Anomalous Connection of Right Pulmonary Veins to Superior Vena Cava with Interatrial Communications

Hemodynamic Data in Eight Cases

By H. J. C. Swan, John W. Kirklin, Luis M. Becu, and Earl H. Wood

Anomalous connection of the right pulmonary veins to the superior vena cava associated with an interatrial communication in an unusually cephalad location has been diagnosed at cardiac catheterization in 8 patients. These patients had the clinical features seen in patients who have atrial septal defects in the region of the fossa ovalis. Differentiation was accomplished by (1) the roentgenographic position of the catheter in the right superior pulmonary vein, (2) demonstration of an abnormally high oxygen saturation of the blood in the superior vena cava, (3) the presence of a small right-to-left shunt from the superior vena cava and the absence of such a shunt from the inferior vena cava, and (4) demonstration of similar drainage of blood from the right superior pulmonary vein and superior vena cava. The syndrome is considered to be an anomaly of pulmonary venous development while the atrial septum forms normally.

RECENT advances in surgical techniques have made possible the closure of communications between the right and the left atria. Study and treatment of patients with such cardiac anomalies have shown that many varieties of interatrial communication may exist. Since the technic of closure may differ considerably according to the type of communication present, the identification of these varieties is of both practical and academic interest.

A variant of interatrial communication considered to be rare by anatomists and pathologists is the association of an anomalous connection of some or all of the veins of the right lung with the superior vena cava, or with the caudal portion of this vessel and the cephalad portion of the right atrium, and an interatrial communication located cephalad to the fossa ovalis near the site of the anomalous connection. The fossa ovalis is usually intact, but an atrial septal defect in this region may coexist. With the relatively large number of cases now reported, it is clear that this anomaly is unusual but not rare. In a series of 35 patients with interatrial communications treated surgically, Lewis and associates1 clearly recognized and described this anomaly in 3 patients and reasoned that it was present in 2 others.

Three anatomic features characterize the anomaly: (1) the defect is above the fossa ovalis and separate from it; (2) no margin of tissue is present superiorly between the defect and the superior vena cava and an incomplete one is seen posteriorly; and (3) there is an associated anomalous connection of the right superior and, at times, the inferior pulmonary veins. We found no information of hemodynamic significance on this condition in the literature.

The hemodynamic state existing in such patients may be clarified by use of the term “connection” to define an anatomic relationship and of the term “drainage” to indicate a functional or physiologic connotation.2,3 Anomalies with normal drainage of blood from the pulmonary vein to the left ventricle by anomalous pathways have been described.4 In contrast, the functional anomaly usually present in patients with large interatrial defects in the region of the fossa ovalis in whom the right pulmonary veins are normally connected to the left atrium is similar to that in patients who have interatrial communications and anomalously connected pulmonary veins. Use of “connection” and “drainage” facilitates the description of the complex functional derangements associated with many of the varieties of interatrial communication, which are not determined on the basis of anatomic malformation alone.

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This paper is concerned with hemodynamic data and their interpretation in 8 patients studied at the Mayo Clinic and considered to have anomalous connections of some or all of the right pulmonary veins to the caudal portion of the superior vena cava, with an associated “high” interatrial communication. The diagnosis in each patient was made at cardiac catheterization. Cardiotomy was carried out in 6 of these patients and the diagnosis was confirmed.

Correction of the anomalies was successfully accomplished with survival in 5 of the 6 patients just mentioned. In the sixth patient (case 6), who had severe pulmonary hypertension, the defect was not repaired because temporary occlusion of the interatrial communication at the time of operation resulted in an increase in pulmonary arterial pressure and a decrease in radial arterial pressure. This patient subsequently died but necropsy was not done. Hence, postmortem studies are not available for any of these patients. A specimen provided by one of our colleagues, Dr. J. E. Edwards, exemplifies the common variant of this anomaly (fig. 1). Hemodynamic studies were not done on this patient. The anatomic findings at the time of thoracotomy in the 6 surgical patients are summarized in table 1 according to the site of connection of the veins from the right lung, the size and location of the principal interatrial communication, and the condition of the fossa ovalis.

In the remaining 2 of the total series of 8 patients, the presence of pulmonary hypertension associated with a great increase in pulmonary vascular resistance and predominant right-to-left shunts was judged to contraindicate surgical repair.

