. Intravenous fluids and medications are given in the saphenous vein at the ankle. The patient is maintained in the head-up position (45 to 60°) for at least 1 year after operation to decrease the venous pressure in the brain.

The Clinical Series

Tricuspid Atresia

Poor results from the treatment of tricuspid atresia with the systemic artery shunt, due to cardiac failure and thrombosis of the shunt, led us to use a shunt that would increase the flow of un oxygenated blood to the lungs without increasing the work of the heart.5

Eight patients in the series, ranging in age from 7 months to 8 years at operation, had tricuspid atresia. All survived the shunting operation and are living 10 months to 4 years and 7 months after operation. All are clinically improved and have no limitation of physical activity. While they are moderately cyanotic on exercise, they are only mildly so at rest. There has been no increase postoperatively in the size of the heart; in fact, in most cases there has been a decrease. Hepatomegaly, which had been present in seven of the eight patients before operation, diminished in all but one of these after operation.

Laboratory studies, repeated at frequent intervals, have confirmed the clinical impression of sustained improvement following operation (table 2). Comparing postoperative data to preoperative, in all eight patients the net right-to-left shunt has diminished, with resultant rise in arterial oxygen saturation and fall

Figure 2

Electroencephalogram in a patient undergoing cava-pulmonary artery anastomosis. Flattening of the brain waves occurred after a total occlusion of the superior vena cava of only 21 seconds. There was no change in the arterial blood pressure or electrocardiogram during the period of caval occlusion.
in the hematocrit level. A substantial rise in venous pressure in the upper extremities has been observed in all patients.

Four patients in this group had had systemic artery-pulmonary artery anastomosis. In three (D.S., P.L., and G.P.), the shunt had thrombosed soon after its establishment. In one (G.P.), in whom it was between the ends of the left subclavian and left pulmonary artery, it was revised unsuccessfully 19 months after establishment; 7 months later, with the aid of hypothermia (34 to 32°C.), the main pulmonary artery (chosen because it was larger than the right branch) was severed at the right ventricle and anastomosed to the side of the superior vena cava by the usual method. In the fourth patient (A.F.) the right pulmonary artery had been too small to be anastomosed with safety to the superior vena cava and was anastomosed to the ascending aorta, end-to-side, to divert more unoxygenated blood to the lungs and, hopefully, to promote its growth. After 20 months it had grown considerably and the aorta-pulmonary artery shunt could be taken down and replaced by a cava-pulmonary artery shunt (fig. 3). Arterial oxygen saturation and the hematocrit level were about the same after the one shunt

**Figure 3**

Method of dealing with a pulmonary artery that is too small for a safe anastomosis to the superior vena cava. The anastomosis of the right pulmonary artery to the ascending aorta is made anterior (or posterior) to the superior vena cava. The diameter of the anastomosis should be exactly 5 mm. A few months later when the pulmonary artery has enlarged as determined by retrograde aortography, the aorta-pulmonary artery anastomosis is taken down and a cava-pulmonary artery anastomosis is made.
Tricuspid Atresia

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<tr>
<th>Patient</th>
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<th>Age at operation</th>
<th>Previous operations</th>
<th>Most recent postop. study (months)</th>
<th>Hematocrit Preop.</th>
<th>Hematocrit Postop.</th>
<th>Art. O₂ sat. (%) Preop.</th>
<th>Art. O₂ sat. (%) Postop.</th>
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<tr>
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<td>86</td>
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</table>

*Breathing 100 per cent O₂*

as after the other. The heart enlarged after the first shunt but after the cava-pulmonary artery shunt it decreased in size (fig. 4). A subsequent study of this patient 26 months after the caval anastomosis showed a fall in arterial oxygen saturation (table 2) and, by angiography, constriction of the right pulmonary artery distal to the anastomosis, re-canalization of the cava-right atrial junction and expansion of the venous connections between superior and inferior venae cavae (fig. 5). This patient also had a small patent ductus on the left side, which closed several months after the pulmonary artery shunt was made.

