Hemodynamic Study of a Case of Anomalous Pulmonary Venous Drainage

By W. Hwang, M.D., O. Prec, M.D., K. Kuramoto, M.D., S. Segall, M.D. and L. N. Katz, M.D.

Complete drainage of the pulmonary veins into the right heart is one of the rarest and most severe congenital cardiac anomalies. The X-ray studies may suggest this diagnosis if pulmonary veins enter either coronary sinus or inferior vena cava. In the case reported, right heart catheterization and angiocardiography were used and the diagnosis of persistent left superior vena cava and partial direct drainage of the pulmonary veins into right auricle was made.

Until recently our knowledge of anomalous drainage of pulmonary veins into the right heart has been drawn mainly from postmortem examinations. Among some hundred reported cases, complete drainage of the pulmonary venous blood into the right auricle or its tributaries occurred in about 35 per cent. Eight patients with such complete anomalous drainage lived for 6 months. Since it is imperative to differentiate this condition clinically from other types of cyanotic congenital heart disease which are amenable to existing surgical treatment, additional information on the diagnosis of such anomalies during life is of practical importance as well as of academic interest.

The following is a case of nearly complete pulmonary venous drainage into the left superior vena cava and right auricle diagnosed with the aid of right heart catheterization and angiocardiography.

Case Report

A 31 month old white boy, known to have congenital heart disease, was admitted for investigation in order to arrive at a more definitive diagnosis. The child had a normal birth. Shortly afterwards, a heart murmur was noted. He was cyanotic from early infancy, especially after excitement or exertion. The cyanosis had been gradually increasing in intensity. Squatting was frequent. In the three-month period prior to admission, the parents noticed that the boy's physical activity had become markedly limited and cyanosis had increased. He had had chicken pox and bronchitis at the age of 18 months. There was no history of convulsive seizures. The family history was essentially negative.

On admission he was found to be fairly well developed. Mentally he was quite alert, although he could only speak a few simple words. There was marked cyanosis of the lips and finger beds. No clubbing of the fingers or toes was present. The chest was not deformed. The lungs were clear. The heart was not enlarged to percussion; there was no precordial thrill. The second pulmonic was louder than the second aortic sound, although there was no accentuation of either. A harsh blowing systolic murmur was heard over the whole precordium with maximal intensity in the third intercostal space to the left of the sternum. Blood pressure was 118/80 and equal in both arms. Pulse rate was 88 per minute. Abdominal organs were essentially normal to palpation. There was no evidence of congestive heart failure.

Laboratory findings: The red blood cell count was 6.22 million per cu. mm. with 14.0 Gm. per cent of hemoglobin (calculated from oxygen capacity of blood). Urinalysis was normal.

Fluoroscopically, the lungs were clear; there were no abnormal parenchymal markings. The silhouette of the heart and great vessels was notable only in that there was a prominent, well-defined bulge directly below the aortic knob in the posteroanterior view (fig. 1). In the partial left anterior oblique position, continuous with this prominence and extending upward, was a paramediastinal shadow which was considered at that time to be a branch of the pulmonary artery. The hilar shadows were not remarkable. The pulsation of the pulmonary arteries were rather small.

The electrocardiogram showed abnormally tall R waves in aVR and in leads over the right precor-
Right heart catheterization was done under light anesthesia with rectal pentobarbital, and the data obtained are presented in table 1. During the procedure, as the catheter was being manipulated in the right atrium, the tip projected beyond the right border of the heart shadow (fig. 2). Analysis of the oxygen content of a blood sample taken at this point, indicated that the catheter had entered a pulmonary vein which drained directly into the right atrium. The catheter was withdrawn from the anomalous vessel and introduced into the right ventricle, but attempts to insert the catheter in the pulmonary artery were finally abandoned because of the irritability of the heart, which led to frequent premature ventricular systoles.

Analysis of the results showed: (1) Blood samples taken at two separate points in the superior vena cava had a high oxygen content (11.2 volumes per cent) as compared to that in the inferior vena cava (8.1 volumes per cent). This indicated an admixture of oxygenated blood in the superior vena cava prior to its entrance into the right atrium. In view of this fact and the abnormal shadow at the left upper paramediastinal region in the chest film (fig. 1), this strongly suggested the presence of a persistent left superior vena cava draining the left lung and eventually emptying into the right auricle via the right superior vena cava.

