Total Anomalous Pulmonary Venous Connection with Severe Pulmonary Venous Obstruction

A Clinical Entity

By Alois R. Hastreiter, M.D., Milton H. Paul, M.D., Marian E. Molthan, M.D., and Robert A. Miller, M.D.

FRIEDLOWSKY, in 1868, was the first to report total anomalous pulmonary venous connection.\(^1,2\) This entity, with its multiple varieties,\(^3\) is receiving increasing attention with the recent encouraging reports of successful surgical therapy.\(^4\) The infradiaphragmatic form was first described by Ghon in 1916,\(^5\) and a total of 37 cases has since been reported.\(^5-24\) Obstruction to the pulmonary venous drainage has been implicated as the basis for the characteristic radiologic features of this form of anomaly, i.e., a relatively small heart associated with diffuse haziness and reticulation of the lung fields. More recently pulmonary venous obstruction complicating the supracardiac type has been reported by Harris et al. (1960)\(^21\) and by Hauck et al. (1960).\(^25\)

This study presents clinical, physiologic, and pathologic data from six male infants with severe pulmonary venous obstruction in otherwise uncomplicated total anomalous pulmonary venous connection and a review of the literature. The clinical picture is remarkably uniform and characteristic, regardless of the site of the pulmonary venous obstruction.

Case Material

Cases 1 to 4. J.D., K.T., B.M., and R.M. These infants had tachypnea, cyanosis, and feeding difficulties on the first day of life. Oxygen produced some relief. Several days or weeks later these babies showed signs of congestive heart failure. Response to therapy was poor, and there was steady deterioration leading to an early death (table 1).

Case 5. T.T. was admitted to the Children's Memorial Hospital on the first day of life because of an imperforate anus. Congenital heart disease of cyanotic type was detected on physical examination. Cyanosis and dyspnea were rapidly progressive. An angiocardiogram was performed, and 2 days later a surgical anastomosis was created between the left common pulmonary vein and the left atrium. After surgery the baby's condition gradually deteriorated with evidence of progressive cardiac failure and apneic spells. Death occurred 10 days after surgery, on the twenty-sixth day.

Case 6. T.H. On the first day of life the baby became cyanotic and dyspneic and was placed in oxygen. He was transferred to the Children's Memorial Hospital at 9 days of age. Following digitalization, there was moderate clinical improvement. Cardiac catheterization and angiocardiograms were performed, and, during the selective angiocardiogram from the right ventricle, perforation of the right ventricle occurred. Dramatic improvement followed a pericardioceotensis. Several days later the baby again showed signs of progressive heart failure and died 11 days following the cardiovascular studies.

The clinical and laboratory findings in these cases are shown in tables 1 and 2 and figures 1, 2, and 3.*

Angiocardiography

Selective cineangiocardiograms were performed on four patients. In all cases a right-to-left shunt was visualized at the atrial level with simultaneous visualization of the pulmonary artery and the aorta. The right-sided chambers and the pulmonary artery and branches were considered larger than normal and the left atrium and ventricle were small.

In case 2 there was a large right-to-left shunt at the ductus level. The pulmonary venous return

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*From the Division of Cardiology, The Children's Memorial Hospital, and the Department of Pediatrics, Northwestern University Medical School, Chicago, Illinois.

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*Cases 1 and 3 were previously included in a radiologic report.*
was markedly delayed. The pulmonary veins entered a vertical trunk behind the heart, and this was followed by faint visualization of the hepatic veins.

In case 4, the pulmonary veins joined to form an ascending common trunk that entered the left innominate vein and then the superior vena cava. At the origin of the anomalous vertical pulmonary vein, there was a definite narrowing to approximately half its original caliber.

In case 5, the drainage of the pulmonary veins was not well visualized, but there was marked persistence of the contrast material in the dilated pulmonary veins.

In case 6, the left main pulmonary artery filled first, and was followed 1.6 seconds later by filling of the right main pulmonary artery—a marked delay. The course of the pulmonary veins was not well defined. At the end of the cycle, after an estimated 10 seconds, a faint amount of contrast material was seen in the portal system.