Defective Development of Right Ventricle with Intact Ventricular Septum

Defective development of the right ventricle characterized by a small tricuspid orifice, a tiny ventricular chamber, an intact ventricular septum, and an atretic or stenotic pulmonary valve, was present in two patients. One of these was aged 1 day and the other 6 days at operation. Both were critically ill. In the younger, who actually was moribund, the right pulmonary artery was large enough for

**Figure 4**

*Patient A. F., tricuspid atresia. Left, Preoperative roentgenogram. Arterial oxygen saturation was 80 per cent and the hematocrit level was 67 per cent. Center. Eighteen months after aorta-pulmonary artery anastomosis. Arterial oxygen saturation was 94 per cent and the hematocrit level 44 per cent. Note enlargement of heart. Right. Two months after aorta-pulmonary artery shunt was taken down and cava-pulmonary artery anastomosis made, the arterial oxygen saturation was 99 per cent and the hematocrit level 42 per cent. The heart has returned to normal size.*

_Circulation, Volume XXXI, February 1965_
### RIGHT-SIDED CIRCULATORY BYPASS

<table>
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<tr>
<th>Right-to-left shunt (% cardiac output)</th>
<th>Venous pressure, postop. (mm. saline)</th>
<th>Postoperative complications</th>
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<td>175</td>
</tr>
<tr>
<td>61</td>
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</tbody>
</table>

| 67 | 22 | 145 | Chylothorax: ligation of thoracic duct |
| 75 | 33 | 150 | Cardiac arrest during endotracheal suctioning: external massage. Recanalization |
| 60 | 47 | 150 | of SVC to right atrium: constriction of RPA |
| 58 | 24 | 210 | Facial edema; temporary paresis of right diaphragm |
| 41 | 190 |  |

Satisfactory anastomosis but the infant died before completion of the operation. In the other, a pulmonary valvulotomy was attempted, but when no flow through the valve was detected a cava-pulmonary artery shunt was made. The patient survived operation but died 2 days later with pulmonary edema and signs of congestive heart failure. The diameter of the right pulmonary artery in this instance was less than half the diameter of the superior vena cava.

**Tetralogy of Fallot**

Cava-pulmonary artery anastomosis establishes a permanent shunt and therefore is indicated in patients with tetralogy of Fallot *only* when it is believed that total repair will

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**Figure 5**

Collateral veins. Patient A. F., tricuspid atresia. Angiogram made 26 months postoperatively demonstrating the passage of dye from the superior vena cava into enlarged hemiazygos and left pericardiohernic veins. Communication between the superior vena cava and the right atrium is demonstrated on the lateral view. Also, there appears to be constriction of the right pulmonary artery distal to the origin of the truncus anterior.
Tetralogy of Fallot

<table>
<thead>
<tr>
<th>Patient</th>
<th>Date of operation</th>
<th>Age at operation (years)</th>
<th>Previous operations</th>
<th>Most recent postop. study (months)</th>
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<th>Hematocrit Postop.</th>
<th>Art. O₂ sat. (%) Preop.</th>
<th>Art. O₂ sat. (%) Postop.</th>
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<td>98</td>
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<td>Left Blalock</td>
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<td>58</td>
<td>76</td>
<td>94</td>
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<td>96</td>
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<td>9</td>
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*Breathing 100 per cent O₂.
†Postoperative study at month enclosed in parentheses.

not be possible (fig. 6).

Eight patients in this series, ranging in age from 16 months to 41 years at operation, had tetralogy of Fallot. All but one survived operation. There was one late death. Six patients are surviving 2 months to 4 years after operation, all are well but still show slight to moderate cyanosis at rest.

Laboratory studies have confirmed the clinical impression of improvement following operation (table 3). The net right-to-left shunt diminished postoperatively with resultant rise in arterial oxygen saturation and fall in hematocrit level (though in one patient (A.G.) after decreasing for 1 year after operation, it has increased). A sustained rise in upper extremity venous pressure has been noted in all but one patient (M.P.).

Corrected Transposition with Ventricular Septal Defect and Pulmonary Stenosis

Definitive repair of the valvular and septal defects associated with corrected transposition may be possible and open-heart exploration is therefore sometimes justified. There were three patients with this diagnosis in our series. Two patients, both aged 14, were moderately cyanotic and a third, aged five, was markedly so. The indications for operation were moderate exercise intolerance and physical underdevelopment. One patient (W.D.) also had complete AV dissociation and aortic insufficiency.

