Total Anomalous Pulmonary Venous Drainage into the Left Innominate Vein Associated with Transposition of the Great Vessels


Total abnormal pulmonary venous drainage into the left innominate vein has been widely reported since it was appreciated that this condition could be recognized by a characteristic "cottage-loaf" cardiovascular shadow on radiologic examination. There appears to be no previous record of this relatively common congenital anomaly occurring in association with transposition of the great vessels, although one case has been reported where all the pulmonary veins entered the right atrium in a heart with complete transposition.1

The patient, a female born July 31, 1953, was blue at birth. She was first seen in June 1956, and has been under supervision since. She has complained of breathlessness on exertion and headaches, but she has never squatted. She has had attacks of faintness but has not lost consciousness. She developed normally mentally and had grown to 41 pounds at 9 years.

On examination there was always marked cyanosis, digital clubbing, and polycythemia. The heart did not appear clinically enlarged and the cardiac impulse seemed normal. The heart sounds were not considered abnormal, but the second sound at the base was loud without detectable splitting. In the pulmonary area there was a soft systolic ejection murmur, which was also audible in the left upper chest posteriorly.

Figure 1

Electrocardiogram showing clockwise rotation of a vertical heart with right ventricular hypertrophy.
ANOMALOUS PULMONARY VENOUS DRAINAGE

The electrocardiogram showed right ventricular hypertrophy (fig. 1). Radiologically the heart was within normal limits, although the upper mediastinal shadow was broad (fig. 2).

When cardiac catheterization was first carried out, the blood samples taken from the superior vena cava and all distal sites showed similar oxygen saturations. These saturations were noted to be about 10 per cent higher than samples from the inferior vena cava (table 1), which suggested that at least part of the pulmonary venous return was into the superior vena cava. An angiogram carried out from the right ventricle, at the time of this first investigation, showed that the aorta arose from this ventricle (fig. 3). There was no evidence to suggest that any blood passed from the right ventricle to the lungs.

When the right ventricular angiograms were examined together with the chest films, it was apparent that the mediastinal shadow was much broader than the aortic arch. This suggested the possibility that the wide mediastinum was due to a left superior vena cava carrying pulmonary venous blood via the left innominate vein to a dilated right superior vena cava.

Cardiac catheterization was repeated, and it was possible to pass the catheter tip from the left innominate vein into a left superior vena cava (fig. 4), and also from the right atrium through an atrial septal defect and the left atrium into the left ventricle. A series of blood samples showed that a stream of pulmonary venous blood entered the left innominate vein from the left superior vena cava (table 2).

An angiogram was recorded following a left ventricular injection. The series of films

**Figure 2**
Posteroanterior teleradiogram. There is a broad upper mediastinal shadow, but the cardiac silhouette is within normal limits.

**Figure 3**
Anteroposterior (left) and left lateral right ventricular (right) angiograms. The aorta arises from the right ventricle. There is no evidence of any blood supply to the lungs from this ventricle.
showed that the pulmonary artery arose from the left ventricle (fig. 5, left). The pulmonary veins joined to form a common pulmonary vein, which drained via a left superior vena cava into the right side of the heart at the junction of the right and left innominate veins (fig. 5, center). The later films showed that the aorta had opacified from the right ventricle (fig. 5, right).

Discussion

From the results of the cardiac catheterization and angiocardiographic studies a diagram was composed of the anatomic abnormalities (fig. 6). The right and left pulmonary veins join a common pulmonary vein behind the heart and this runs into a persistent left superior vena cava, or anomalous vertical vein, which passes upwards in front of the left pulmonary artery into the left innominate vein. The blood in the pulmonary veins and in the left superior vena cava is almost fully saturated with oxygen (table 2) and this stream of saturated blood enters the right heart through the left innominate vein. Systemic and pulmonary venous bloods mix in the right atrium and blood from this common pool passes to the systemic and pulmonary circulations. The systemic circulation is supplied by blood which passes to the right ventricle and then out into the transposed aorta. The pulmonary circulation is supplied exclusively by blood that enters the small left atrium through the atrial septal defect and passes to the transposed pulmonary artery from the left ventricle. Receiving blood from a common pool, the blood in the aorta and pulmonary artery has the same oxygen saturation.