(2) Direct drainage from at least one pulmonary vein from the right lung into the right atrium was demonstrated by the insertion of the catheter in the anomalous vessel.

(3) The relatively small difference between the oxygen contents of the arterial blood and that of the right ventricle indicated that nearly all the pul

**Table 1.—Results of Right Heart Catheterization**

<table>
<thead>
<tr>
<th>Catheter Location</th>
<th>O2 Content (Vol.%</th>
<th>Pressure (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right ventricle</td>
<td>15.6*</td>
<td>108/5</td>
</tr>
<tr>
<td>Right auricle</td>
<td>15.5†</td>
<td>9-1</td>
</tr>
<tr>
<td>Anomalous Pulmonary Vein</td>
<td>17.1</td>
<td>12-5</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>11.2</td>
<td></td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>8.1</td>
<td></td>
</tr>
</tbody>
</table>

Arterial oxygen content: 16.6 vol. %
Arterial oxygen capacity: 18.5 vol. %
Cardiac output:
- Systemic flow: 0.94 liters/min.
- Pulmonary flow: 5.3 liters/min.
Distribution of pulmonary flow:
- To the right heart: 4.6 liters/min.
- To the left auricle: 0.7 liters/min.
Interauricular right to left shunt: 0.24 liters/min.
* Average of three blood samples from the chamber.
† Average of two blood samples from the chamber.
‡ The oxygen consumption is assumed to be 80 cc./min. as the patient was not sufficiently cooperative to give a consistent oxygen consumption test.

were interpreted as right heart strain in an electrically vertical heart.
monary drainage was into the right atrium. The oxygen contents indicate that about seven-eighths of the pulmonary venous blood drained into the right heart and one-eighth into the left auricle.

(4) Pulmonary stenosis, though not directly demonstrated by an increased pressure gradient across the orifice, was inferred from the small pulmonary arterial pulsation despite the high right ventricular systolic pressure and the high pulmonary blood flow.

(5) An interauricular septal defect was necessarily postulated as the path is essential for the maintenance of systemic flow.

Angiocardiography was done to demonstrate the presence of an interauricular septal defect and any associated anomalies in the cardiovascular system. Thirty-five cubic centimeters of 70 per cent Diodrast was injected. Figure 3 shows clearly the interauricular communication. The pulmonary arteries were found to be unusually wide and tortuous. No definite area of stenosis could be identified, partly due to an overlapping of several structures at the point where the stenosis should be expected.

**DISCUSSION**

The embryonic development of anomalous pulmonary veins has been adequately reviewed and discussed by MacManus,\textsuperscript{12} Brody\textsuperscript{3} and Brantigan.\textsuperscript{2} It is generally contended that anomalous pulmonary veins causing partial shunt of blood into the right heart may not give rise to any symptoms or signs, and are compatible with a normal life. Their presence occasionally give rise to grave consequence when the contralateral lung with normal drainage is therapeutically obliterated (thoracoplasty, lobectomy, pneumonectomy) for some underlying pulmonary disease. It is conceivable that some of the immediate accidents following pneumothorax or other collapse therapy may be due to such anomalous drainage of the lung. On the other hand, the prognosis is very grave\textsuperscript{14} when more than 50 per cent of the pulmonary blood is shunted back to the right side of the heart. When the drainage of the lungs into the right atrium is complete, with few exceptions, patients die early, either because of insufficient systemic supply or right ventricular failure. The hemodynamics involved in this anomaly merit further comment.