In 4 of the 6 surgical patients (cases 2, 4, 5, and 6), the anomaly was essentially similar to that described by Lewis and associates.1 In
Table 1.—Anatomic Characteristics of Interaltrial Communication and Anomalous Connection of Right Pulmonary Veins

<table>
<thead>
<tr>
<th>Case</th>
<th>Location of cardiac connections of right pulmonary veins*</th>
<th>High interatrial communication, location and size (cm.)</th>
<th>Fossa ovalis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>SVC-RA RA</td>
<td>Posterosuperior aspect of atrium; 3 x 1 (slit)</td>
<td>&quot;Tiny hole&quot;</td>
</tr>
<tr>
<td>2</td>
<td>SVC-RA LA</td>
<td>Superior aspect of atrium; 2.5 x 2</td>
<td>Intact</td>
</tr>
<tr>
<td>3</td>
<td>Small diverticulum of left atrium</td>
<td>SVC straddled both atria and diverticulum; 2.5 (circle)</td>
<td>Intact</td>
</tr>
<tr>
<td>4</td>
<td>SVC-RA LA</td>
<td>SVC straddled right and left atria; 2.2 x 1.5</td>
<td>Patent foramen ovale</td>
</tr>
<tr>
<td>5</td>
<td>SVC-RA LA</td>
<td>From SVC caudally to the level of inferior vein connection; 4 x 3</td>
<td>Small defect</td>
</tr>
<tr>
<td>6</td>
<td>SVC-RA LA</td>
<td>Superior aspect of atrium; 1 x 0.5</td>
<td></td>
</tr>
</tbody>
</table>

*SVC-RA = junction of superior vena cava and right atrium; RA = right atrium; LA = left atrium.

case 1, the superior and inferior pulmonary veins joined to enter the heart through a common orifice at the junction of the superior vena cava with the right atrium. In case 3, a true anomalous connection did not exist; instead, the pulmonary veins all drained to a diverticulum located behind (dorsal to) the junction of the superior vena cava and right atrium. The anterior (ventral) wall of this diverticulum was absent, so that the superior vena cava appeared to straddle both right and left atria and was in free communication with each. Therefore, the only communication between the atria in this patient was by way of the lower part of the superior vena cava above the superior margin of the atrial septum. In case 5, the right superior pulmonary vein connected to the superior vena cava at a point 2 cm. cephalad to the right atrium, while the right inferior pulmonary vein connected to the left atrium. The defect in this patient was larger than in the others, and a separate communication in the fossa ovalis was also present. The principal defect involved the posterior and superior (dorsocephalad) aspect of the interatrial septum, extending 2.5 to 3 cm. caudally from the region of the superior vena cava to the level of the left atrial orifice of the inferior pulmonary vein.

Each patient was sent to the laboratory with the tentative diagnosis of an atrial septal defect. No clinical feature has been recognized that might serve to differentiate this condition from an atrial septal defect of the usual type. Clinical evidence of pulmonary hypertension was present in 3 patients.

Methods

Cardiac catheterization was carried out in the manner already described from this laboratory. Intracardiac and intravascular pressures were measured by means of strain-gage manometer-catheter systems with adequate frequency and damping characteristics. The oxygen saturation of samples of blood drawn from the chambers of the heart and from the great vessels was determined by means of a cuvette oximeter. Analysis of blood from the heart and from the radial artery as to oxygen content and oxygen capacity was carried out by the method of Van Slyke and Neill. The pulmonary (Qp) and systemic (Qs) blood flows (in liters per minute) were determined from the relations:

\[ Q_p = \frac{V_{O_2}}{C_{pv} - C_{pa}} \quad \text{and} \quad Q_s = \frac{V_{O_2}}{C_{sa} - C_{mvb}} \]

where \( V_{O_2} \) is the oxygen consumption in milliliters per minute, and \( C_{pv}, C_{pa}, C_{sa} \) and \( C_{mvb} \) represent the oxygen content of pulmonary vein, pulmonary artery, systemic artery, and mixed venous blood, respectively, in milliliters per liter. \( C_{mvb} \) was taken as the average oxygen content of superior and inferior caval blood.

The fraction of pulmonary arterial flow representing recirculated blood (\( S_{t-r} \)) is given by the relation:

\[ S_{t-r} = \frac{C_{pa} - C_{mvb}}{C_{pv} - C_{mvb}} \]

The fraction of systemic artery flow representing mixed venous blood (\( S_{r-1} \)) is given by:

\[ S_{r-1} = \frac{C_{pv} - C_{sa}}{C_{pv} - C_{mvb}} \]

Dye-dilution curves were recorded by ear and cuvette oximeters after injection of Evans blue (T-1824) or methylene blue at different locations in the heart and great vessels and are considered in detail in the following section.
RESULTS

The basic hemodynamic state in these patients is similar to that found in patients who have atrial septal defects. In this small group are examples of large pulmonary blood flow, without pulmonary hypertension, and of severe pulmonary hypertension with bidirectional shunting. It is in particular hemodynamic details that these patients differ from those who have atrial septal defects of the usual type.