In one patient (D.B.) cardiotomy with the aid of cardiopulmonary bypass was carried out but the ventricular defect was found to be so large as to preclude success from closure; furthermore, complete heart block developed during the course of exploration of...
the defect and severe postvalvular pulmonary artery stenosis was found. A cava-pulmonary artery anastomosis therefore had to be made, the patient still on cardiopulmonary bypass.*

Laboratory studies (table 4) confirm the clinical impression of improvement. The net right-to-left shunt has diminished with resultant rise in arterial oxygen saturation and fall in hematocrit level. Upper extremity venous pressure has increased; in the patient with the AV dissociation, it has increased more than usual and may reflect elevated left atrial pressure.

Transposition of the Great Vessels with Pulmonary Stenosis

The operative plan for this anomaly should enable the patient to survive until the technics for a total repair have been perfected. It would be an advantage, of course, if the palliative operation could later be utilized as a feature of the total repair.6

Eight patients in the series had transposition of the great vessels, four with congenital pulmonary stenosis. One of the latter (K.M.), the first patient of our entire series, is now more than 6 years postoperative and is maintaining his initial excellent result. There were two operative deaths.

Three patients had pulmonary stenosis produced by banding the pulmonary artery 8 months to nearly 3 years before the shunting procedure. One in this group, an infant (D.M.), did not survive the shunting procedure. Three patients had an atrial septal defect created (Blalock-Hanlon technic) during the shunting operation but prior to making the cava-pulmonary artery anastomosis. One of these (P.D.), aged 4 months, who had no pulmonary stenosis and pulmonary hypertension did not survive enlargement of the atrial septal defect and cava-pulmonary artery anastomosis.

Laboratory studies (table 5) indicate that the net right-to-left shunt has diminished, with resultant rise in oxygen saturation and fall in hematocrit level. An elevated upper extremity venous pressure has been maintained by all patients since operation.

Single Ventricle with Pulmonary Stenosis

The cava-pulmonary artery shunt increases the flow of venous blood through the lungs without increasing the work of the single ventricle. Patients with this deformity who have a systemic artery-pulmonary artery shunt may die soon after operation from cardiac failure,2 or, if the shunt is a small one, eventually outgrow its maximal beneficial effect.

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*Operation performed by Dr. H. C. Stansel.
Corrected Transposition of the Great Vessels with Pulmonary Stenosis

<table>
<thead>
<tr>
<th>Patient</th>
<th>Date of operation</th>
<th>Age at operation (years)</th>
<th>Previous operations</th>
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<th>Hematocrit</th>
<th>Art. O₂ sat. (%) *</th>
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*Breathing 100 per cent O₂.

Table 5

Transposition of Great Vessels with Pulmonary Stenosis

<table>
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<th>Patient</th>
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<th>Age at operation (years)</th>
<th>Previous operations</th>
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*Breathing 100 per cent O₂.
†Patient P.D. had transposition of the great vessels but without pulmonary stenosis.

Three patients in this series, aged 2, 10, and 19 years at operation, had single ventricle with pulmonary stenosis. Indications for operation were, in the youngest, failure to thrive and, in the other two, poor exercise tolerance. All three patients were cyanotic at rest. One (Z.S.) had had several operations earlier to improve oxygenation. At 3 years of age, transposition of the great vessels was suspected and an atrial septal defect created; little, if any, improvement was noted and the following year, at 4 years of age, a subclavian artery-pulmonary artery anastomosis was made on the left; initially, there was considerable clinical improvement, but over a period of years the child failed to develop at a normal rate and though the shunt remained patent cyanosis increased. At age 10 a superior vena cava-right pulmonary artery shunt was made. Patency of the left subclavian-pulmonary artery shunt was confirmed. The pressure in the main pulmonary artery, prior to division of the right, was 14 mm. Hg.

A second patient (C.C.) had had an endo-to-side right subclavian-pulmonary artery anastomosis performed at the age of 4 which reduced cyanosis and improved exercise tolerance. Growth and development were satisfactory until adolescence, when cyanosis and clubbing of the digits became marked and fatigability increased. It became difficult for the patient to obtain employment because of his appearance and the history of congenital malformation of the heart. At the age of 19 it was decided to perform a cava-pulmonary artery shunt. Operation was performed through a median sternotomy. The presence of a single ventricle was confirmed. The pulmonary outflow tract was atretic though the main pulmonary artery and right and left branches appeared to be of normal size. The end-to-side shunt between the right subclavian and right pulmonary artery was patent. The pressure in the right pulmonary artery distal to the shunt was 15 mm. Hg with the right pulmonary artery proximal to the

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shunt occluded and 7 mm. Hg with it open. The right pulmonary artery was divided proximal to the subclavian artery-pulmonary artery shunt causing the shunt flow to be diverted entirely into the right lung. The main pulmonary artery now communicating only with the left pulmonary artery branches was divided just distal to the heart and anastomosed to the medial side of the superior vena cava. The latter was then doubly ligated just above the right atrium. Thus blood from the superior vena cava was routed to the left lung through the main pulmonary artery and its left branch (figs. 7 and 8).