Postmortem Findings

The general characteristics of the hearts are listed in table 3. An unusual feature was a completely closed foramen ovale in case 3. The detailed pathologic anatomy of the anomalous pulmonary veins is illustrated in figure 4.

Case 1. The four pulmonary veins joined near the midline and posterior to the left atrium to form a common trunk, which descended into the abdomen anterior to the esophagus, piercing the diaphragm and connecting to the portal vein at
its origin behind the head of the pancreas. The portal vein was much enlarged and the liver was moderately large. The ductus venosus was obliterated.

Case 2. The pulmonary veins formed a common vertical trunk, which descended into the abdomen, as in case 1, and then connected to the left branch of the portal vein. The widest diameter of the common trunk was 0.6 cm. At the point of entrance into the portal vein, there was a definite narrowing due to obliterative changes, constricting the orifice to 0.1 cm. in diameter. The ductus venosus was closed. The left lobe of the liver showed much more intense congestion than the right lobe and there was a sharp color line of demarcation between them.

Case 3. The four pulmonary veins formed a common trunk, which descended like the others, and penetrated the diaphragm through the esophageal hiatus. The infradiaphragmatic portion of this common venous channel became bulbous, measuring 0.7 cm. in diameter, and connected with the patent ductus venosus, which was 0.3 cm. in diameter. The ductus venosus drained into the inferior vena cava at the confluence of the left hepatic vein. The umbilical vein was patent, 0.2 cm. in diameter, and its hepatic portion was considerably larger, measuring 0.8 cm. in diameter.

Case 4. The four pulmonary veins formed a common trunk, which drained into an anomalous vertical pulmonary vein that entered the superior vena cava via the left innominate vein. At the junction of the common pulmonary vein, the vertical vein exhibited a rather prominent constriction and a saccular aneurysmal dilatation just proximal to the stenotic area.

Case 5. The four main pulmonary veins joined in the midline behind the heart to form an apparently blind venous trunk. An accessory pulmonary vein arose from the right upper lobe and emptied into the superior vena cava 2 cm. proximal to its connection to the right atrium. On careful dissection it was established that the branches of the pulmonary veins from the right middle lobe anastomosed with branches of the accessory pulmonary veins from the right upper lobe. A surgical anastomosis had been created between the common pulmonary vein and the left atrial appendage; the anastomotic orifice was rather small, measuring 0.2 cm. in diameter.

Case 6. The pulmonary veins from the right lung and from the left upper lobe joined in the midline forming a blind venous channel. These veins were small, thick-walled, and had a narrow lumen. An accessory pulmonary vein of moderately large caliber extended from the lower mediastinal surface of the left lower lobe, posterior to the esophagus, piercing the diaphragm by way of the esophageal hiatus, terminating in the portal vein along its midlateral aspect. A very narrow cord-like structure extended from the right lower lobe into the portal vein; it had no lumen. This probably represented a vestigial accessory pulmonary vein.

Figure 1

Typical radiologic aspect of the heart and lungs (case 6—anteroposterior and lateral views).
TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

Discussion

Extensive studies of the normal embryogenesis of the pulmonary veins and their connections to the heart have been reported.\textsuperscript{3, 26-28} In brief, as the lung buds develop, they are enveloped by a venous plexus that communicates with the cardinal and umbilicovitelline systems. A common pulmonary vein is first recognizable in the 4-mm. embryo as an evagination of the superior wall of the left atrium.

Figure 2

Electrocardiograms of cases 1 (J.D.), 3 (B.M.), 4 (R.M.), 5 (T.T), and 6 (T.H.).
Table 3
Postmortem Data in Six Cases of Total Anomalous Pulmonary Venous Connection with Severe Pulmonary Venous Obstruction