Values of pulmonary flow varied from 10.6 to 2.4 L./min./M.² of body surface, while pulmonary arterial pressures correlated inversely with flow, ranging from 27/13 to 113/52, expressed as mm. Hg (table 2). The systemic blood flow was below the range of normal only in case 6. Left-to-right shunts of 67 to 24 per cent were calculated in 7 patients and right-to-left shunts of 9 to 31 per cent were found in 5 patients. These shunts were determined from data on blood flow in cases 5 through 8 (table 2); in case 4, the shunt was calculated from indicator-dilution curves after injection of T-1824 into the superior and inferior venae cavae. The evaluation of a right-to-left shunt in this situation is considered in detail later. In 1 instance (case 8), the pulmonary artery was not entered at the time of catheterization, and the wedge pressure in a pulmonary vein was not greatly increased. A considerable gradient existed in case 4 between the right ven-

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### Table 2.—Data on Intracardiac Shunts in Anomalous Connection of Right Superior Pulmonary Vein

<table>
<thead>
<tr>
<th>Case</th>
<th>Intravascular pressures, mm. Hg*</th>
<th>Blood flow (L./min./M²)</th>
<th>Intracardiac shunt, per cent†</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>LV Systemic PA PA wedge FV RA RV</td>
<td>Pulmonary Systemic L-R R-L</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>† 105/70 28/16 28/18 13/7 14/10 28/5 5/3</td>
<td>8.4 2.8</td>
<td>67 0</td>
</tr>
<tr>
<td>2</td>
<td>117/11 130/75 36/13 12/5 8/2 † 34/8 7/1</td>
<td>10.6 3.7</td>
<td>66 0</td>
</tr>
<tr>
<td>3</td>
<td>107/6 125/70 33/18 25/16 10/6 12/8 28/2 11/8</td>
<td>10.1 5.6</td>
<td>43 0</td>
</tr>
<tr>
<td>4</td>
<td>124/11 102/50‡ 27/13 † † 17/10 58/10 10/0</td>
<td>7.3 3.1</td>
<td>57 0</td>
</tr>
<tr>
<td>5</td>
<td>91/0 106/51 86/32 † † 8/0 7/3 81/-2 8/1</td>
<td>4.8 3.5</td>
<td>26 18</td>
</tr>
<tr>
<td>6</td>
<td>109/11 113/67 116/43 † † 10/4 11/9 113/6 9/4</td>
<td>2.4 2.5</td>
<td>26 31</td>
</tr>
<tr>
<td>7</td>
<td>† 122/68 113/52 † † 9/4 5/0 113/2 6/2</td>
<td>2.6 2.9</td>
<td>24 30</td>
</tr>
<tr>
<td>8</td>
<td>† † 145/84 † 23/14 8/3 † 95/8 10/3</td>
<td>† † 0 20</td>
<td></td>
</tr>
</tbody>
</table>

* LV = left ventricle; PA = pulmonary artery; PV = pulmonary vein; RV = right ventricle; RA = right atrium.
† L-R = left-to-right shunt as per cent of pulmonary flow; R-L = right-to-left shunt as per cent of systemic flow.
‡ Not obtained.
§ Systemic arterial pressure recorded 1 hour after left ventricular pressure.
Fig. 2. Roentgenograms obtained at cardiac catheterization: a and b, from case 3; c and d, from case 6. The tip of the catheter was in the right superior pulmonary vein (a and c) and in the left ventricle (b and d). Note (a and c) that the catheter entered the pulmonary vein above the main bulge of the right atrial chamber. This high location of the shaft of the catheter makes it extremely unlikely that it traversed an atrial septal defect of the usual type before entering the pulmonary vein.

In b, the catheter passes into the left ventricle; although the position of the shaft of the catheter is more cephalad than frequently seen, it could be that of a catheter traversing an atrial septal defect in the usual location (fossa ovalis). The position of the shaft of the catheter in d is more typical for that of a catheter entering the left ventricle via an interatrial communication at the fossa ovalis.
tip persistently entering either a pulmonary vein or the left atrium and ventricle.

Oxygen Saturation Values

The oxygen saturation of radial arterial blood was within the range of normal in cases 1, 2, 3, and 5, minimally reduced in cases 4 and 8 and significantly depressed in cases 6 and 7 (table 3). The saturation of pulmonary arterial blood was increased, as is consistent with the presence of the left-to-right shunts, and was always less than that of systemic arterial blood. The saturation of blood from the pulmonary vein was determined by cuvette oximeter in every instance except case 5; however, 2 patients (cases 4 and 7) were breathing 100 per cent oxygen and the values of 100 per cent that were obtained do not indicate the quantity of oxygen present in physical solution. In the remaining cases, the patients were breathing room air and the saturation values of 98 or 99 per cent exceeded the saturation of blood from the radial artery in every instance, although in cases 1, 2, and 3 the saturation of radial arterial blood was within the range of normal.