Since establishment of the cava-pulmonary artery shunt cyanosis in all three patients has decreased and has remained minimal.

Laboratory studies (table 6) reveal that the net right-to-left shunt has diminished, with resultant rise in arterial oxygen saturation and fall in hematocrit level. These changes are more evident in the child (R.M.) who had no previous shunt.

### Origin of Both Great Vessels from Right Ventricle with Pulmonary Stenosis

This deformity is difficult to correct because the ventricular defect is usually large and the left ventricular outflow tract to be reconstructed lies far anterior. In one case we explored, using cardiopulmonary bypass, the defect seemed irreparable. There have been a few successful repairs reported, however, and thus actual repair should always be the first consideration.

In two patients, aged 8 and 10 years at operation, both major arterial trunks arose from the right ventricle and the pulmonary artery was stenotic. One patient (P.S.) had, in addition, supravalvular pulmonary stenosis. Both patients were underdeveloped, complained of exercise intolerance, and were moderately cyanotic.

After the establishment of a cava-pulmonary artery shunt the follow-up examinations have shown an increase in exercise tolerance and near disappearance of cyanosis. Labora-
Table 6

Single Ventricle with Pulmonary Stenosis

<table>
<thead>
<tr>
<th>Patient</th>
<th>Date of operation</th>
<th>Age at operation (years)</th>
<th>Previous operations</th>
<th>Most recent postop. study (months)</th>
<th>Hematocrit Preop.</th>
<th>Hematocrit Postop.</th>
<th>Art. O₂ sat. (%)* Preop.</th>
<th>Art. O₂ sat. (%)* Postop.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Z.S.</td>
<td>10/31/60</td>
<td>10</td>
<td>Blalock-Hanlon</td>
<td>30</td>
<td>57</td>
<td>48</td>
<td>89</td>
<td>96</td>
</tr>
<tr>
<td>R.M.</td>
<td>11/27/61</td>
<td>2</td>
<td>Left Blalock</td>
<td>25</td>
<td>50</td>
<td>40</td>
<td>78</td>
<td>99</td>
</tr>
<tr>
<td>C.C.</td>
<td>5/1/64</td>
<td>19</td>
<td>Right Blalock</td>
<td>3</td>
<td>59</td>
<td>45</td>
<td>91</td>
<td>95</td>
</tr>
</tbody>
</table>

*Breathing 100 per cent O₂.

Table 7

Origin of Both Great Vessels from the Right Ventricle with Pulmonary Stenosis

<table>
<thead>
<tr>
<th>Patient</th>
<th>Date of operation</th>
<th>Age at operation (years)</th>
<th>Previous operations</th>
<th>Most recent postop. study (months)</th>
<th>Hematocrit Preop.</th>
<th>Hematocrit Postop.</th>
<th>Art. O₂ sat. (%)* Preop.</th>
<th>Art. O₂ sat. (%)* Postop.</th>
</tr>
</thead>
<tbody>
<tr>
<td>M.W.</td>
<td>11/26/62</td>
<td>8</td>
<td></td>
<td>14</td>
<td>49</td>
<td>43</td>
<td>93</td>
<td>99</td>
</tr>
<tr>
<td>P.S.</td>
<td>1/24/63</td>
<td>8</td>
<td></td>
<td>10</td>
<td>52</td>
<td>51 (3)†</td>
<td>91</td>
<td>98</td>
</tr>
</tbody>
</table>

*Breathing 100 per cent O₂.
† Postoperative study at month enclosed in parentheses.

tory studies (table 7) have revealed that the net right-to-left shunt has diminished, with resultant rise in arterial oxygen saturation in both patients and fall in hematocrit level in one patient and no change in the other. The upper extremity venous pressure has increased in both patients and has remained elevated over the relatively short follow-up period.