<table>
<thead>
<tr>
<th>Case no.</th>
<th>1 (J.D.)</th>
<th>2 (K.T.)</th>
<th>3 (B.M.)</th>
<th>4 (R.M.)</th>
<th>5 (T.T.)</th>
<th>6 (T.H.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart size</td>
<td>N</td>
<td>Mod. L</td>
<td>Sl. L</td>
<td>Sl. L</td>
<td>Sl. L</td>
<td>N</td>
</tr>
<tr>
<td>Right atrium</td>
<td>Mod. D</td>
<td>Mod. D</td>
<td>Mark. D</td>
<td>Mod. D</td>
<td>Mark. D</td>
<td>Mod. D</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>Mod. H</td>
<td>Mod. H</td>
<td>Mod. H</td>
<td>Mod. H</td>
<td>Mark. H</td>
<td>Mark. H</td>
</tr>
<tr>
<td>Left atrium</td>
<td>Sl. D</td>
<td>Mod. D</td>
<td>Mark. H</td>
<td>Mod. D</td>
<td>Sl. D</td>
<td>Mod. D</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>Mod. T</td>
<td>Mod. T</td>
<td>Very S</td>
<td>Mod. S</td>
<td>Mod. S</td>
<td>Mod. S</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>Mod. T</td>
<td>Mod. T</td>
<td>N</td>
<td>Mod. T</td>
<td>Mod. T</td>
<td>Mod. T</td>
</tr>
<tr>
<td>Aorta</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mod. L</td>
</tr>
<tr>
<td>Tricuspid valve</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mark. D</td>
<td>Mark. D</td>
</tr>
<tr>
<td>Mitral valve</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>N</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mod. L</td>
</tr>
<tr>
<td>Foramen ovale</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mark. D</td>
</tr>
<tr>
<td>Ductus arteriosus</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mark. D</td>
</tr>
<tr>
<td>Pulmonary venous trunk</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mark. D</td>
</tr>
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<td>Mod. L</td>
<td>Mod. L</td>
<td>Mark. D</td>
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<td>Mark. D</td>
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</tr>
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<td>Mark. D</td>
<td>Mod. L</td>
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<td>Mark. D</td>
</tr>
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<td>Mod. L</td>
<td>Mark. D</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mark. D</td>
</tr>
<tr>
<td>Ductus arteriosus</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mark. D</td>
</tr>
<tr>
<td>Pulmonary venous trunk</td>
<td>Mod. L</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mod. L</td>
<td>Mark. D</td>
<td>Mark. D</td>
</tr>
</tbody>
</table>

*Measurements in centimeters.
†Largest dimension of patent foramen ovale.

Abbreviations: N, normal; Mod., moderately; Sl., slightly; Mark., markedly; Diam., diameter; Circ., circumference; L, large; D, dilated; H, hypertrophied; S, small; T, thin.

It later connects to the pulmonary venous plexus, establishing the pulmonary venous return into the left atrium.

The clinical picture of total anomalous pulmonary venous connection with severe pulmonary venous obstruction varies markedly from the unobstructed form. Table 4 lists the main differential characteristics of these two groups. Of the 37 cases of the infracardiac variety reported only seven occurred in female patients, and all the supercardiac cases concerned male infants. Parsons et al.11 pointed out the intensification of cyanosis and pulmonary congestion with straining in the infradiaphragmatic group, which they attributed to compression of the long and narrow pulmonary venous trunk by increased intra-abdominal and intrathoracic pressures. The increase in cyanosis and respiratory distress with feeding may be explained on the basis of the proximity of the vertical pulmonary venous trunk to the esophagus.24

All types of total anomalous pulmonary venous connection usually present a moderate to severe right axis deviation in the electrocardiogram. The obstructed group lacks the tall, peaked P waves so often recorded in the usual case of total anomalous pulmonary ve-
nous connection. The pattern of right ventricular hypertrophy characteristically is represented by a tall R or qR complex in the right precordial leads and deep S waves with little or no R wave over the left chest (reversal of the R/S ratio). No instances of the rsR' pattern, which is not uncommonly described in total anomalous pulmonary venous connection, have been observed in the obstructed group.

In the obstructed variety, the anteroposterior and lateral roentgenograms of the chest are typical and are considered the key to the diagnosis. A number of excellent descriptions are provided in the literature dealing with the infradiaphragmatic type. In our experience the supradiaphragmatic variety with severe obstruction shows identical findings. The heart is normal in size and configuration, or only slightly enlarged. The lung fields exhibit a diffuse haziness throughout, giving them a ground-glass appearance. The pulmonary markings are prominent with a distinctly reticulated appearance that suggests interlobular edema and is best seen in the lateral views. These markings sometimes obscure the borders of the heart (fig. 1). Diffuse air trapping, areas of atelectasis, or suggestive pneumonitis may be present. The appearance of the lung fields in association with a small heart size strongly suggests pulmonary venous obstruction.