With regard to the right side of the heart, blood of the highest saturation was obtained from the lower part of the superior vena cava.

Table 3.—Average Oxygen Saturation of Blood in Great Vessels and Cardiac Chambers in Anomalous Connection of Right Superior Pulmonary Vein

<table>
<thead>
<tr>
<th>Case</th>
<th>Blood oxygenation, per cent*</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>IVC</td>
</tr>
<tr>
<td>1</td>
<td>73</td>
</tr>
<tr>
<td>2</td>
<td>77</td>
</tr>
<tr>
<td>3</td>
<td>†</td>
</tr>
<tr>
<td>4</td>
<td>78</td>
</tr>
<tr>
<td>5</td>
<td>81</td>
</tr>
<tr>
<td>6</td>
<td>45</td>
</tr>
<tr>
<td>7</td>
<td>67</td>
</tr>
<tr>
<td>8</td>
<td>66</td>
</tr>
</tbody>
</table>

* IVC = inferior vena cava; MRA = midportion of right atrium; RA-SVC = right atrial-superior caval junction; SVC = superior vena cava.
† Samples unsuitable or not obtained.
‡ Site not entered at catheterization.
§ Saturation of right ventricular blood; pulmonary artery not entered in this patient.

In 6 cases. Samples were not drawn from this region in cases 1 and 2, while in case 8 a sample was not obtained upstream to the site of arterialization in the superior vena cava.

Indicator-Dilution Curves

The characteristics of indicator-dilution curves obtained after injection of T-1824 into the right and left pulmonary arteries in patients with atrial septal defects and with anomalous pulmonary venous connections have been the subject of a recent report from this laboratory. The dilution curves noted after injection of indicator at these sites in the patients under present consideration showed similar features. Anomalous drainage (left-to-right shunts) of small or moderate magnitude was demonstrated from the left lung, while anomalous drainage of a much greater degree occurred from the right lung. In this respect, these 8 patients were similar to many of those who have atrial septal defects of the more usual type.

Two unusual features in the dilution curves of these patients provided considerable information concerning the pattern of drainage from the right lung in this condition.

1. Preferential Right-to-Left Shunting of Superior as Opposed to Inferior Cavai Blood. Small right-to-left shunts from the inferior vena cava are common in patients with atrial septal defects of the usual type, whereas right-to-left shunts from the superior vena cava, when present, are of smaller magnitude. This difference is due to the anatomic relationship of the inferior vena cava to a defect located in the region of the fossa ovalis; subsequent experience has shown conclusively that this is the usual occurrence in patients who have atrial septal defects. In 6 of our 8 patients, indicator-dilution curves were recorded after injection of dye into both the superior and the inferior vena cava, while in the remaining 2 patients (cases 3 and 8), only injections into the superior vena cava were done. In 2 patients (cases 1 and 3), right-to-left shunts were not detected from the superior vena cava (table 4); right-to-left shunts ranging from 5 to 50 per cent were demonstrated in the remainder. In 4 of the 6 patients with injections...
TABLE 4.—Comparison of Circulation Times* and Shunt Values in Anomalous Connection of Right Superior Pulmonary Vein

<table>
<thead>
<tr>
<th>Case</th>
<th>Superior vena cava</th>
<th>Inferior vena cava</th>
<th>Right pulmonary vein</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>AT</td>
<td>BT</td>
<td>R-L. shunt, per cent</td>
</tr>
<tr>
<td>1</td>
<td>9.0</td>
<td>5.0</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>4.4</td>
<td>2.4</td>
<td>5</td>
</tr>
<tr>
<td>3</td>
<td>7.2</td>
<td>4.5</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>4.0</td>
<td>2.8</td>
<td>5</td>
</tr>
<tr>
<td>5</td>
<td>5.0</td>
<td>5.0</td>
<td>16</td>
</tr>
<tr>
<td>6</td>
<td>3.1</td>
<td>3.6</td>
<td>50</td>
</tr>
<tr>
<td>7</td>
<td>7.6</td>
<td>5.4</td>
<td>17</td>
</tr>
<tr>
<td>8</td>
<td>5.2</td>
<td>3.2</td>
<td>26</td>
</tr>
</tbody>
</table>

