Total Anomalous Pulmonary Venous Return, Surgically Corrected in Two Patients beyond 40 Years of Age*

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Two adult patients over 40 years of age with total anomalous pulmonary venous return, which was recognized preoperatively and successfully corrected surgically, are described here. The clinical diagnosis of this condition in adults may not be difficult if it is kept in mind in patients with findings of a large atrial septal defect. The associated cyanosis in a patient with a large atrial septal defect without accompanying, marked elevation of pulmonary vascular resistance or pulmonic stenosis may be a clue to this diagnosis. On cardiac catheterization, the findings of identical O₂ saturations of blood samples from various right and left chambers of the heart, with a slightly higher oxygen saturation in pulmonary artery than the systemic artery, are typical features. These were seen in both of our patients. The pulmonary angiogram, of great importance to the surgeon, is essential to delineate the exact anatomy of the anomalous veins. These two cases support previous observations that a favorable prognosis in this condition is associated with a large interatrial communication, a short common pulmonary venous trunk, and a normal pulmonary vascular resistance.

The survival of patients with total anomalous pulmonary venous return beyond infancy is uncommon.¹ ² Nevertheless, when a large atrial septal defect is also present, enough of the pulmonary venous drainage may return via the septal defect to the left side of the heart to permit normal levels of systemic blood flow and survival into adult life.² Two patients over the age of 40 years, believed to be the first of this age to be described, were found to have this condition and were successfully operated upon. Their clinical and physiologic picture most nearly resembled that of an atrial septal defect. In such patients the associated venous anomaly may not be evident at catheterization unless angiography is performed. Relative arterial oxygen saturations in the pulmonary and femoral arteries are a valuable clue to the coexistence of total anomalous pulmonary venous return to the right side of the heart. Because of the importance of the venous anomaly from the standpoint of surgical closure of the defect, the anomalous pulmonary veins must be correctly identified preoperatively.

Case Reports

A 50-year-old housewife gave a history of known heart murmur since birth and frequent respiratory infections during childhood. She had possible acute rheumatic fever at age 21 years. At age 45, she began having exertional dyspnea and intermittent chest pain. After an episode of bronchopneumonia at age 48, she was considered to be cyanotic for the first time. Following this she became increasingly breathless and had repeated respiratory infections.

Physical examination showed a normally developed but chronically ill-appearing woman with a blood pressure of 100/65 mm Hg. There was slight cyanosis of the distal extremities. The precordial impulse was diffuse and indicative of primarily right ventricular enlargement. Delay of the pulmonic closure sound was persistent during expiration, although decreased. There was a grade 2/6 ejection systolic murmur over the pulmonic area and a short, low-pitched diastolic rumble at the lower left sternal border. In addition, at the apex there was a grade 3/6 holosystolic murmur with lateral and posterior radiation.

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TOTAL ANOMALOUS PULMONARY VENOUS RETURN

Comment: This patient had been considered for many years to have rheumatic valvular disease, probably because of the mitral regurgitant murmur. Although she was then considered to have become cyanotic, the amount of right-to-left shunt at the basal state during catheterization was sufficient to lower her arterial oxygen saturation only from the 97.5 percent level in pulmonary vein to 90.0 percent in the left ventricle. The clinical findings in general were those of an atrial septal defect. She had had a predominantly left-to-right shunt for many years without developing obstructive pulmonary hypertension. Because of the anomalous pulmonary venous drainage into the coronary sinus, the conven-

Figure 1. Case 1. X-ray film of heart, posteroanterior view, shows enlargement, especially of the main pulmonary artery, increased pulmonary vascularity, and increased transverse cardiac diameter due to a combination of right ventricular enlargement and scoliosis. There is no specific suggestion of total anomalous pulmonary venous return.

The hematocrit was 52 percent. The electrocardiogram showed right axis deviation, incomplete right bundle branch block, and diffuse repolarization changes. Chest x-ray film (Fig 1) showed an appearance of cardiac enlargement and pulmonary vascularity consistent with atrial septal defect.

Cardiac catheterization on March 2, 1967 provided the measurements shown in Table 1. The increase in oxygen concentration on the right side occurred at the level of the right atrium. Hydrogen curves were positive in the right atrium and negative in the superior vena cava. The oxygen saturation in the pulmonary artery was equal to or greater than that in the left ventricle, and the pulmonary venous saturation was higher than that in the left atrium. Biplane angiograms (Fig 2 A, B) taken after pulmonary artery injection showed the entire pulmonary venous return to enter a common chamber, which drained via the coronary sinus into the right atrium. Mild mitral insufficiency because of prolapse of the posterior mitral leaflet was demonstrated by left ventricular cineangiogram. Surgery was performed because of the symptoms of exertional dyspnea and repeated respiratory infection.