Although it is always difficult to make accurate calculations of blood flows in patients with complicated shunts, the pulmonary ar-

### Table 1

<table>
<thead>
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<th>Sampling site</th>
<th>Oxygen saturation, per cent</th>
<th>Pressure mm. Hg</th>
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<tr>
<td>Inferior vena cava</td>
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<td>Right atrium: Low</td>
<td>63</td>
<td></td>
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<tr>
<td>High</td>
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<tr>
<td>Superior vena cava</td>
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<tr>
<td>Right ventricle</td>
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<td>84/0</td>
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<td>92/68</td>
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<td>Left atrium</td>
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</table>

### Table 2

<table>
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</thead>
<tbody>
<tr>
<td>Aorta</td>
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<tr>
<td>Right ventricle</td>
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<tr>
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<tr>
<td>Inferior vena cava</td>
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<td></td>
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<tr>
<td>Left ventricle</td>
<td>20/5</td>
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</tbody>
</table>


**Figure 4**

Posterolateral chest film showing a cardiac catheter passing from the left innominate vein through the left superior vena cava into the pulmonary vein.
teriovenous oxygen difference is about three times the systemic arteriovenous oxygen difference. Therefore the systemic blood flow is about three times the pulmonary, indicating that only about a quarter of the blood from the right atrium passes through the atrial septal defect to the left atrium. The reduced pulmonary blood flow in this patient makes a dramatic contrast with the torrential pulmonary blood flow when this sort of abnormal pulmonary venous drainage occurs without transposition of the great vessels. Undoubtedly, this difference in pulmonary blood flow explains why the pulmonary venous channels in this patient are not dilated to the extent seen without transposition of the great vessels, where there is characteristically a broad upper mediastinal shadow formed on the left by a persistent left superior vena cava and on the right by a dilated right superior vena cava.

The possibility of performing a plastic operation to direct the systemic venous blood exclusively into the left atrium and the pulmonary venous blood into the right atrium has been considered. This would leave the child with a corrected transposition and normal arterial oxygen saturations. Although

Figure 5
Anteroposterior left ventricular angiograms. The pulmonary artery arises from the left ventricle (left). The pulmonary veins join to form a common pulmonary vein which drains into a left superior vena cava (center). The blood passes from the left superior vena cava via the left innominate vein into the right superior vena cava and thence to the right ventricle and aorta (right). The left pulmonary artery is seen to arch around and behind the left superior vena cava (left). PA, pulmonary artery; LV, left ventricle; RSVC, right superior vena cava; LSVC, left superior vena cava; IV, left innominate vein; RPV, right pulmonary veins; LPV, left pulmonary veins; CPV, common pulmonary veins; RV, right ventricle; A, aorta.

Figure 6
Diagram of the anatomical abnormalities occurring in this patient with total pulmonary venous drainage into the right heart through a persistent left superior vena cava and transposition of the great vessels.

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such an operation is bound to be hazardous, it may be inevitable to prevent deterioration.

Summary

The diagnosis of a patient with total anomalous pulmonary venous drainage into the left innominate vein associated with transposition of the great vessels is described. It is suggested that the characteristic “cottage-loaf” cardiovascular shadow is absent because transposition of the great vessels leads to pulmonary oligemia, in contrast with the increased pulmonary blood flow that is present when the great vessels are not transposed with this type of abnormal pulmonary venous drainage.

References


Methodology of Science

Can we hope to be guided in the right way by experience when there exist theories (such as classical mechanics) which to a large extent do justice to experience, without getting to the root of the matter? I answer without hesitation that there is, in my opinion, a right way, and that we are capable of finding it. Our experience hitherto justifies us in believing that nature is the realization of the simplest conceivable mathematical ideas. I am convinced that we can discover by means of purely mathematical constructions the concepts and the laws connecting them with each other, which furnish the key to the understanding of natural phenomena. Experience may suggest the appropriate mathematical concepts, but they most certainly cannot be deduced from it. Experience remains, of course, the sole criterion of the physical utility of a mathematical construction. But the creative principle resides in mathematics. In a certain sense, therefore, I hold it true that pure thought can grasp reality, as the ancients dreamed.—Albert Einstein. Essays in Science. New York, Philosophical Library, Inc. 1934, p. 17.
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