* AT = appearance time (second); BT = build-ups time (seconds).
† Per cent of pulmonary vein blood draining to left atrium.
‡ Curves from pulmonary vein were of methylene blue. In cases 3 and 6, proportion of blood draining normally was calculated thus: \( C_{P(V)} / C_{P(I)} \), where \( C_{P(V)} \) and \( C_{P(I)} \) refer to the peak concentration after injection into the pulmonary vein and left ventricle, respectively.
§ Not obtained.
|| This indicates that 9 per cent of the systemic blood flow is represented by shunted blood, since the superior vena cava contributes about one third of the venous return.

into both venae cavae, the right-to-left shunt from the superior vena cava exceeded that from the inferior vena cava (fig. 3). In case 5, a larger right-to-left shunt was demonstrated from the inferior vena cava than from the superior vena cava and, as was predicted, a defect in the region of the foramen ovale was found at operation in addition to the defect situated high in the atrial septum (table 1).

It is apparent that the presence of preferential right-to-left shunting of blood from the superior vena cava is the consequence of the anatomic relation of the superior vena cava to the interatrial communication. The superior free border of the interatrial septum acts as a “dividing crest” to direct a portion of the superior caval blood into the left atrium, just as the limbus of the fossa ovalis acts in regard to inferior caval blood in the usual case of atrial septal defect.

Fig. 3. Diagram of potential flow patterns from superior and inferior vena cavae in anomalous pulmonary venous connection and high atrial septal defect. The solid circles represent sites of injection of indicator. The solid lines indicate direction of passage of dye from the respective sites of injection. The dilution curves (left) are representative of the drainage pattern from each site of injection. Note that the presence of a high interatrial defect permits right-to-left shunting of a small amount of superior caval blood, whereas location of the defect in relation to the inferior vena cava makes occurrence of such a shunt from the inferior vena cava unlikely.

2. Comparison of Dilution Curves Obtained from the Right Pulmonary Vein and from the Superior Vena Cava. The contour of the dilution curve of an indicator is determined largely by the path or paths traversed by the blood from the site of its injection. In deciding whether or not a pulmonary vein entered at cardiac catheterization is anomalously connected, it is of considerable importance to compare the dilution curves obtained after injection of indicator into that vein with the curve recorded after injection of dye into the superior vena cava. Identity or near similarity with regard to appearance time and contour of these curves indicates that the blood from each location drains in the same manner and is strong evidence that a common pathway is traversed by both streams (fig. 4). Conversely, dissimilarity indicates that the blood drains in an essentially different manner and suggests absence of a vascular path common to blood from the pulmonary vein and from the superior vena cava (fig. 5).

Essentially similar dye curves were obtained
FIG. 4. Demonstration of anomalous drainage of pulmonary vein by identity of dilution curves recorded after injection into anomalously connected pulmonary vein (left) and into superior vena cava (right). Typical dilution curves after injection of indicator are given above the respective diagrams. If the streams of blood share a common pathway for a distance sufficient for mixing to occur, then the drainage pathways of blood from the superior vena cava and the pulmonary vein will be similar, and identical dilution curves will result from injection of indicator at each of these sites. The identical dilution curves after injection of indicator into the superior vena cava and into a pulmonary vein are considered conclusive evidence that the vein is anomalously connected.

FIG. 5. Different dilution patterns after injection of indicator into the right pulmonary veins and into superior vena cava. When the vein is normally connected to the left atrium (left panel), indicator passes preferentially from this location to the left ventricle and only a small fraction flows from the left atrium into the right atrium. When indicator is injected into the superior vena cava, the dilution pattern is fundamentally different. In this example, a large proportion of right pulmonary vein blood and a small quantity of superior caval blood were considered to pass directly to the left ventricle. These fundamental differences in drainage are seen clearly in the dilution patterns after injection at these sites. Thus, it can be concluded that the pulmonary vein into which the injection of indicator was made is not connected to the superior vena cava.

<table>
<thead>
<tr>
<th>CASE 1</th>
<th>TIME (sec.)</th>
<th>APPEARANCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>RIGHT UPPER LOBE PULMONARY VEIN</td>
<td>6.8</td>
<td></td>
</tr>
<tr>
<td>SUPERIOR VENA CAVA</td>
<td>7.0</td>
<td>T-1824 0.13 mg./kg.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>CASE 2</th>
<th>TIME (sec.)</th>
<th>APPEARANCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>RIGHT UPPER LOBE PULMONARY VEIN</td>
<td>5.0</td>
<td></td>
</tr>
<tr>
<td>SUPERIOR VENA CAVA</td>
<td>4.4</td>
<td>Methylene Blue 0.20 mg./kg.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>CASE 7</th>
<th>TIME (sec.)</th>
<th>APPEARANCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>RIGHT UPPER LOBE PULMONARY VEIN</td>
<td>6.0</td>
<td></td>
</tr>
<tr>
<td>SUPERIOR VENA CAVA</td>
<td>7.2</td>
<td>T-1824 0.16 mg./kg.</td>
</tr>
</tbody>
</table>

