Anomalous Systemic Venous Drainage with Hypoplasia of the Right Ventricular Myocardium

By H. R. Overy, B.M., M.R.C.P., P. G. Steinbicker, Jr., M.D., and S. Gilbert Blount, Jr., M.D.

Congenital anomalies of systemic venous drainage are not uncommon. The benign nature of the majority of these anomalies has been well documented and the usual course described. Most commonly, a persistent left superior vena cava drains into the coronary sinus and then into the right atrium without making connections with the pulmonary veins. Numerous examples of this condition have been reported.1-5 Rarely the left superior vena cava drains into the left atrium.1, 2, 4, 8-11 This situation may be associated with a low posterior septal defect and absence of the coronary sinus.10 Also rarely seen is the condition in which the right superior vena cava drains into the left atrium.6-7 In the present case anomalies of both systemic and pulmonary veins result in drainage of systemic blood into the left atrium with an additional complication that has not been described previously.

Report of Case

W.J.U., a 4-year-old white boy (CGH no. 179058) presented on March 20, 1964, at this institution for evaluation of cyanosis. By history he was the product of a normal dizygotic twin pregnancy and uncomplicated delivery. Cyanosis was noted at the age of 6 weeks. Growth and development had always lagged behind that of his twin. His physical activities were diminished by exertional fatigue and dyspnea. The parents had noted increase in the cyanosis with exertion. No history of congestive failure was obtained. The patient’s five siblings, including his twin, have always appeared to be normal.

Physical examination revealed a small cyanotic young white male, weighing 33 lb. Pulse rate was 105 per minute. Positive physical findings included prominent a and v waves in the jugular venous pulse and a widely split second sound at the upper left sternal border. There was no evidence of hyperactivity of either ventricle, and no murmurs were audible. No hepatosplenomegaly was noted and the lungs were clear.

Electrocardiogram showed a leftward P vector suggesting an ectopic atrial pacemaker but was otherwise within normal limits. Chest x-ray (fig. 1) showed minimal cardiomegaly, normal pulmonary vascularity, and widening of the superior mediastinum.

Cardiac catheterization and angiocardiography were performed on June 2, 1964, via the left arm. The catheter entered the right atrium via a normally situated right superior vena cava and passed

![Figure 1](http://circ.ahajournals.org/)

Chest roentgenogram.
through an interatrial defect and later in the procedure entered a left superior vena cava or vertical vein from the left subclavian vein. Within this vessel the catheter followed a course vertically, then turned horizontally at the level of the main pulmonary artery, and entered the left atrium and a pulmonary vein in the right lung. Hydrogen curves were negative in the vertical part of the course and positive in the horizontal. These findings were interpreted as indicating downward flow of systemic venous blood in the left superior cava or vertical vein into the left upper lobe pulmonary vein. Oxygen saturation data (table 1) demonstrated a flow of desaturated systemic venous blood into the left atrium. Positive hydrogen curves in the right atrium and distally to the pulmonary artery without oxygen step-up indicated only an insignificant left-to-right shunt at the atrial level. Variations in systemic oxygen saturation, right atrial pressure, and end-diastolic right ventricular pressure were noted during catherization apparently with minor changes in physical activity of the child. The right atrial pressure rose from 4/0 when the child was asleep, to 10/0 when he was awake and slightly restless. Angiograms (fig. 2) demonstrated an anomalous venous connection from the junction of the left subclavian and jugular veins which initially descended vertically on the left then crossed horizontally and entered the left atrium in its right upper quadrant. In addition angiocardiography from the inferior vena cava (fig. 2) showed considerable flow of contrast medium from the right to left atrium.

On June 4, 1964, on cardiopulmonary bypass, it was found that the left superior vena cava or vertical vein joined the left upper lobe pulmonary vein. The vertical vein was ligated and divided immediately above this junction. A 2 by 2.5 cm secundum type interatrial septal defect was closed with running silk sutures. The coronary sinus was normally situated, and no abnormality of the tricuspid valve was noted. The right ventricle was inspected after closure of the atrial septal defect and ligation of the vein. It appeared to be normal in size but thin walled, especially over the infundibulum where contraction was definitely weak.

The immediate postoperative course was uncomplicated. However, on June 15, 1964, 11 days after operation, when the patient had been home for 3 days, he was readmitted because of lethargy, abdominal swelling, and peripheral edema. Physical examination revealed elevated jugular venous pressure, ascites, hepatomegaly, large bilateral pleural effusions, and a prominent third heart sound at the cardiac apex. The patient was digitalized, given diuretics, and responded with a 4½-lb. loss of weight. Thoracentesis removed the bulk of the pleural fluid which had a specific gravity of 1.012 and was sterile. Right heart catheterization was performed on this admission with hydrogen curves which revealed no evidence of left-to-right shunt. The resting cardiac output was low as shown by the A-V difference of 5.36

Table 1

Cardiac Catherization Data on 4-Yr.-Old Male

<table>
<thead>
<tr>
<th>Catheter course</th>
<th>RSVC</th>
<th>RA</th>
<th>LA &amp; LV</th>
<th>PV</th>
<th>RV &amp; PA</th>
<th>Hydrogen</th>
<th>Pressure, mm Hg</th>
<th>O2 sat., %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Also descended</td>
<td>LSVC</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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</table>