Figure 7

Superior vena cava–left pulmonary artery shunt. Patient C.C., single ventricle with pulmonary atresia. A systemic artery-pulmonary artery anastomosis (A) was made on the right side when the patient was 4 years old. At the age of 19 the right pulmonary artery was divided (B) diverting the systemic artery flow entirely to the right lung and a superior vena cava-main pulmonary artery anastomosis (C) was made to divert the superior vena cava flow into the left lung.
**Right-sided Circulatory Bypass**

<table>
<thead>
<tr>
<th>Right-to-left shunt (% cardiac output)</th>
<th>Venous pressure, postop. (mm. saline)</th>
<th>Postoperative complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preop. 33</td>
<td>Postop. 31</td>
<td>105</td>
</tr>
<tr>
<td>65</td>
<td>20</td>
<td>130</td>
</tr>
<tr>
<td>44</td>
<td>24</td>
<td>230</td>
</tr>
</tbody>
</table>

**Ebstein's Anomaly**

The rationale of this operation for Ebstein's anomaly is reduction of the venous return by diversion of about one third of it, with consequent reduction of the work load of the greatly dilated, malfunctioning right atrium. This, in turn, may lower the incidence of serious arrhythmias, which commonly occur with this anomaly.

Four patients in the series had Ebstein's anomaly. In three, an atrial septal defect was present: in two of these (H.H. and E.F.) the shunt at the atrial level was right-to-left and in one (R.C.) it was bidirectional but predominantly left-to-right. One patient (R.H.) had no demonstrable shunt; this patient, aged 38, the oldest with Ebstein's anomaly in the series, had worked full time until the age of 36, when increasing fatigue caused him to give up his job. All four patients demonstrated exercise intolerance. The youngest patient (H.H.), aged four, had sustained a hemiplegia and complete AV dissociation as a result of a cardiac catheterization and another patient (R.C.) had severely disabling flutter-fibrillation. There was one operative and one late death (table 8).

Three patients survived the shunting operation. In one case (R.C.), cardiopulmonary bypass was used as an aid in completing the cava-pulmonary artery anastomosis, closing the atrial septal defect and resecting a por-

![Venous angiogram 4 months after superior vena cava-left pulmonary artery shunt in patient C.C. showing passage of dye from the superior vena cava on the right to the pulmonary artery on the left. The dye was injected into the right brachial vein.](image-url)
Ebstein's Anomaly

<table>
<thead>
<tr>
<th>Patient</th>
<th>Date of operation</th>
<th>Age at operation (years)</th>
<th>Previous operations</th>
<th>Most recent postop. study (months)</th>
<th>Hematocrit Preop.</th>
<th>Art. O2 sat. (%)* Preop.</th>
<th>Postop.</th>
</tr>
</thead>
<tbody>
<tr>
<td>H.H.</td>
<td>6/13/60</td>
<td>4</td>
<td></td>
<td>61</td>
<td>86</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>R.H.</td>
<td>6/19/61</td>
<td>37</td>
<td></td>
<td>45</td>
<td>99</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>R.C.</td>
<td>5/29/62</td>
<td>17</td>
<td></td>
<td>49</td>
<td>99</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>E.F.</td>
<td>11/18/63</td>
<td>9</td>
<td></td>
<td>38</td>
<td>97</td>
<td>100†</td>
<td></td>
</tr>
</tbody>
</table>

*Breathing 100 per cent O₂.

† Postoperative study at month enclosed in parentheses.

The postoperative changes in this group have been difficult to evaluate: objective evidence, such as increase in cardiac output or decrease in the work of the right side of the heart, is lacking. The longest follow-up is 30 months (R.H.): this patient states that his exercise tolerance is definitely improved and that he is again gainfully employed.

Discussion

Understanding of the altered circulatory dynamics that bring about improved arterial oxygen saturation following superior vena cava-right pulmonary artery anastomosis is facilitated if the patients are considered in two groups: those with the tetralogy of Fallot and those with anomalies in which there is complete intracardiac mixing of systemic and pulmonary venous blood. The relationship of preoperative and postoperative net right-to-left shunts to flow through the operative fistula is different in these two groups.

In seven patients with the tetralogy of Fallot the fall in the net right-to-left shunt ranged...
from 5 to 42 per cent after operation. The average decrement of 28 per cent corresponds closely with the figure for the fraction of systemic venous return assumed to be carried by the superior vena cava, namely, 33 per cent. This correspondence suggests that right ventricular output into the overriding aorta was lessened by an amount equal to the reduction in right ventricular inflow—i.e., the flow through the operative fistula—while output into the pulmonary artery continued undiminished.