FIG. 6. Demonstration of anomalous drainage of blood from right upper lobe pulmonary veins to superior vena cava in cases 1, 2, and 7. Note in each instance the near identity between the dilution curves obtained after injection at each of these sites, indicating that the vein was anomalously connected in each instance. Note also the absence of a right-to-left shunt of superior caval blood in case 1, the slight degree of shunting in case 2, and the moderate shunt present in case 7.

in cases 1, 2, and 7 (fig. 6) and in case 6 (not shown). Fundamentally dissimilar curves were obtained in cases 3 and 5. A dilution curve from the right pulmonary vein was not obtained in case 4. The magnitude of the right-to-left shunt of superior caval blood was quantitated from the dilution curve.\(^{15}\) In cases of similar drainage of superior caval and pulmonary venous blood, that portion of the pulmonary venous blood that passes directly to the left atrium is referred to as “normally drained blood,” although it circulates in a manner identical to the portion of superior caval blood that is shunted from right to left. Calculation of the “right-to-left shunt” from these curves permits approximate quantitation of the fraction of pulmonary blood draining normally (table 4). In cases 2, 3, 5, and 6, the only indicator injected into the pulmonary vein was methylene blue, whereas T-1824
was used for injection into the superior vena cava. While small right-to-left shunts can be estimated with reasonable accuracy from curves obtained by use of methylene blue, the quantitative evaluation of large shunts by this method may be subject to considerable error, possibly due to loss of this dye in the pulmonary vascular bed. Dilution curves had been obtained in cases 3 and 6 after injection of methylene blue into the left ventricle. In case 5, the catheter did not enter the left ventricle and hence it was not possible to estimate the proportion of pulmonary vein blood draining normally from the pulmonary vein in this patient; however, a large initial deflection resulted from the injection of dye in the pulmonary vein, indicating the presence of normal drainage of considerable magnitude. The dilution curves in case 8 were unusual, indicating that a greater proportion of superior caval blood than of blood from the pulmonary vein passed directly to the left ventricle.

Table 1 shows that a similar anatomic arrangement existed in cases 1, 2, 4, and 6. In case 3, a true anomalous connection was absent. The relation of the anatomic aspects of the lesion to the degree of anomalous drainage demonstrated by the dilution curves is noteworthy.

DISCUSSION

The fact that this report includes 8 patients studied over a relatively short span of time indicates that the association of high interatrial communications with anomalous connection of pulmonary veins to the right atrial-superior vena caval junction is not a rare lesion. The 6 patients operated on were in a group of 90 patients who had interatrial communications and who underwent surgical treatment. An incidence of approximately 10 percent may represent the frequency of this anomaly among a group of patients presenting the clinical and hemodynamic features of atrial septal defect. This incidence indicates that the anomaly should be sought for diligently, since correction of such a lesion may be difficult or impossible by certain surgical techniques.

Repair of atrial septal defects can be accomplished safely by a number of operative procedures. The atrial-well technic now has been used at the clinic in 90 cases for repair of atrial septal defect and left-to-right shunts. To December 1, 1955, this technic had been used in 59 such cases, with 3 deaths. Since then, an additional 31 patients have been operated on, with no deaths. This and other techniques allow operation to be done safely in such instances. A persistent common atroioventricular canal should be identified preoperatively, because use of a different procedure, namely extracorporeal circulation, is preferable for such patients. Although the condition of high atrial septal defect and anomalous connection of the right pulmonary veins to the superior vena cava has been repaired successfully by the atrial-well technic and by other procedures, its preoperative identification likewise appears surgically desirable.

These 8 patients presented the clinical features associated with atrial septal defects of the usual type, and distinctive signs that might aid in the differential diagnosis were absent. At cardiac catheterization, passage of the catheter into the pulmonary vein of the right upper or middle lobe from the superior vena cava apparently above the right atrium was an important diagnostic feature. The use of indicator-dilution curves to establish the similarity or dissimilarity of the pattern of drainage from this vein to the pattern from the superior vena cava was a noteworthy feature. Identical drainage patterns were taken as good evidence that the pulmonary vein entered was anomalously connected to the superior vena cava. Dissimilar patterns suggested that the vein was not directly connected to the superior vena cava.

