Anomalous Pulmonary Venous Return with Intact Atrial Septum
Diagnosis and Pathophysiology

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SUMMARY Twenty-one patients with partial anomalous pulmonary venous drainage with intact atrial septum have been studied. These include 13 patients not previously reported from our laboratories and eight patients with complete hemodynamics reported by others. Methods for identification of this abnormality and for identification of an intact atrial septum are described, including differential indicator dilution curves, catheter probing of the atrial septum and pulmonary angiography. Blood flow through anomalously draining lobes of the lung is usually higher than through normally draining lobes attributable to the higher pressure difference across the anomalous lung, right atrial pressure being uniformly lower than left atrial pressure. The pulmonary vascular resistance when "standardized" to the flow of blood normally present in different portions of the lung indicated that no significant differences existed between normally and anomalously draining lobes. Six patients had coexisting rheumatic mitral stenosis and one had congenital mitral stenosis. Its influence on the hemodynamic changes produced by PAPVD is discussed.

THE LUNGS arise from a portion of the foregut and initially pulmonary veins drain the splanchnicplexus and empty into the systemic venous system, i.e., the superior vena cava, innominate veins, inferior vena cava, portal veins, ductus venous, and coronary sinus. The veins of that portion of the splanchnicplexus that are destined to become pulmonary veins then connect with the atrial portion of the heart. If one or more of these pulmonary veins fail to connect with the left atrium, the original drainage into the systemic venous system may persist.

Partial anomalous pulmonary venous drainage (PAPVD) occurs when one or more but not all the pulmonary veins drain into the systemic venous circulation instead of into the left atrium. The abnormally draining veins are often from the whole right lung or the whole left lung but sometimes from only one or two lobes. Right anomalous pulmonary veins usually empty into the superior vena cava, occasionally into the inferior vena cava or other sites. Anomalous left pulmonary veins usually drain into a left superior vena cava and occasionally into innominate or subclavian veins or coronary sinus.1,2

PAPVD is usually associated with an atrial septal defect (ASD) and occasionally with congenital mitral stenosis.3 Rarely PAPVD is not accompanied by an ASD. The present report includes 13 cases of the latter variety which were personally observed and eight cases reported by others wherein hemodynamic findings were included.4-10 The purpose of this paper is to try to explain some of the hemodynamic consequences of PAPVD with intact atrial septum with and without associated mitral stenosis.

Material and Methods

Thirteen patients with the diagnosis of PAPVD with intact atrial septum in whom adequate catheterization data could be obtained were reviewed. This represented all patients with this syndrome studied at the Peter Bent Brigham Hospital during a 20-year period (1954-1974) and at the San Diego Naval Hospital during a 10-year period (1966-1976) (table 1). A diagnosis of PAPVD with intact septum was made only when at least two of the following three criteria were present: 1) a significant oxygen step-up at the site of termination of the anomalous vein together with a dye curve demonstrating a left-to-right shunt from one lung or from a more proximal site and a normal dye curve from the other lung; 2) clear angiographic visualization of the origin, course and termination of the anomalous vein; 3) surgical examination of the atrial septum and the anomalous vein.

Eight cases reported in the literature4-10 presented clinical and hemodynamic data in sufficient detail that they could be included in the analysis (table 1).

Pulmonary and systemic blood flows were calculated from oxygen content of samples obtained from the anomalous vein (PVO), the pulmonary artery (PAO), a systemic artery (SAO), and the superior and inferior vena cavae (SVCO, IVCO). When the anomalous vein entered the superior or inferior vena cava, samples for calculation of systemic blood flow were obtained from an area well proximal to the entrance of the anomalous vein. When no sample could be obtained for the anomalous vein, 98% saturation was assumed. Pulmonary (PBF), systemic (SBF), and left-to-right shunt (L→R) flows were calculated as follows: 

\[
PBF = \frac{3 \text{SVCO} + 1 \text{IVCO}}{4}
\]

that portion of the lungs with normal venous drainage

\[
L \rightarrow R \text{ shunt} = PBF - SBF = \text{flow through that portion of the lung with anomalous venous drainage}
\]

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Mixed venous oxygen content was calculated using the formula of Flamm et al.\textsuperscript{11} (3 SVC + 1 IVC)/4.