<table>
<thead>
<tr>
<th></th>
<th>RSVC</th>
<th></th>
<th>4/0-10/0</th>
<th>29/0-26/0-7</th>
<th>23/12</th>
<th>96</th>
</tr>
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<tbody>
<tr>
<td>RV</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>PA</td>
<td>+</td>
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<td>PV</td>
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<tr>
<td>LV</td>
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</table>

<table>
<thead>
<tr>
<th>A-V difference, vol %</th>
<th>Pulmonary</th>
<th>Systemic</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>8.21</td>
<td>4.13</td>
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</table>

<table>
<thead>
<tr>
<th>Flow, L/min/m²</th>
<th>Pulmonary</th>
<th>Systemic</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>2.16</td>
<td>4.15</td>
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</table>

Abbreviations: RSVC and LSVC = right and left superior vena cava; RA and LA = right and left atrium; RV and LV = right and left ventricle; PA and PV = pulmonary artery and pulmonary vein; IVC = inferior vena cava.
volumes per cent and the right ventricular systolic pressure was 24 with an end diastolic pressure of 7 to 12 reflecting right heart failure. Brachial arterial oxygen saturation was 88% which is at the lower limit of normal for this altitude. The patient's condition stabilized, and he was discharged on June 30, 1964, two weeks later. At discharge, a small right pleural effusion and cardiomegaly persisted and use of digitalis, oral diuretics, and a low salt regimen was continued.

When last seen as an out-patient on January 21, 1965, the patient was asymptomatic and had no fluoroscopic evidence of any residual pleural fluid. His over-all heart size was normal and digitalis therapy has been discontinued without complications.

**Discussion**

The anatomical situation in this patient is due to anomalies of both systemic and pulmonary veins. The first part of the anomalous connection to the left atrium appears to be a left superior vena cava or vertical vein derived embryologically from the left anterior cardinal vein. The horizontal part is probably a persistent common pulmonary vein that is normally absorbed into the posterior wall of the left atrium. Therefore, the upper lobe pulmonary veins join a common channel which then opens into the left atrium. The upper lobe veins also connect with the systemic veins via the vertical vein. No connection was detected between these anomalous veins and the coronary sinus. Such a connection is unlikely to have been present because after operation the negative hydrogen curve in the pulmonary artery indicated absence of drainage of pulmonary venous blood into the right atrium.

This case illustrates the importance of the relative right and left ventricular impedance in determining systemic and pulmonary flows and the importance of an interatrial communication functioning as an "escape valve" in the presence of altered ventricular compliance. In our patient, the magnitude of right-to-left shunt is shown by the pulmonary and systemic A-V differences which indicate a pulmonary flow approximately half normal and a normal systemic output. Thus, the total right-to-left shunt was equal to half the systemic output. A significant part of this right-to-left shunt occurred through the atrial defect as shown in the angiogram (fig. 2). This shunt suggested reduced compliance of the right ventricle as the right-to-left shunt occurred without tricuspid valve stenosis.

During cardiac catheterization an abnormal rise in right atrial pressure occurred with minimal physical exertion together with a rise in the end diastolic pressure in the right ventricle. At surgery, a hypodynamic thin-walled right ventricle was noted. Closure of the atrial "escape valve" resulted in an increase of the work load on the right ventricle with the postoperative appearance of right
ventricular failure. Since the flow to the right ventricle is normally derived from the superior vena cava in fetal life, the basis of the abnormal thin-walled right ventricle in this case might be explained by the anomalous venous drainage which could have caused decreased right ventricular flow prior to, and after birth resulting in a right ventricular myocardium which was inadequate to accept a normal work load after correction of the defects. A similar argument has been advanced to explain left ventricular hypoplasia when the flow has been reduced in fetal life by premature narrowing or closure of the foramen ovale.

Hypoplasia of the right ventricular myocardium in association with anomalous drainage of the systemic veins does not appear to have been previously reported, although Tuchman and associates⁶ indicate that the chest roentgenograms in their case suggested a small right ventricle or atrium. The possibility that the hemodynamics in this case might be explained by a mild form of Ebstein’s anomaly was entertained, but this appears unlikely since at cardiac catheterization simultaneous pressure and intracardiac electrocardiographic tracings showed synchronous change in form during withdrawal of an electrode catheter from the right ventricle to right atrium. In addition at operation no anomaly of the tricuspid valve was seen. The occurrence of an inadequate right ventricle in the present case considerably complicated the postoperative course.

In their recent communication on persistent left superior cava emptying into the left atrium, Meadows and Sharp¹¹ commented that the lack of the expected correlation between the degree of peripheral unsaturation and the fraction of the total systemic venous return emptying into the left atrium eludes any certain explanation at present and that the variability of the ratios of calculated systemic to pulmonary flow (in reported cases) is a parallel problem lacking explanation. The present case suggests that the nature of the right ventricular myocardium may provide this explanation.

Summary

A case of anomalous systemic venous drainage into the left atrium with right-to-left shunt across an atrial septal defect is presented. Hemodynamic studies, observation during surgical correction of the abnormalities and the occurrence of postoperative heart failure indicated inadequacy of the right ventricular myocardium.

References

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