The demonstration that the saturation of blood in the lower part of the superior vena cava or upper part of the right atrium consistently exceeded the values in blood from other parts of the heart strongly pointed to an interatrial communication located high in the atrial septum or an anomalous pulmonary venous connection or both. It has been demonstrated that the proportion of superior caval blood shunted in the right-to-left direction is greater than that of inferior caval blood. In only 1 patient was the right-to-left shunt from
the superior vena cava demonstrated to be of lesser magnitude than that from the inferior vena cava; a defect in the region of the fossa ovalis coexisted in this patient. On the basis of these several findings alone or in combination, the presence of an anomalously connected pulmonary vein was diagnosed before operation in each case. It was soon recognized that this anomalous connection usually was associated with a high interatrial communication, and the significance of preferential right-to-left shunting of superior caval as opposed to inferior caval blood was then apparent. When an anomalous connection of the right pulmonary veins to the superior vena cava accompanies a defect in the region of the fossa ovalis, then preferential shunting of inferior caval as opposed to superior caval blood frequently will suggest the true nature of the anomaly.

**Developmental Anatomy**

Interatrial communications involving the tissue lying posterosuperior to the region of the fossa ovalis are frequently, if not always, associated with some type of anomaly involving the pulmonary veins. In most of the cases reported in the literature, as well as in those presented in this paper, there is an anomalous connection of the upper right pulmonary veins to the superior vena cava. As a result of the location of the defect, the superior vena cava almost invariably overrides the atrial septum and drains into both atria. In addition to the afore-mentioned 3 certain and 2 probable cases described by Lewis and associates,1 16 additional cases have been reported in the literature. Necropsy was done in all these latter cases. Overriding of the atrial septum by the superior vena cava was an obvious feature in many of them. In 1 case of Rokitansky17 and in a case of Stoebcr,18 cor triatriatum was associated with this type of anomaly. A specimen in the pathologic collection at the clinic furnishes another example of this anomaly associated with cor triatriatum.19 These last 3 cases are of considerable importance in that they suggest the true nature of the interatrial communication in this malformation. The defect in these 3 cases can be considered correctly as an anomalous connection between the common pulmonary vein and the lowest portion of the superior vena cava. All the components of the true atrial septum, including the septum primum and the septum secundum, had been formed normally. The foramen ovale in each of these cases was valve-competent but patent, opening into the lower chamber of a double left atrium.

The anomalies of the venous system suggested to Edwards and Helmholz19 that the distinctive type of interatrial communication reported in the present paper can be considered as a persistence of one of the venous connections that normally exist in the fetus between the splanchnic plexus and the cardinal system of veins (fig. 7). By differential growth, the original communication between both systems of veins loses any appreciable length, forming an anomalous connection between the elements originally derived from the pulmonary vein (left atrium or the superior chamber of a cor triatriatum) and the elements derived originally from the adjacent portion of the cardinal system (superior vena cava). Unlike the condition in cor triatriatum, the left pulmonary veins open freely into the left atrium. The process of differential growth that reduces the length of communication between the common pulmonary veins and the superior vena cava allows a shifting of the right pulmonary vein, which now may connect with the superior vena cava in the region of its entry into the right atrium (fig. 8B and C) or with the posterosuperior wall of the left atrium (fig. 8A). In 3 of the patients herein reported (cases 2, 4, and 6), the right superior pulmonary vein drained blood from the upper and middle lobes, whereas in case 5, this vein drained blood from the upper lobe only. The anomalous connection in these patients was of the type depicted in figure 8C. Case 3, in which the right pulmonary veins joined to form a small diverticulum of the left atrium, was an example of a connection to the posterosuperior wall of the left atrium (fig. 8A), while the anatomic arrangement in case 1 was of the type pictured in figure 8B.

**Summary**

An unusual malformation consisting of anomalous connection of one or more of the
FIG. 7. Top. Developmental anatomy of the pulmonary venous sinus (P.V.S.), which is formed by union of the right superior (R.S.P.V.) and inferior (R.I.P.V.) pulmonary veins with the left superior (L.S.P.V.) and inferior (L.I.P.V.) pulmonary veins, and of the common pulmonary vein (C.P.V.). Note persistence of a connection (asterisk) with the primordium of the superior vena cava (S.V.C.), as seen in panel A. When the common pulmonary vein is absorbed normally into the left atrium (L.A.) and the connection persists, then its relative length is lost and an interatrial communication exists above the fossa ovalis, as shown progressively in panels B and C. This anomaly represents class B, subtype b, in the classification of total anomalous pulmonary venous connection by Edwards and Helmholz.