The decrement in net right-to-left shunt achieved by operation was actually less than the flow through the operative fistula in any subject with (1) a pulmonary blood flow derived in part from extracardiac collateral channels or (2) a bidirectional shunt at the ventricular septal defect. This is so because some of the blood flowing to the lungs would be recirculating in these patients, and the fraction of recirculated blood would be increased by operation. With of course exception of the special case of transposition of the great vessels, recirculation is maximum when intracardiac mixing of systemic and pulmonary venous blood is complete.

Data are available for 11 patients with complete intracardiac mixing. These are seven patients with tricuspid atresia and four with single ventricle and pulmonary stenosis, the latter including two patients who also have transposition of the great vessels (K.M. and S.D.). In these 11 subjects the postoperative net right-to-left shunt was 2 to 45 per cent.
lower than the preoperative, the average fall being 25 per cent.

In order for these reductions in net right-to-left shunt to have been achieved, flows of greater magnitude through the operative fistula were necessary, as pointed out above. It is calculated that flow through the operative fistula in this group of patients ranged from 6 to 69 per cent of systemic flow. The average figure is 40 per cent, in close correspondence with the assumed 33 per cent contribution of the superior vena cava to systemic venous return.

The variability of individual results among the patients of these two groups presumably reflects, in addition to true variation, additive errors in the basic assumptions and variation in basal state from one examination to the next.

In both groups of patients the volume of blood flowing to both lungs preoperatively appears to have been fully accommodated by the left lung postoperatively.

The hemodynamic pathways following operation in transposition of the great vessels with four cardiac chambers, in double outlet right ventricle with pulmonary stenosis, and in corrected transposition with ventricular septal defect and pulmonary stenosis are open to speculation, but presumably more nearly resemble the tetralogy of Fallot than a defect with complete intracardiac mixing. The postoperative fall in net right-to-left shunt in seven patients representing these three anomalies was from 10 to 34 per cent, the best results being achieved in the two patients with transposition, in whom the superior vena cava-right pulmonary artery anastomosis was combined with a Blalock-Hanlon procedure so as to permit return to the right atrium of that volume of blood diverted proximally to the right lung (R.H. and M.Y.).

While the objective benefits from superior vena cava-right pulmonary artery anastomosis are considerable as measured in patients at rest, the demonstration of improvement during muscular exercise is perhaps even more striking, as illustrated by patient E.M. with tricuspid stenosis. This infant gave evidence of progressive decrease in pulmonary blood flow and exercise tolerance from birth till operation at 7 months of age. Postoperatively an angiogram demonstrated substantial flow to the right lung. The baby's arterial oxygen saturation during inhalation of 100 per cent oxygen at rest rose, however, from 89 per cent preoperatively only to 90 per cent 1 month postoperatively, figures corresponding to net right-to-left shunts of 41 and 38 per cent, respectively. But while his arterial blood studies at rest changed very little, he exhibited marked and sustained increase in exercise tolerance. His arterial oxygen saturation during air inhalation actually rose from 56 per cent during feeble resistance preoperatively to 73 per cent during vigorous and prolonged crying 1 month postoperatively. The failure to demonstrate significant improvement in arterial oxygen saturation at rest is presumably explained by progressive diminution of pulmonary blood flow through channels other than the operative shunt. The flow through these channels, though relatively large, appears to have increased little or not at all in response to muscular activity. The right pulmonary flow resulting from the surgical anastomosis, on the other hand, clearly augments pari passu with systemic blood flow during muscular exercise.

In addition to the assurance of augmented pulmonary flow with exercise, particular emphasis should be laid on two further features of this operation of considerable physiologic significance: (1) only systemic venous, not mixed arterialized and venous blood, is shunted to the lungs; (2) substantial reduction in net right-to-left shunt is achieved with no added burden on the heart. Left atrial flow is increased by an amount exactly equal to the decrement in right atrial flow. In situations of complete intracardiac mixing, ventricular flow is unchanged. In the tetralogy of Fallot, right ventricular flow decreases while left ventricular increases by the same amount.

