730 gm. and retain 2 "enlarged spleens" and 1 that weighed 400 gm.

Excluding the cases noted above, 12 acceptable cases of thymoma associated with anemia and only 1 case of thymoma associated with pancycopenia remain.

The association of isolated anemia and thymoma seems, as Chalmers has stated, "most unlikely to be one of mere chance." This seems particularly true because the anemia in 4 cases was cured by excision of the thymoma, and in several cases the thymoma was known to be present before the anemia developed. The more-than-chance association of thymoma and pancycopenia is not established because only 1 well established case and 1 questionable case are available and the effect of thymectomy on pancycopenia is unknown. However, both diseases are so unusual that their coexistence is a striking clinical and pathological phenomenon. In our patient it is impossible to say which disease antedated the other, and no postulate of causal interrelation is warranted.

An attempt was made to correlate the microscopical type of the reported thymomas as well as could be determined from published descriptions and illustrations with the hematologic abnormality in 12 cases. Of these cases, 1 was omitted for lack of microscopical description. The remaining cases showed that 4 spindle-cell and 5 small-cell thymomas were associated with isolated anemia and that pancycopenia was associated with 1 spindle-cell thymoma.

The apparent frequency of spindle-cell thymomas in the reported cases of associated hematologic disorders appears unusual. The usual thymoma is composed of lymphocytes and epithelial cells in variable proportions. Less commonly encountered are thymomas composed of spindle cells, which are thought to arise from epithelial-cell progenitors. The available number of cases is too small to establish a significant relation between thymoma-cell type and hematologic abnormality.

**Summary**

An autopsied case of pancycopenia associated with thymoma is described, and the previously published cases of this rare clinical syndrome are reviewed. The unusual frequency of spindle-cell thymomas associated with this syndrome is discussed.

**References**


**CIRCULATORY BYPASS OF THE RIGHT SIDE OF THE HEART*†**

**IV. Shunt between Superior Vena Cava and Distal Right Pulmonary Artery — Report of Clinical Application**

WILLIAM W. L. GLENN, M.D.†

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In the first publication of this series of papers attention was called to the need of a method for the direct delivery of venous blood into the pulmonary arterial circulation. The congenital anomalies of the heart that might be remedied from this operation are characterized by malfunction of the right atrium or right ventricle or both. More specifically, the cardiac conditions that would benefit from circulatory bypass of the right side of the heart include stenosis or atresia of the tricuspid and pulmonary outflow tracts, Ebstein's anomaly, single ventricle, bilocular heart and transposition of the great vessels with an associated pulmonary valvar stenosis. Also, in certain cases of pulmonary hypertension in which the changes in the pulmonary arterioles have not become irreversible, the direct delivery of systemic venous blood into the pulmonary arterial circulation may be beneficial at some time after temporary ligation of the right pulmonary artery. Finally, bypass of the right side of the heart may be indicated when there is obstruction of the cavae where they join the heart, or when there is an abnormal insertion of either cava into the left atrium. Other literature pertinent to this problem has been reviewed previously.1-3
In the first paper 9 experiments were reported in which the superior vena cava was anastomosed to the distal end of the right pulmonary artery, and observations were made on venous pressure, oxygen saturation of the venous and arterial blood and angiography. It was evident from these early experiments that an anastomosis performed between the superior vena cava and the distal end of the right pulmonary artery usually remained patent postoperatively, and venous-pressure studies in the superior vena cava before and after the anastomosis was opened indicated that at least some of the blood from the superior vena cava passed through the anastomosis into the right pulmonary artery.