From the limited data available it can be concluded that angiocardiographic studies may be extremely helpful in determining the anatomy of the anomalous pulmonary venous connection, in indicating the site of the obstruction (as in case 4 of this series) and in furnishing information on the circulation time in the obstructed pulmonary venous segment. In the infradiaphragmatic group, the vertical descending pulmonary venous trunk can be visualized. The pulmonary veins fill very late with contrast material at 4½ and 5 seconds in cases of Lucas et al. The time elapsed between the visualization of the main pulmonary artery and the portal veins is 8 seconds (case 2), 10 seconds (case 5), 9 seconds, and 10½ seconds. Although this method of measuring the pulmonary circulation time is crude, the evidence of a marked delay in the drainage of the obstructed pulmonary venous circuit is clear. The angiocardiograms in the obstructed supracardiac cases of this series also show a marked prolongation of the pulmonary venous return. In case 6 there was a significant delay from the filling of the left to the filling of the right.
main pulmonary arteries; this could be explained by the different degree of pulmonary venous obstruction in each lung.

There has been no detailed report in the literature of a cardiac catheterization performed in the infradiaphragmatic group of total anomalous pulmonary venous connection. A complete study was possible in one of our patients of this type (case 2). The discussion of the obstructed supracardiac type is based on the findings in our cases 4 and 6 and the data obtained from the report of Hauck et al.25 The hemodynamic findings in the obstructed and unobstructed cases are listed in Table 5. The site of the left-to-right shunt (infra- or supracardiac) is usually readily recognized. The main differential characteristics in the obstructed group are those of a lower peripheral arterial saturation, a markedly elevated pulmonary artery pressure level (higher than systemic pressures) (fig. 3), and a very high pulmonary artery wedge pressure. There is a pronounced gradient between the mean pressures of the proximal portion of the pulmonary veins (pulmonary artery wedge pressure) and the right atrium, indicating the existence of an obstruction in the course of the pulmonary veins. The estimated pulmonary blood flow is normal or near normal. In our experience, these patients seem especially susceptible to the development of arrhythmias during cardiac catheterization.

At autopsy, in the cases of obstructed total anomalous pulmonary venous connection, the cardiac size is described as normal or as only slightly enlarged. The right atrium and right ventricle are usually moderately enlarged and

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### Table 4

<table>
<thead>
<tr>
<th></th>
<th>Total pulmonary venous connection without obstruction</th>
<th>Total pulmonary venous connection with obstruction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td>Nearly equal distribution</td>
<td>Almost all males</td>
</tr>
<tr>
<td>Onset of symptoms</td>
<td>Usually early</td>
<td>Very early (0-3 weeks)</td>
</tr>
<tr>
<td>Age at death</td>
<td>Approximately 20% survive the first year of life</td>
<td>Infradiaphragmatic, 2 da. to 4 mo.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Supradiaphragmatic, 2 wk. to 1 mo.</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Tachypnea, dyspnea, congestive heart failure</td>
<td>Same</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>Very mild</td>
<td>Moderate</td>
</tr>
<tr>
<td>Heart size</td>
<td>Large, hyperdynamic</td>
<td>Normal, quiet</td>
</tr>
<tr>
<td>Thrills</td>
<td>Occasionally at the upper left sternal border</td>
<td>Absent</td>
</tr>
<tr>
<td>Systolic murmurs</td>
<td>Present in 75%—usually soft, occasionally loud at the upper left sternal border</td>
<td>Absent in 50%; very soft 'ejection' murmurs along left sternal border in 50%</td>
</tr>
<tr>
<td>Diastolic flow murmurs</td>
<td>Almost always present beyond 6 mos. of age</td>
<td>Absent</td>
</tr>
<tr>
<td>Continuous 'vascular'</td>
<td>Occasionally along the left sternal border (usually associated with drainage into an anomalous vertical pulmonary vein)</td>
<td>Present only in the supradiaphragmatic variety; its location varies with the site of obstruction</td>
</tr>
<tr>
<td>murmurs</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Second heart sound</td>
<td>Variable intensity—usually well split and fixed with respiration</td>
<td>Increased—usually well split and fixed with respiration</td>
</tr>
<tr>
<td>Other heart sounds</td>
<td>Commonly present (triple or quadruple rhythm)</td>
<td>Absent</td>
</tr>
</tbody>
</table>
Figure 4