In evaluating the shunt several questions remain to be answered. One of the most important, since many of these shunts have been made in small children, is—does the anastomosis grow? Unrestricted blood flow from the
cava into the pulmonary artery depends upon a large anastomosis growing apace with the individual. To determine the size of the anastomosis, angiography, with dye injected into the brachial vein (usually the left), to enable one to visualize flow to the inferior vena cava through the hemiazygous venous system (fig. 5) has been employed usually 1 to 2 years after operation, and is planned to be repeated usually at intervals of 5 years. Thus far no constriction of the anastomosis has been demonstrated on studies carried out as long as 5 years postoperatively, but much more time must pass before the question of growth of the anastomosis can be answered definitely. We believe the shunt should be established between the end of the pulmonary artery and the side of the superior vena cava in order to incorporate the azygous vein to take advantage of its bell-shaped opening and any favorable growth factor that may be present at the junction of the azygous vein with the superior cava.

Another important question is—how much of the superior cava flow passes directly to the inferior cava through collateral veins? A study of venous pressure measurements of the upper extremity in 31 patients in our series has revealed a small average decline over the 6-year period, from approximately 170 mm. of saline at the end of 1 year to approximately 150 mm. of saline at the end of 4 or more years after operation. This change is statistically insignificant due to the vagaries of the testing methods, but should it reflect a true trend it may signify enlargement of collateral veins to the inferior vena cava or a decrease in pulmonary vascular resistance.

Expansion of the collateral circulation between the superior and inferior venae cavae postoperatively has been demonstrated on angiograms in many patients and is particularly striking in those who at some time prior to cava-pulmonary artery anastomosis had an increase in pulmonary vascular resistance. (This increased resistance may persist for at least several years after pulmonary artery banding; in such patients creation of the shunt should be delayed as long as possible.) The expansion of collateral veins has usually involved the hemiazygous, pericardiophrenic, and internal mammary veins. Occasionally, in the small child, the superficial veins over the chest wall have been prominent. To what extent the development of these collateral veins will ultimately compromise the effectiveness of the cava-pulmonary artery shunt is not known at present, but it is certain from the clinical and laboratory evidence of a decrease in right-to-left shunt in the cyanotic patient that, for at least the follow-up period we are reporting, the majority of the blood in the superior vena cava is diverted through the cava-pulmonary artery shunt to the right lung.

Chylothorax following a cava-pulmonary artery shunt is commonly seen in the experimental animal, but not often in man. Five patients in the total series developed chylothorax. Three of these also had a moderate superior or vena caval syndrome. The two patients having the most severe chylothorax (P.L. and W.D.), however, had no superior caval syndrome on observation, which suggests that the chylothorax was due to direct injury of the thoracic duct or its branches in the area of the operative dissection. In two of the five patients the chylothorax was controlled by aspiration alone; in three patients with large collections of chyle uncontrollable by aspiration and restriction of fluids by mouth ligation of the thoracic duct was required.

The superior vena caval syndrome, as evidenced by swelling of the head, neck, and upper trunk, was seen in 15 patients or nearly one half of the operated group; it was usually mild and lasted only a few days. Upright positioning of the body relieved it in most cases. All showing this syndrome, except for the 41-year-old woman (M.P.) who had a thrombosis, were under 5 years of age.

In some patients the shunted flow will not be adequate to support growth and development, and other procedures will have to be resorted to to divert more venous blood to the lungs. In most patients it will, of course, be possible to create a systemic artery-pulmonary artery shunt to the contralateral lung. In the laboratory we have explored two other
methods of increasing the venous flow to the lungs in dogs with cava-pulmonary artery shunts. In one series of experiments the inferior vena cava was ligated below the renal veins to promote flow from the inferior to the superior vena cava. An increase in flow to the right lung was observed, though only temporarily.9 Recently, in another series of experiments, a fistula was established between artery and vein in the neck and a significant increase in flow through the cava-pulmonary artery shunt was demonstrated.10 (The AV fistula is in effect an extrathoracic Blalock shunt.) Several of these animals have now been followed for more than 2 years and continue to demonstrate an increased flow to the right lung. These results are encouraging and it is likely that this method of increasing flow through the cava-pulmonary artery shunt will find clinical application.

The operative and late deaths according to the age of the patients undergoing the shunting procedure are summarized in figure 11. A review of the six operative deaths revealed two cases of defective development of the right ventricle, at 1 day and 6 days of age; two of transposition of the great vessels, one with pulmonary stenosis from pulmonary artery banding and minimal intracardiac mixing and the other without pulmonary stenosis and with pulmonary hypertension; one of a tetralogy of Fallot with pulmonary atresia and nearly complete blockage of the ventricular septal defect; and one of Ebstein's anomaly with a hemiplegia and complete AV dissociation.