### Table 1. Preoperative Catheterization of the Right Side of the Heart.*

<table>
<thead>
<tr>
<th>Catheter Position</th>
<th>Blood Oxygen Content</th>
<th>Capacity</th>
<th>Saturation</th>
<th>Pressures (Corrected)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>vol. %</td>
<td>vol. %</td>
<td>%</td>
<td>mm. Hg</td>
</tr>
<tr>
<td>Right ventricle (mid)</td>
<td>18.7</td>
<td>67</td>
<td>82/10</td>
<td></td>
</tr>
<tr>
<td>(?) Right ventricle (mid)</td>
<td>23.4</td>
<td>85</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right atrium (low)</td>
<td>14.2</td>
<td>51</td>
<td>10/4</td>
<td></td>
</tr>
<tr>
<td>Right atrium (mid)</td>
<td>16.9</td>
<td>61</td>
<td>7/2</td>
<td></td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>17.1</td>
<td>62</td>
<td>9/6</td>
<td></td>
</tr>
<tr>
<td>Left femoral artery</td>
<td>20.7</td>
<td>75</td>
<td>88/99</td>
<td></td>
</tr>
</tbody>
</table>

*Performed by Pediatric-Cardiology Staff on 11/9/56.

In the second and third papers in this series further studies were carried out to determine longer-term effects of direct anastomosis of the vena cava to the distal pulmonary artery in the laboratory animal. A series of 75 animals was studied in which the anastomosis was made between the superior vena cava and the right pulmonary artery. Studies were also made on a second group of 46 animals, with an anastomosis made between the inferior vena cava, severed from connection with the right atrium, and the distal end of the right pulmonary artery. As a result of these experiments, it is believed that most and possibly all of the blood returning to the heart through the superior vena cava can be made to pass directly into the pulmonary circulation bypassing the right side of the heart. In this limited sense, the right side of the heart is not necessary to propel the venous return through the pulmonary circulation when the pulmonary vascular resistance is normal. Because of the regular occurrence of splanchnic venous congestion and ascites where the shunt between the inferior vena cava and pulmonary artery is made and because no greater flow of blood through the lung is obtained with this shunt than with that between the superior vena cava and right pulmonary artery, it was believed that the former shunt will probably have no practical clinical application except when there is congenital insertion of the inferior vena cava into the left atrium.

The ideal candidate for the shunt between the superior vena cava and right pulmonary artery appears to be a child or adult with cyanosis due to decreased pulmonary blood flow with normal or diminished pulmonary arteriolar resistance and with a condition that cannot be treated adequately by other methods. Such a patient was recently presented for consideration for surgery, and the shunt was advised on the basis of extensive experimental work carried out in this laboratory over a period of three and a half years. The present illness revealed that the child's birth was uneventful. In the neonatal period the color was poor, and oxygen was required. His color was described as "blue," and a "murmur" was heard at this time. After discharge from the hospital development appeared to be normal, although his color continued to be blue and this was more evident when he cried. Some difficulty in breathing was observed when he was lying flat.

Over the next 5 years the child developed and gained weight normally. Cyanosis was marked at all times, and exercise tolerance was poor. Between the 5th and 6th years a steadily increasing polycythemia was observed, and on October 19, 1956, the hemoglobin was found to be 25.2 gm. per 100 ml., the red-cell count 7,150,000, and the hematocrit 71 per cent of packed cells. An electrocardiogram, as interpreted by the Pediatric Staff, revealed right- axis deviation, normal sinus rhythm, high-peaked R waves, a vertical heart and right ventricular hypertrophy as evidenced by a high R wave in Lead V1. One month later cardiac catheterization and angiocardiography were carried out by the Pediatric Staff. The findings of the catheterization are shown in Table 1. Angiocardiography confirmed the suspicion of the existence of a single ventricle and also demonstrated a transposition of the great vessels, with decreased pulmonary blood flow. It was the consensus of members of the combined service conference that these findings represented a single ventricle, a transposition of the great vessels and a pulmonary stenosis.

Phlebotomy was done about 2 months before operation to reduce the hematocrit from 70 to 64 per cent of packed cells. No vein punctures were permitted in the upper extremities for 5 days before surgery.