When all the pulmonary venous blood is returned to the right atrium, the only pathways through which the systemic circulation can be supplied would be one or more intracardiac shunts. In Brody's analysis of 105 autopsies with anomalous drainage of the lung to the right heart, 38 cases had total aberrant drainage. Aside from 5 cases of cor bilocularis, which do not strictly belong to this group, a patent foramen ovale, when this defect was looked for, was found in all cases. Only 4 had an interventricular septal defect, and 10 had a patent ductus arteriosus. Obviously, by far the most important shunt in this condition is the interauricular septal defect. When interauricular septal defects exist alone or in association with other congenital heart defects the direction of the shunt is almost always from left to right.\textsuperscript{1,7,10,16} This has been explained on the basis of the relative positions of the auricles and the greater distensibility of the right auricle.\textsuperscript{12} In cases of complete or nearly complete drainage of the pulmonary blood into the right atrium, on the contrary, the return flow to the right auricle is so much greater than that to the left side (7 times greater in the present case) that the pressure gradient between the auricles is reduced and the shunt becomes right to left.

Furthermore the pulmonary stenosis that is considered to have been demonstrated in this case elevated the end diastolic pressure in the
right ventricle. This further facilitated the shunting of blood from right to left.

The prognosis of this patient is, of course, very grave. Brody's analysis showed that in spite of its vital role the foramen ovale gradually closed in all cases. Moreover the strain on the right heart, produced both by the high volume flow and the pulmonary stenosis, must be great. The increased cyanosis in the past 3 months and the abnormally low (about 8.0 volumes per cent) systemic venous oxygen content indicates that either the interauricular communication is closing or physical activity has increased out of proportion to that which can be maintained by the amount of blood coming from the lung and that passing through the interauricular septal defect. The electrocardiogram, as presented above, confirmed the presence of right ventricular strain. A downhill course leading to early death seems inevitable, if no drastic procedure is done to correct the anomaly. Brantigan mentioned the possibility of a surgical anastomosis between the anomalous pulmonary vein and the left auricle in such cases. The production of such an anatomic correction indeed seems worth attempting as it may offer the only hope for these patients. If the anatomic structure does not lend itself to this type of correction, enlargement of the interauricular septal defect may increase the amount of systemic blood supply resulting in a physiologic correction.

A definite diagnosis of complete or partial drainage of pulmonary veins into the right heart during life was made possible only after the utilization of right heart catheterization and angiocardiography. To date 10 cases have been reported in which the diagnosis was made with one or both of these technics. Dotter and his associates reported two cases of partial drainage of the pulmonary veins into the inferior vena cava; both were diagnosed with the aid of angiocardiography and one was confirmed by right heart catheterization. Grishman and associates recently reported three similar cases diagnosed by chest x-ray and angiocardiography. Both groups of authors emphasized the diagnostic significance of the laterally convex falciform shadow in the ordinary posteroanterior chest film. In fact, the studies of all 5 reported cases were initiated because of the presence of such abnormal shadows in the lower lung fields. Bruce and his associates made the diagnosis in their two cases by direct catheterization of the anomalous veins. One of their diagnoses was later confirmed by operation and autopsy. Johnson and McRae demonstrated by catheterization what was probably an anomalous right pulmonary vein, although the possibility of the catheter having entered a hemangioma could not be completely ruled out. Courmand's and Chapman's series of catheterized cases both included one case each of such anomalous pulmonary vein.

Grishman and co-workers remarked that vascular structures have to be of a certain size and density before becoming roentgenologically demonstrable. In the case presented, the anomalous vessels were not demonstrated either in the ordinary x-ray films or by angiocardiography. But even when the latter method is successful in demonstrating one or more such vessels, the information is inadequate. Right heart catheterization, we believe, is essential in such cases. Beside confirming the diagnosis, a comparison of oxygen contents of blood from the great vessels, chambers of the heart and peripheral artery help to quantitate the amount of anomalous drainage. This approximate quantitation, as demonstrated in our case, is obviously more important in determining the prognosis and management of the patient than the mere demonstration of the presence of such an anomaly.

**Summary**

1. A case of nearly complete anomalous drainage of pulmonary veins into the right atrium and persistent left superior vena cava with associated interauricular septal defect and pulmonary stenosis diagnosed by right heart catheterization and angiocardiography is reported.
2. The hemodynamics in this anomaly, the prognosis and possible management of this patient are discussed.
3. The relative merits of catheterization and angiocardiography as diagnostic procedures for this condition are presented.
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