At operation on March 15, 1967 a large atrial septal defect of the septum secundum variety was found in addition to the pulmonary venous anomaly. The atrial septum was repositioned to enlarge the left atrium in order to encompass the coronary drainage site, and the defect was closed, leaving a persistent drainage of the coronary sinus into the left atrium. The mitral valve did not require repair. Among several, postoperative problems were brief periods of A-V dissociation and atrial fibrillation. At a follow-up visit six weeks postopera-

Figure 2 A, B. Case 1. Frontal (top) and lateral (bottom) views of the pulmonary angiogram, showing the common pulmonary venous trunk entering the coronary sinus.
Table 1—Cardiac Catheterization Data.

<table>
<thead>
<tr>
<th>Site</th>
<th>Case 1</th>
<th>Case 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>O₂ Saturation %</td>
<td></td>
<td></td>
</tr>
<tr>
<td>O₂ Content Vol %</td>
<td></td>
<td></td>
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<tr>
<td>Pressures (mm Hg)</td>
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<tr>
<td>O₂ Saturation %</td>
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<td>O₂ Content Vol %</td>
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<tr>
<td>Pressures (mm Hg)</td>
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<tr>
<td>Right subclavian vein</td>
<td>70.5</td>
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<tr>
<td>Superior vena cava</td>
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<tr>
<td>Right atrium</td>
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<td>38/16</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>23.4</td>
<td>86.0</td>
</tr>
<tr>
<td>Pulmonary vein</td>
<td>97.5</td>
<td>18.0</td>
</tr>
</tbody>
</table>

O₂ capacity (Vol %) 23.9 20.47

O₂ consumption (ml/min/M²) 125 (assumed) 128

Blood flows (L/min/M²)
  Pulmonary (Qp) 5.5 7.5
  Systemic (Qs) 2.4 3.7

Qp:Qs 2.3:1 2:1

Shunts (L/min/M²)
  L → R 3.4 5.1
  R → L 0.3 1.3

Resistances (units)
  Pulmonary 3.8 5.6
  Systemic 33 20

Figures in parenthesis denote mean pressures.

Tional procedures of obtaining oxygen samples and hydrogen curves would not have revealed the important venous anomaly.

The patient, a 46-year-old salesman, was known to have had a murmur since childhood and intolerance for strenuous exertion for many years. During a recent hospitalization elsewhere for a bleeding peptic ulcer the presence of cardiac enlargement and an abnormal electrocardiogram prompted further evaluation.

Physical examination showed a thin, normally developed man with a blood pressure of 105/70 mm Hg and no obvious cyanosis. There was a precordial impulse of increased right ventricular activity, constant separation of the aortic and pulmonic closure sounds, and a low-grade, pulmonic ejection murmur.

The electrocardiogram (Fig 3) showed right axis deviation, right bundle branch block, and right ventricular hypertrophy. X-ray films (Fig 4 A, B) showed enlargement of the main pulmonary artery segment and its branches, and pulmonic plethora.

The results of cardiac catheterization are shown in Table 1. The increased oxygen saturation between the right subclavian vein and the superior vena cava suggested the presence of an anomalous pulmonary venous return into the superior vena cava. Oxygen saturations were almost identical in all four chambers and great vessels, although slightly higher in the pulmonary artery than in the femoral artery. Blood flow calculations made, using an assumed pulmonary venous saturation of 97 percent, showed bidirectional, predominantly left-to-right, shunts, as indicated in Table 1. Pulmonary angiograms (Fig 5 A, B) showed that all the pulmonary veins joined to form a common trunk posterior and slightly superior to the left atrium, and drained anteriorly into the superior vena cava just above its junction with the right atrium. The common trunk was only 2 cm in length and 2 cm in diameter. Surgery was recommended because of the presence of pulmonary hypertension.

At operation an atrial septal defect of the secundum variety measuring at least 4 cm in diameter was found. The anomalous, common pulmonary vein entered the superior vena cava as described above. This anomalous vein was anastomosed side-to-side with the atria, and the atrial septal defect was closed on the right side of this orifice. Postoperation he had episodes of intermittent atrial flutter-fibrillation alternating with a slow nodal rhythm. Later, because of persistent nodal bradycardia, a permanent transvenous pacemaker was implanted. Since having surgery the patient has had improved exercise tolerance.

Comment: The clinical picture in this patient was also essentially that of an atrial septal defect. Although the pulmonary artery pressure was moderately elevated, the pulmonary vascular resistance was normal, and there was no hemodynamic reason for him to have the right-to-left shunt indicated by a femoral artery oxygen saturation of 88 percent.

Discussion

The clinical spectrum of findings associated with total anomalous pulmonary venous return is very wide. It varies from a cyanotic newborn infant who dies during the first few weeks of life to a