Oxygen consumption, intracardiac pressures, and indicator dilution curves using indocyanine green were obtained in the usual manner. Pulmonary vascular resistances (PVR) in dynes-sec-cm\textsuperscript{-5} in normally and anomalously draining lung segments were calculated as follows:

\[
PVR_{\text{normal lung}} = \frac{[\bar{P}A - \bar{P}C]}{SBF} \times 80
\]
\[
PVR_{\text{anomalous lung}} = \frac{[\bar{P}A - \bar{R}A]}{L \to R \text{ shunt}} \times 80
\]

where \(\bar{P}A\) = mean pulmonary arterial pressure, \(\bar{P}C\) = mean pulmonary capillary wedge pressure from a normally draining lung segment and \(\bar{R}A\) = right atrial mean pressure.

When resistances are in parallel, as in the different lobes of the lung, each lobe has a higher resistance than the total. A normal individual, for example, has a cardiac output of 5 L/min, a \(\bar{P}A\) pressure of 15, and a \(\bar{P}C\) of 10. The PVR is \([15 - 10]/5 \times 80 = 80\) dynes-sec-cm\textsuperscript{-5}. Assuming equal flow to all lobes of the lungs, the PVR of each of the five lobes is \([(15 - 10)/(0.2 \times 5)] \times 80 = 400\) dynes-sec-cm\textsuperscript{-5}. As the example illustrates, it is difficult to determine by the usual method whether one or more lobes with anomalous pulmonary venous drainage received more or less than the usual quota of blood flow and whether the pulmonary vascular resistance was abnormally greater or less than in the lobes with normal venous drainage into the left atrium.

An attempt has been made to standardize these segmental resistances by dividing them by the percentage of the total pulmonary blood flow which would normally go to that segment. In order for this standardization to be valid, pulmonary arterial flow is assumed to be distributed uniformly to normally and anomalously drained lung segments alike. We felt this assumption was valid since pulmonary arterial vasculature to anomalously drained lung segments appears normal in chest roentgenograms and pulmonary angiograms. Moreover, the distribution of intravenously administered \textsuperscript{99m}Tc albumin macroaggregates is uniform in the pulmonary arterial and capillary phases of patients with PAPVD (unpublished observations, S. Treves, Children's Hospital Medical Center, Boston).

Bryan and co-workers have previously estimated flow distribution to the various lobes of each lung in normal individuals in the supine position. Flow to upper zones was 36%, middle 35% and lower 29%.\textsuperscript{12} Since both lung weight and ventilation is about 54% for the right lung and 46% for the left lung, blood flow must have a similar distribution.\textsuperscript{13} Flow to the various lobes has been estimated as shown in table 2. Flows to any given lung segment (specifically the anomalously draining lobe or lobes) were standardized by dividing by the percentage of the lung so drained. This standardized flow was then inserted into the formula for pulmonary vascular resistance. These standardized PVRs were calculated as follows:

\[
\text{Flow to upper zones} = \frac{0.36 \times \text{total flow}}{0.54}
\]
\[
\text{Flow to middle zones} = \frac{0.35 \times \text{total flow}}{0.46}
\]
\[
\text{Flow to lower zones} = \frac{0.29 \times \text{total flow}}{0.46}
\]
TABLE 3. Hemodynamic Data for Patients with PAPVD

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<th>No.</th>
<th>Anom. PV (vol %)</th>
<th>PA (vol %)</th>
<th>IVC (vol %)</th>
<th>SVC (vol %)</th>
<th>Ao (vol %)</th>
<th>Anom. PV (%)</th>
<th>Ao (%)</th>
<th>O2 Content</th>
<th>O2 saturation</th>
<th>Blood Flow</th>
<th>L → R shunt (anom. lung) (L/min/m²)</th>
<th>Predicted flow thru anom. lung + (L/min/m²)</th>
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Mean ± SD

A = O2 consumption assumed.

+ predicted anomalous flow = SBF × % of lung with abnormal PV drainage.

** O2 content in PV assumed = 98%.

α = values reported are in ml/min or L/min because body surface area not available.