At operation on February 25, 1958, an intravenous infusion was started in the long saphenous vein at the ankle. The right side of the chest was entered anteriorly through the bed of the 4th rib. The right pulmonary artery was isolated and found to be large and easily compressible. The superior vena cava was isolated within the pericardium. Cardiac arrest developed during the intrapericardial dissec-
tion but responded promptly to massage. Atrial tachycardia was controlled by digitalization. The azygos was ligated and divided. The pressure in the right pulmonary artery was found to be 270 mm. of saline solution. The artery was ligated medially and divided. Rubber-band bulldog clamps were placed on the distal branches. An anastomosis was performed between the distal end of the right pulmonary artery and the side of the superior vena cava at the level of its junction with the azygos vein (Fig. 1). The clamps to the superior vena cava, the pulmonary artery and the right lung.

Immediately after operation there was a marked improvement in the patient's color, and the cyanosis that had been quite evident before operation was now barely discernible. Angiography was begun on the 6th postoperative day.

The patient returned to the hospital 4 weeks after discharge for a postoperative superior caval angiogram and venous-pressure and oxygen-saturation studies. The angio- gram revealed the rapid passage of dye (diatrizoate sodium, 50 per cent) from the left brachial vein and superior vena cava into the distal right pulmonary artery through the anastomosis (Fig. 2). The dye rapidly passed through the lungs into the left atrium. There was no evidence of obstruction or venous distension. A venous-pressure determination was performed through a 15-gauge needle placed in the left brachial vein with the patient at rest. The pressure varied with each quiet respiration from 118 to 140 mm. of saline solution. Oxygen-saturation studies (Table 2) indicated a marked improvement during rest, with and without 100 per cent oxygen inhalation, and after exercise.

TABLE 2. Arterial Oxygen Studies before and after the Shunt.

<table>
<thead>
<tr>
<th>Period of Test</th>
<th>Femoral-Artery Saturation Before Operation</th>
<th>Femoral-Artery Saturation After Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>After exercise</td>
<td>42.8</td>
<td>71.6</td>
</tr>
<tr>
<td>With rest (20-25 min.)</td>
<td>70.7</td>
<td>86.9</td>
</tr>
<tr>
<td>After breathing oxygen for 1 min.</td>
<td>78.4</td>
<td>96.2</td>
</tr>
<tr>
<td>Hematocrit (packed red cells)</td>
<td>70.0(2 mo.)</td>
<td>47.0(2 mo.)</td>
</tr>
</tbody>
</table>

*Operation, 2/25/58.‡/14/58.‡/23/58.

**DISCUSSION**

The successful performance of the shunt in this patient is significant, for his complex problems could

*The angiogram was supervised by Dr. Richard Barach.

†Dr. N. K. Ordway made the following calculations on the basis of the calcieterization data and studies of femoral-artery oxygen saturation.

In all the calculations the arteriovenous difference of 3.8 vol. per cent observed at the time of catheterization on November 9, 1956, was used. Complete mixing of the blood in the common ventricle is assumed so that the oxygen content of systemic arterial and pulmonary arterial blood is the same at any time. With this as a basis the right-to-left shunt on November 9, 1956, was calculated to have been 38 per cent of the systemic venous return to the heart, and the pulmonary blood flow 73 per cent of the systemic blood flow. On February 14, 1956, the right-to-left shunt calculated out at 53 per cent of the systemic venous return to the heart, and the pulmonary flow at 58 per cent of the systemic blood flow. After operation the net right-to-left shunt is now 28 per cent of the systemic venous return to the heart. The amount of bypass to the right lung brought about by the anastomosis of the superior vena cava to the distal right pulmonary artery is now exactly the difference between the preoperative and postoperative net shunts, or somewhere between 29 per cent and 35 per cent of the systemic blood flow, depending on which of the preoperative figures one uses for net right-to-left shunt. This is in striking concordance with the prediction based on observation on animals. To have produced the same result with a systemic-to-pulmonary shunt procedure, one would have had to increase the pulmonary flow by a flow through the operative fistula equal to one or one and a quarter times the systemic flow, depending on which of the preoperative figures for right-to-left shunt is used. In either case, this means a pulmonary flow of about one and eight-tenths times the systemic flow, or a pulmonary flow of two and a half times the preoperative pulmonary flow, depending again on what figure is used for that. This would amount to a one-and-a-half-fold to twofold increase in output of the common ventricle as opposed to no increase in output from the common ventricle with the superior-vena-cava-pulmonary-artery shunt. A further assumption made is that the flow through the stenotic pulmonary valve to the left lung postoperatively is the same as the total flow to both lungs preoperatively. This is a reasonable assumption since the pressure difference must be about the same because the return to the common ventricle is also the same.