Anatomic course of the anomalous pulmonary veins. 1. Drainage into the portal vein or its left branch (cases 1 and 2 respectively). 2. Drainage into the ductus venosus (case 3). 3. Drainage into a left anomalous vertical vein having a zone of stenosis (case 4). 4. Drainage by means of an accessory pulmonary vein from the upper lobe of the right lung into the superior vena cava or azygos vein (case 5). 5. Drainage by means of a hypoplastic accessory pulmonary vein from the lower lobe of the left lung into the portal vein (case 6). APV, accessory pulmonary vein; AVPV, anomalous vertical pulmonary vein; CPV, common pulmonary vein; CPVT, common pulmonary venous trunk; DV, ductus venosus; IVC, inferior vena cava; LHV, left hepatic vein; LIV, left innominate vein; LPV, left portal vein; PV, portal vein; RHV, right hepatic vein; RLL, right lower lobe; RPV, right portal vein, RUL, right upper lobe; SVC, superior vena cava.
Table 5
Effect of Severe Pulmonary Venous Obstruction on Hemodynamic Findings in Total Anomalous Pulmonary Venous Connection

<table>
<thead>
<tr>
<th>Total pulmonary venous connection</th>
<th>Peripheral arterial saturation</th>
<th>Pulmonary arterial wedge pressure</th>
<th>Pulmonary artery pressure</th>
<th>Pulmonary blood flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>Without obstruction</td>
<td>80-95%</td>
<td>Variable; usually normal</td>
<td>Variable</td>
<td>Increased; moderate to large L-R shunt</td>
</tr>
<tr>
<td>With obstruction</td>
<td>Below 65%</td>
<td>Very high</td>
<td>Markedly elevated; higher than systemic</td>
<td>Nearly normal</td>
</tr>
</tbody>
</table>

thickened. The left-sided chambers are almost always described as moderately small, and in one half the cases have a thinner than normal wall. The tricuspid valve ring is usually slightly enlarged, and that of the mitral valve normal or slightly narrowed. The pulmonary artery is mildly dilated, and the aorta is either normal or slightly small. The interatrial communication is in almost all cases a foramen ovale, which is patent to an extent of 0.3 to 1.0 cm. The ductus arteriosus is described as a probe-patent or "closing" in two thirds of the cases. The remaining third is equally divided into completely obliterated or widely patent (up to 1.4 cm. in diameter). An important surgical consideration is the disproportion between the common pulmonary vein and the waist of the left atrial appendage, as pointed out by Edwards and Du Shane for the general group of total anomalous pulmonary venous connection. Sherman and Bauersfeld suggested that the smallness of the left side of the heart is due to atrophy and not hypoplasia, hence the possibility of an increase in its size when hemodynamically stimulated.

In the infradiaphragmatic variety, the four pulmonary veins usually join slightly to the left of the midline, posterior to the left atrium to form a common pulmonary venous trunk that descends anterior to the esophagus and traverses the diaphragm to connect to either the portal venous system or the ductus venosus. Direct connection of the pulmonary venous trunk to the inferior vena cava is extremely rare but it has been mentioned. The common pulmonary trunk often decreases in diameter as it progresses toward the abdomen, and in several instances a definite saccular dilatation is present at the site of its junction with the abdominal vessel. Histologic obliterative changes were shown to take place at its caudal end, capable of constricting its entrance into the receiving vein. The trunk most commonly traverses the diaphragm via the esophageal hiatus, but in some cases it reaches the abdomen through an accessory hiatus between the esophageal and the caval openings. It connects either to the portal vein (25 of 34 cases, four of which drain into its left branch), or to the patent ductus venosus (6 of 34 cases). When it enters the ductus venosus, this structure, which is patent, drains the total pulmonary venous return into the inferior vena cava. The patent ductus venosus is always significantly narrower than the incoming pulmonary venous trunk, and obliterative changes affecting this venous structure have been demonstrated. The umbilical vein may or may not be patent, but is usually obliterating. When the common pulmonary venous trunk connects to the portal vein or its left