* = patient with mitral stenosis.

standardized PVR (normal lung segments) = 

\[ \frac{PA - PC}{SBF \times (1 - \% \text{ of lung drained anomalously})} \]

standardized PVR (anomalous lung segments) = 

\[ \frac{PA - RA}{L \rightarrow R \text{ shunt}} \times \% \text{ of lung drained anomalously} \]

An example of such a calculation in an individual with an anomalous vein from the right upper lobe to the superior vena cava follows:

\[ L \rightarrow R \text{ shunt} = 6.5 \text{ L/min}, \quad PA = 14, \quad RA = 4 \]

Standardized PVR (anomalous lung segments) = 

\[ \frac{14 - 1}{6.5} \times 80 = 23 \text{ dynes-sec-cm}^{-4}. \]

Results

Recognition of anomalous pulmonary venous drainage at the time of catheterization was made by the following maneuvers. 1) The finding of a high O2 saturation in the right heart (RA or PA) aroused suspicion of a left-to-right shunt which when assessed by withdrawal of samples from each chamber and from several levels of both venae cavae (even distal to the innominate vein) usually revealed the approximate position of the emptying site of the anomalous vein. 2) Dye curves were performed in nine of the 21 cases. In most cases these were performed sequentially with injections into left and right pulmonary arteries and sampling from a systemic artery. Curves from normally draining lungs were unremarkable while those from lungs with anomalous veins demonstrated left-to-right shunts. In patient 10 early appearance of inhaled hydrogen was noted in the inferior vena cava and throughout the right heart but not in the superior vena cava. This patient had anomalous pulmonary venous drainage from the right lung to the IVC. 3) In many instances, the catheter was introduced directly into an anomalous vein. This demonstrated the site of drainage. 4) In eleven patients, pulmonary angiography identified the location of the anomalous vein, its lung segment, and its site of emptying. It is usually impossible, however, to distinguish left atrium from right atrium as the emptying site.

Demonstration of an intact atrial septum depended on the following: 1) Probing of the septum for several minutes with a catheter introduced through a femoral vein practically assures patency or intactness of the atrial septum. Employing a catheter which enters the right atrium from the superior vena cava is not as accurate. 2) Unless angiography is performed by injection of opaque media into the left atrium — a maneuver practically never performed — intactness of the atrial septum cannot be confidently demonstrated angiographically. 3) Differential dye curves together with probing of the atrial septum invariably predicted the presence of anomalous venous drainage with intact atrial septum in the cases reported here.

The hemodynamics were in general those of an ASD except that on the diagnostic O2 run, the increase in O2 satura-
tion was usually found in either the SVC or IVC with no further increase in the RA. Pulmonary arterial blood samples demonstrated high oxygen saturations in all patients (table 3) except those with small shunts. Left-to-right shunts (i.e., through those parts of the lung with anomalous pulmonary venous drainage) varied from small (2 L/min or less in six patients) to large (5 L/min or more in seven patients) with the remainder between these two values in eight patients (table 3).

When flows were standardized for that segment of the lung with anomalous pulmonary venous drainage, it can be seen (table 3) that in the great majority of cases, pulmonary blood flow was higher than it theoretically should have been. In one patient with mitral stenosis, it was much higher (8.9 L/min versus a theoretical 1.0 L/min). In seven patients, flow through the anomalously draining lung was 2.0 L/min or more than predicted. In the remaining 14 patients there was reasonable agreement. The mean increase of flow predicted was 1.5 ± 2.1 L/min.

Right and left atrial (PC) pressures varied widely but in all cases the RA pressure was lower than the PC pressure. The mean difference was 8.1 ± 6.8 mm Hg with a range from 1 to 22 mm Hg.

Pulmonary vascular resistances as usually calculated were normal or mildly elevated in normally draining lung segments (mean 174 ± 137 dyne·sec·cm⁻¹, table 1). In anomalously draining lung segments, PVR was considerably higher in all but three cases. The mean value was 580 ± 939 dyne·sec·cm⁻¹.

Pulmonary vascular resistances standardized to correct for the size of the lung segments with normal and abnormal pulmonary venous drainage were almost the same in normally and anomalously draining lung segments. The mean for normal lung segments was 120 ± 98 dyne·sec·cm⁻¹ (range 23 to 341), and for anomalous lung segments it was 163 ± 234 dyne·sec·cm⁻¹ (range 30 to 1132).

Arterial O₂ saturation was normal in almost all cases, as it was in the anomalous pulmonary vein in nine of the 10 cases from which samples were withdrawn.