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noncyanotic adult with minimal disability. Burroughs and associates\(^5\) first suggested that the size of the interatrial communication and the pulmonary vascular resistance may be the factors influencing the behavior of the circulation in this anomaly. The presence of increased pulmonary vascular resistance was associated with poorer prognosis in the cases of total anomalous pulmonary venous return (TAPVR) described by Gathman and Nadas.\(^1\) Darling and associates\(^4\) reviewed 80 patients with TAPVR, including 63 from the literature, and 17 of their own. They found that only 12 patients had survived infancy, of whom only three reached adulthood (ages 27, 31, 37 years). Each of these three had large interatrial defects at the time of autopsy. Later, Burroughs and Edwards\(^2\) reviewed a total of 188 patients with this anomaly. Eighty-one patients among those having a single receiving vessel or chamber, and no other critical cardiac malformation, were examined with respect to the relationship between longevity and the anatomical findings. Depending upon the length of the anomalous pathways, the patients were grouped into three categories: (a) long (infracardiac drainage); (b) intermediate (drainage via left vertical vein); and (c) short (drainage into right atrium or superior vena cava). It was clearly seen that patients with short common pulmonary venous segments and larger interatrial communications had a longer life span than those with longer common pathways and smaller interatrial communications. The oldest patient reported from the total series of 188 patients was 39 years old. Cooley and associates\(^5\) reported surgical experience in 82 patients with total anomalous pulmonary venous return. The oldest patient in their series was again 39 years old. Both of our patients had large atrial shunts and short common pathways. In one the common anomalous venous trunk joined the right atrium through the coronary sinus, and in the other it joined the superior vena cava near its junction with the right atrium.

Cyanosis also varies considerably among patients with total anomalous pulmonary venous connection. In general, patients who are cyanotic at birth die early, and those who have no or delayed cyanosis survive longer.\(^7\) Burroughs and Edwards\(^2\) noted that cyanosis appeared earlier when total anomalous pulmonary connection was associated with longer routes and smaller interatrial communications. In one of our patients, cyanosis was a very insignificant feature and appeared only occasionally on exertion, and in the other patient cyanosis was first noticed only at the age of 48 years.

All of the previously observed factors of large interatrial communication, the short common pulmonary venous trunk, normal pulmonary vascular resistance, and insignificant cyanosis associated with a favorable prognosis in TAPVR were found in these two adult patients. The influence of size of the interatrial shunt on the course of this disease has led
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Thus, in a patient surviving to adult life the clinical picture and natural history may be expected to resemble closely that of an uncomplicated atrial septal defect. If pulmonary angiography were not performed, cases such as the first in this report would not be recognized preoperatively, and a major, unexpected anatomic problem would be encountered in the operating room. One clue to the existence of this associated venous anomaly, however, is the presence of slight-to-moderate systemic arterial desaturation in the absence of severe pul-

to the use of balloon atrial septostomy as a pallia-
tive procedure in infants with total anomalous pulmonary venous return.6

The surgical outcome is related to the degree of increase in pulmonary vascular resistance, cyanosis, and age.5,6,7 Muller9 performed the first surgical correction of total anomalous pulmonary drainage. In the largest experience of Cooley's 62 cases, there were 19 deaths in 36 infants under age one, and five

FIGURE 4 A, B. Case 2. X-ray films of heart, posteroanterior (top) and right anterior oblique (bottom) views, show enlargement of the right ventricle and pulmonary artery characteristic of atrial septal defect, without evidence of total anomalous pulmonary venous drainage.

FIGURE 5 A, B. Case 2. Frontal (top) and lateral (bottom) views of the pulmonary angiogram, showing the common pulmonary venous trunk entering the superior vena cava.
monary hypertension or some other cause for right-to-left shunt. In both of our cases the systemic arterial oxygen saturation was less than that in the pulmonary artery, a circumstance which would not occur from random mixing in even a large, isolated atrial septal defect. Oxygen saturation higher in pulmonary artery than in systemic artery has been observed in cases of TAPVR with drainage into right atrium or superior vena cava, in contrast to the cases with drainage into the portal system or inferior vena cava. In cases of infradiaphragmatic anomalous pulmonary venous drainage, the systemic arterial oxygen saturation is higher than that from the pulmonary artery. This phenomenon has been explained on the basis of streaming of blood flow in the right atrium, so that the blood from the inferior vena cava is directed more towards the foramen ovale, and that from the superior vena cava or coronary sinus is directed more towards the tricuspid valve. Obliterative pulmonary arterial hypertension is estimated to occur in 14 percent of all atrial septal defect. Neither of our patients had developed that lesion. In infants, high pulmonary resistance may be related to obstruction of the pulmonary veins, such as a long segment or one passing through the diaphragm.

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Beatrice of Dante's Divine Comedy

The mysterious figure of Beatrice is, of course, of central importance in any attempt to understand Dante (1265-1321) and his works. There used to be a great deal of controversy about whether the Beatrice of the Commedia is a real woman, Beatrice Portinari of Florence, or only a sign for something else, the Church, or Divine Wisdom or Theology or the Holy Spirit. It is now generally recognized that the question in this form is wrongly conceived. Beatrice could reach Dante because she was a real woman—and just that woman. And so, in the upper reaches of the Purgatorio, she approaches as the woman Dante had known on earth: a name and a memory that tempts him as an apple does a child; as one to be found in the garden of youthful delights; then, finally, shaking his whole being as Aeneas' shooed Dido's, and burning him with Venus' ancient flame. She is, of course, only a "shadow" but Dante forgets that. Beatrice meets her erring lover, first of all, as the profoundly disturbing young woman of Florence.