FIG. 8. Bottom. Types of congenital malformation of the heart resulting from persistence of a communication between the pulmonary venous sinus and the primordium of the superior vena cava. A. The superior vena cava (S.V.C.) straddles the cephalad margin of the atrial septum, in communication with the left atrium (L.A.) behind and the right atrium (R.A.) in front. The right pulmonary veins join the cephaladorsal portion of the left atrium to form a sinus communicating with the left atrium, the superior vena cava and the right atrium across the cephalad margin of the atrial septum. This anomaly existed in case 3. B. The right superior (R.S.P.V.) and inferior (R.I.P.V.) pulmonary veins connect with the lower portion of the superior vena cava and the upper portion of the right atrium, respectively. Case 1 was an example of this type of malformation. C. The right superior pulmonary veins connect to the lower portion of the superior vena cava at the level of the interatrial communication, while the right inferior pulmonary veins connect to the left atrium. This was the commonest variant seen, being present in cases 2, 4, 5, and 6.
right pulmonary veins with the superior vena cava at its junction with the right atrium associated with a high interatrial communication has been diagnosed by means of cardiac catheterization in 8 patients at the Mayo Clinic. In 1 of the 6 patients on whom operation was done, an additional interatrial communication was found in the region of the fossa ovalis. In 4 of the patients, a significant defect in this region was absent. In all 6 patients interatrial communications of moderate size were found localized to the superior aspect of the right atrium close to the entry of the superior vena cava into this chamber. As a result of such location, the superior vena cava overrode the atrial septum in a number of instances. This malformation is believed to be a form of anomalous pulmonary venous connection, in contrast to atrial septal defects of the usual type.

The patients had the symptoms, physical signs, and basic hemodynamic patterns common in atrial septal defect. Features of diagnostic value at cardiac catheterization were as follows:

1. A cardiac catheter could be passed from the superior vena cava into the right pulmonary vein draining the upper or middle lobe from a location above or near the junction of the superior vena cava with the right atrium.

2. The oxygen saturation of samples of blood withdrawn from the juncture of the superior vena cava with the right atrium was equal to or greater than that of other samples from the right side of the heart.

3. There was a preferential right-to-left shunt of the superior caval blood as opposed to inferior caval blood.

4. Anomalous venous drainage of the right upper lobe was demonstrated by the similarity of indicator-dilution curves recorded after injection of dye into the superior vena cava and into the vein and artery of the right upper lobe.

**SUMMARIO IN INTERLINGUA**

Catheterisation cardiac de octo patientes al Clinica Mayo resultava in le diagnose de un malformation inusual, consistente del connexion anormal de un o plures del venas dextero-pulmonar con le vena cave superior al puncto de su junction con le atrio dextere, in association con le presentia de un communication alti-interatrial.

Operationes esseva execute in 6 del patientes. In 1 del 6, un communication interatrial additional esseva trovate in le region del fossa oval. In 4 del patientes, nulle defecto significative esseva trovate in iste region. In omne 6 casos, communicationes interatrial de dimensiones moderate esseva trovate localisate al aspecto superior del atrio dextere, proxime al entrata del vena cave superior in iste camera. Como resultato de iste location, le vena cave superior passava in ultra del septo atrial in un numero de casos. Es formulate le opinion que iste malformation es un genere de anormal connexion pulmono-venose, in contrasto con le typo usual de defecto del septo atrial.

Le patientes habeva le symptomas, signos physic, e basic conformation hemodynamic que es le regula in defectos del septo atrial. Observationes de valor diagnostic, facite per catheterisation cardiac, esseva le sequente:

1. Le catheter cardiac poteva esser passate ab le vena cave superior a in le vena dextero-pulmonar, con drainage del lobo superior o medial ab un loco supra o proxime al junction del vena cave superior con le atrio dextere.

2. Le saturation oxygenic de specimens de sanguine obtenite al junction del vena cave superior con le atrio dextere esseva equal o superior al saturation oxygenic de altere specimens obtenite al latere dextere del corde.

3. In le derivation dextero-sinistre le sanguine esseva supero-caval plus tosto que infero-caval.

4. Anormalitate de drainage venose del lobo dextero-superior esseva demonstrate per le similaritate de curvas de dilution de substantia indicatori obtenite post injection del substantia in le vena cave superior con curvas obtenite post injection in le vena e arteria del lobo dextero-superior.

**REFERENCES**


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In the mean time I shall say so much, that there are many things allowed and received in Physiologie, Pathologie, and Medicine, that no body knows the cause of; yet that there are such things no body is ignorant, namely, of rotten feavers, revulsion, purgation of excrement, yet all these things are known by the help of Circulation.—William Harvey, _de Circulis Sanguinis_, 1649.
Anomalous Connection of Right Pulmonary Veins to Superior Vena Cava with Interatrial Communications: Hemodynamic Data in Eight Cases
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