**Discussion**

The diagnosis of partial anomalous pulmonary venous return with intact septum is frequently difficult to make even at the time of cardiac catheterization. This is especially true in cases with small left-to-right shunts. Even if the shunt is large, the catheterization data are frequently interpreted as being consistent with the diagnosis of atrial septal defect. Clues to the diagnosis at that time include an oxygen step-up at the level of the venae cavae and wide disparity between right and left atrial (pulmonary capillary wedge) pressures. In the presence of an ASD, these pressures are usually identical. Differential dye curves, pulmonary angiography and inability to cross the interatrial septum with the catheter help to confirm the diagnosis. The principle of differential dye curves is that indicator dilution curves from normally draining areas of the lung appear normal while curves obtained from anomalously draining lung regions demonstrate a left-to-right shunt. Differential dye curves are usually performed with injection of dye into each pulmonary artery with sampling from a systemic artery. A curve demonstrating early recirculation (left-to-right shunt) is inscribed when dye is injected proximal to or into the lung with anomalous venous return. Curves from the other lung are normal in appearance. Injection of dye into each pulmonary artery with sampling in the right atrium demonstrates early appearance of dye following injection into the anomalously drained lung. Differential dye curves may yield a false negative result if dye is injected into the pulmonary artery beyond the pulmonary arterial branch which supplies the anomalously draining lobe.

Following indicator dilution studies, pulmonary angiog-
raphy is frequently helpful in defining the anatomical location of anomalous pulmonary venous return. However, the integrity of the atrial septum can be difficult to identify even in the presence of clear-cut differential dye curves from both main pulmonary arteries. Probing of the atrial septum with a catheter from the right femoral vein or left atrial angiography can be helpful in proving that the atrial septum is intact. A marked difference in left and right atrial pressures also constitutes hemodynamic evidence of the integrity of the atrial septum.

Therefore, in our experience, it is possible to recognize PAPVD with an intact atrial septum by cardiac catheterization. However, it requires more than routine diagnostic methodology. False positive differential dye curves have been obtained in an occasional patient with atrial septal defect. In such cases, it appears that the pulmonary veins are so situated that all the blood returning from one lung shunts across the atrial septal defect while blood returning from the other lung passes into the left heart chambers and aorta.

Considerable disagreement exists concerning the hemodynamic pathophysiology associated with PAPVD with intact atrial septum. Some authors, noting high flows through anomalously drained lung segments, have suggested that the pulmonary vascular resistance (PVR) is decreased in these regions compared with normally draining lung segments. Others have noted elevated PVRs in anomalous pulmonary venous drainage regions as compared with normally draining areas.

Our observations indicate that flow through any given segment of the lung with anomalous drainage is almost uniformly slightly greater and, in some instances with pulmonary hypertension in association with mitral stenosis, considerably greater (case 3) than expected (see table 3). Flow through a vascular bed is directly related to the pressure difference across the bed and inversely related to the pulmonary vascular resistance. The increase of flow through anomalously drained lung segments can most readily be explained by a greater pressure difference across the anomalously draining lung. Pulmonary arterial pressure is identical for both lung segments. The normally draining lung segments empty into the left atrium; the anomalously draining lung segments into the right atrium. Right atrial pressure was uniformly lower than left atrial pressure (table 3) by a mean of 8.1 mm Hg. Thus, the pressure gradient across the anomalously draining lung was greater than that across the normally draining lung.

The standardized (in relation to the size of the lung segment involved) pulmonary vascular resistance for anomalously drained lung segments was quite variable. In some individuals it appeared to be higher (in three cases by over 100 dynes-sec-cm⁻⁵) and in some it was lower (in two cases by more than 100 dynes-sec-cm⁻⁵) than in the normally draining lung but in general standardized PVRs were approximately the same for normal and anomalous lung segments. It would seem, therefore, that PVR in the intact human lung is held relatively constant despite regional differences in perfusing pressures and flows — at least in the ranges occurring in these patients.

Seven of the 21 patients reported here had coexistent mitral stenosis. Congenital mitral stenosis has been reported to occur in association with anomalous pulmonary venous drainage but in all but one of the cases reported here the lesion was rheumatic. Most of the patients with mitral valve disease reported here were studied because they were thought clinically to have Lutembacher's syndrome. These patients sought medical advice because of symptoms related to mitral stenosis rather than to anomalous pulmonary venous drainage. As in Lutembacher's syndrome, the coexistence of rheumatic mitral valve disease is coincidental.