All patients except one died within 37 hours after operation with moderate to severe signs of obstruction of flow through the superior vena cava; the exception was, of course, the patient who died during operation. In three cases the pulmonary artery was small and in a fourth the artery was large enough but there was pulmonary hypertension; death in these four was probably due to cerebral edema secondary to obstruction of the venous return. From this we conclude that a small right pulmonary artery (less than one half the diameter of the superior vena cava) and a high pulmonary arterial resistance, usually on the basis of pulmonary hypertension, are definite contraindications for cava-pulmonary artery anastomosis. The fifth patient, a 1-day-old infant with hypoplasia of the right ventricle, was moribund at the time of operation. The sixth patient, with transposition and pulmonary stenosis from pulmonary artery banding and minimal intracardiac mixing, developed within 24 hours after operation a total atelectasis, unrelieved by bronchoscopy and tracheostomy, and a pleural effusion of the left side, which accentuated a severe hypoxia, causing death.

Death in two of these, in the 1-day-old child and in the child with Ebstein's anomaly, was probably not preventable. In the others certain different procedures might have proved of some benefit: temporary establishment of an aorta-pulmonary artery shunt in the 6-day-old infant with hypoplasia of the right ventricle; creation of an interatrial septal defect in addition to establishment of the shunt or an open-heart procedure in the child with tetralogy of Fallot (though correction of pulmonary atresia in a 16-month-old infant is doubtful); banding of the pulmonary artery in the child with transposition without pulmonary stenosis; and enlargement of the interatrial communication in the other patient with transposition and poor intracardiac mixing.

There were two late deaths, 13 months postoperatively in an 18-year-old with Ebstein's anomaly and 4 years postoperatively in a 44-year-old woman with tetralogy of Fallot, con-

**Figure 11**

*Superior vena cava–right pulmonary artery anastomosis. Age and mortality in 38 patients.*

*Circulation, Volume XXXI, February 1965*
genital heart block, and Stokes-Adams attacks. Both deaths were due to arrhythmias related to the patient’s basic anomaly. The cava-pulmonary artery shunt was widely patent at autopsy.

Conclusions

We have used the vena cava-pulmonary artery shunt in animals for more than 10 years and in patients for more than 6 years and can now draw the following conclusions:

1. The ideal caval pulmonary artery anastomosis is made between the side of the superior vena cava and the distal end of the right or left pulmonary artery. The vena cava is doubly ligated at its junction with the right atrium.

2. Patency of the anastomosis made as above is sustained. There has been only one case of thrombosis or closure in 38 cases and that was relieved by thrombectomy.

3. Growth of the anastomosis probably keeps pace with the adjacent vessels. Angiograms up to 5 years postoperatively have shown no evidence to the contrary.

4. Superior vena caval pressure remains moderately elevated for at least several years after operation, as shown by brachial venous pressure determinations.

5. In all patients in this series with a right-to-left intracardiac shunt there is improvement in oxygen saturation and hematocrit level following operation and in nearly all patients the initial excellent results have been sustained. In one patient 2 years after operation a reduction in blood flow through the shunt due to development of collateral channels between the two venae cavae and re-establishment of flow from the cava to the right atrium has been demonstrated and in a few others a slight reduction in blood flow through the shunt after several years is suspected. As is seen in all of the postoperative brachial vein angiograms, however, the route of upper extremity venous blood flow is still predominantly through the cava-pulmonary artery shunt.

Acknowledgment

The authors gratefully acknowledge the help of their colleagues, particularly Dr. Allan V. N. Goodyer in the Department of Medicine, Drs. Marie J. Browne and Ruth Whittenmore in the Department of Pediatrics, Drs. Richard H. Greenspan and Florencio A. Hipona in the Department of Radiology, and Drs. John E. Fenn, Andrew J. Graham, and Allan L. Toole in the Department of Surgery, for their many contributions to the diagnosis and care of the patients reported in this paper.

References


Circulatory Bypass of the Right Side of the Heart: VI. Shunt between Superior Vena Cava and Distal Right Pulmonary Artery; Report of Clinical Application in Thirty-eight Cases

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Circulation. 1965;31:172-189
doi: 10.1161/01.CIR.31.2.172

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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