If this operation is applied to a patient with intact ventricular septum such as Epstein's anomaly, the right ventricular output would be reduced by about 29 to 35 per cent whereas the left would not be altered.
not, at least with present knowledge, be corrected by open-heart technics. Also, the use of a shunt between the systemic artery and the pulmonary artery would have had the undesirable effect of increasing the work of the single ventricle. Furthermore, because the shunt could be made as large as the diameter of the vessels would permit, it is likely, if the animal experiments can be used as a guide, that the anastomosis will remain sufficiently wide as the child grows to transmit the total return to the superior vena cava without a further significant increase in venous pressure. The anastomosis performed in this patient was identical to the first one done on a laboratory animal more than three and a half years ago. That first animal is living and well, with a widely patent anastomosis, at the present time.

It is probable that the venous flow through the shunt will not exceed 30 to 40 per cent of the total systemic venous return. But, as is evident from the postoperative clinical and laboratory studies in this patient, this amount of flow will result in marked improvement in arterial oxygenation without increasing the work of the heart. The reduction of 30 to 40 per cent of the venous return to the right side of the heart incident to the creation of the shunt may be beneficial to conditions such as Ebstein's anomaly characterized by right-sided heart failure with normal or hypotensive pulmonary-artery pressure. There was no evidence of increase in the heart size in this patient after operation—if any change took place, the heart appeared slightly smaller.

The most striking change after operation was the increase in exercise tolerance. Cyanosis, which was severe before operation, was barely discernible with the child at rest after operation. Cyanosis increased with exercise, but diminished rapidly on resting. His family noted that he now had no limitation of activity and that only on strenuous exercise was cyanosis marked. When he was asked how the operation helped him, he replied, “Now I can walk up the hill.”

The criteria for the selection of patients for the performance of the shunt cannot be too strongly emphasized. The two most important local anatomic features are a pulmonary artery and superior vena cava of large size and a normal or decreased resistance to flow in the pulmonary circulation.

The gratifying result obtained in this first patient encourages me to apply this technic to other patients when there is strict adherence to the criteria for selection and management. Remarkable as this operative procedure is, from the standpoint both of the alteration of the basic circulatory pattern and of the improvement of the arterial oxygen saturation in the cyanotic patient, the possibly fatal consequence of thrombosis of the anastomosis must not be forgotten and no effort spared to avoid this complication.

**SUMMARY AND CONCLUSIONS**

On the basis of experimental work carried out over a period of three and a half years, the clinical application of anastomosis between the superior vena cava and the distal right pulmonary artery was successfully accomplished. The patient was a seven-year-old boy with a single ventricle, transposition of the great vessels and pulmonary stenosis.

Clinical studies and laboratory tests indicate significant improvement in arterial oxygen saturation and exercise tolerance since operation. Attention is called particularly to the strict criteria for the selection of patients for this operative procedure, and the exceptional precautions taken to prevent thrombosis at the site of anastomosis. For a successful shunt of this type, it is believed to be essential to have a pulmonary artery and superior vena cava of large size and a normal or low resistance to flow through the pulmonary circulation.

A number of congenital malformations of the heart, not previously amenable to adequate treatment, may be benefited by a shunt between the superior vena cava and pulmonary artery.

It is due to the experimental contributions of my surgical colleagues, Drs. J. F. Patoño, S. B. Nuland, P. H. Guilfoil and M. Hume, and Mr. J. E. Penn that this procedure in the human subject was made possible. I am also indebted to members of the Pediatric Staff, Drs. H. S. Harned, R. Whitemore, R. J. Waters, R. Sunico and N. K. Ordway, for their help in the diagnosis and management of the patient reported herein.

**REFERENCES**