The combination of partial anomalous pulmonary venous drainage and mitral valve disease has interesting hemodynamic implications. 1) Flow across the mitral valve (systemic cardiac output) is not increased by anomalous pulmonary venous drainage. In fact, the systemic output would tend to be somewhat lower than normal at every level of exertion. In this respect PAPVD can have a beneficial action in lessening the flow across the mitral valve and therefore the pressure in the left atrium at any given size of mitral valve orifice. 2) Right ventricular failure, as indicated by a right atrial mean pressure above 6 mm Hg was present in seven patients, four of whom had mitral stenosis. Both mitral stenosis and PAPVD act independently to increase the burden on the right ventricle: the former by pressure, the latter by flow and, if the flow is large enough, eventually by pressure. 3) Pulmonary vascular disease can be caused by both PAPVD and mitral stenosis, assuming the mechanism of the former to be the same as it is in ASD. Only two patients (8 and 12) with PAPVD without MS had any elevation of standardized PVR in the anomalously draining lung. Standardized PVR in the normally draining lungs of these patients was normal. In two patients with mitral valve disease (3 and 13), standardized PVR was elevated in the normally draining lung and normal in the lobes with PAPVD. Mean standardized PVR was not significantly different in normally as compared with anomalously draining lobes of the lung. 4) In mitral stenosis, PVR rises in most patients when the mitral valve area is about 1.0 cm² or less. At this point left atrial, pulmonary venous, and pulmonary capillary pressures are at or near 25 mm Hg at rest, i.e., at the pulmonary edema threshold. It has been postulated that elevation to the pulmonary edema level of either the pulmonary venous or possibly pulmonary capillary pressure is responsible for initiating the rise in pulmonary arteriolar resistance. However, it could be elevations in pulmonary arterial diastolic pressure that initiate increases in PVR. It was thought that these patients might give insight as to the locus of stimulation for the pulmonary arteriolar constriction because both the normally and anomalously draining lungs are subjected to the same pulmonary arterial pressure, but only the normally draining lung is subjected to the high left atrial, pulmonary venous, and pulmonary capillary pressures secondary to mitral stenosis. The findings are shown in Table 4.

As can be seen, the results are inconclusive. In patients 3

<table>
<thead>
<tr>
<th>Patient</th>
<th>Normal lung</th>
<th>PAPVD lung</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>220</td>
<td>43</td>
</tr>
<tr>
<td>14</td>
<td>336</td>
<td>1132</td>
</tr>
<tr>
<td>15</td>
<td>60</td>
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<td>171</td>
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<td>18</td>
<td>290</td>
<td>239</td>
</tr>
<tr>
<td>19</td>
<td>341</td>
<td>113</td>
</tr>
</tbody>
</table>
and 19 PVR was elevated in the normally draining lung but not in the lungs with PAPVD. In patients 14 and 18 it was elevated in each. In patients 15, 16 and 17 it was normal in the normally draining lung and elevated in the lobes with PAPVD. Thus, these studies give no clear-cut answer to the question of the location of the stimulus for the increase of PVR in mitral stenosis.

References

ELEVATED CONCENTRATIONS of serum cholesterol and triglycerides have both been implicated in the pathogenesis of coronary artery disease (CAD). They are the major risk factors for coronary atherosclerosis. The relationship between hypercholesterolemia and CAD is less well defined. A study of familial hyperlipidemia in relatives of survivors of myocardial infarctions suggested that hypercholesterolemia in combination with hypertriglyceridemia was more common in these patients than was isolated hypercholesterolemia or hypertriglyceridemia.

Although hyperlipidemia is generally recognized as a risk factor, the physician may be uncertain as to its significance in a given patient. From the results of the Framingham study, it appears that cholesterol is a risk factor for CAD over the entire range of concentrations studied. Hence the physician cannot define a single threshold above which the patient is at risk. In managing patients with hyperlipidemia,
Anomalous pulmonary venous return with intact atrial septum: diagnosis and pathophysiology.
J S Alpert, L Dexter, W V Vieweg, F W Haynes and J E Dalen

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