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*We have recently studied an additional case of infradiaphragmatic total anomalous pulmonary venous connection with drainage directly into the inferior vena cava, at the level of the entrance of the hepatic veins. The common pulmonary venous trunk narrowed to an orifice of 1 mm. in diameter at the site of the connection (fig. 5).
Postmortem specimen showing infradiaphragmatic total anomalous pulmonary venous connection directly to the inferior vena cava (15-day-old girl). The inferior vena cava has been cut and the heart reflected upwards to expose the anomalous pulmonary venous system. An, surgical anastomosis between left atrium and pulmonary vein; CPVT, common pulmonary venous trunk; HV, hepatic veins; IVC, inferior vena cava; J, junction of common pulmonary venous trunk and inferior vena cava; LA, left atrium; PV, pulmonary veins.

Figure 5

branch, the pulmonary venous return reaches the inferior vena cava, either by traversing the hepatic sinusoids and hepatic veins or in a retrograde manner, through the patent ductus venosus, reaching the heart without traversing the liver. In some cases, with connection to the portal vein the ductus venosus was open, although undergoing obliteratorive changes. The majority of authors, however, do not comment on the status of this structure.

In those cases with drainage into the portal vein in which the ductus venosus is obliterated, a marked dilatation of the portal venous system is observed distal to the entrance of the pulmonary venous trunk and there is exaggerated hepatomegaly (cases 1 and 2). In case 2, where the pulmonary venous trunk entered the left branch of the portal vein, these findings were limited to the left lobe of the liver. They confirm the idea that the pulmonary venous return traverses the liver in order to reach the inferior vena cava.

In the supradiaphragmatic variety with obstruction the following variations in the path of the pulmonary veins have been observed:

1. The four pulmonary veins connect to a persistent anomalous vertical vein that drains into the right atrium via the innominate vein and superior vena cava. A severe anatomic constriction and poststenotic dilatation are present near the origin of the anomalous vertical pulmonary venous trunk (case 4). One of the infants reported by Hauck et al. belongs in this group.
2. The four pulmonary veins join near the midline forming what appears to be a blind sac (cases 5 and 6). In both these cases, however, an accessory pulmonary vein is present, draining respectively from the right upper lobe of the lung into the superior vena cava (case 5), and from the left lower lobe of the lung into the portal vein (case 6). On careful dissection, it can be demonstrated that these accessory pulmonary veins connect with branches of the normal pulmonary vein of the lobe of the lung in which the accessory pulmonary vein originates, thus resulting in drainage of the entire pulmonary venous system of both lungs through the accessory vein. In one of the cases reported by Hauck et al., a similar disposition of the pulmonary veins was described, the accessory vein originating in the right upper lobe of the lung and draining into the azygos vein.

In one of our patients (case 6) the pulmonary veins and the accessory pulmonary veins are very hypoplastic and can hardly have allowed efficient pulmonary venous drainage.

The lungs show similar gross anatomic and histologic findings in all cases. They are moderately voluminous, pale, except for small areas that appeared congested and showed a pink external discoloration. The external surfaces exhibit a peculiar reticulated pattern outlining the pulmonary lobules, which appear to be separated by thick septa. Cut sections through the lungs show a moderate amount of congestion and patchy, small, atelectatic areas.

Microscopic sections show patchy areas of atelectasis and congestion, with scattered, small focal hemorrhages. The interlobular septa are widened with interstitial edema, marked lymphangiectasis, and, on occasions, an increase in interlobular connective tissue. In the areas of collapse there is a marked macrophage infiltration. The pulmonary arterioles are slightly thickened.

Nature of the Pulmonary Venous Obstruction

In the infradiaphragmatic type, several factors may participate in the production of pulmonary venous obstruction: (a) the marked cross-sectional difference between the main pulmonary veins, the common venous trunk, and the ductus venosus or the portal vein; (b) a pressure gradient between the portal venous system and the pulmonary venous system; (c) the obligatory passage of the pulmonary venous return through the liver sinusoids, and (d) obliterative changes in the distal end of the pulmonary venous trunk or the ductus venosus, which may progress in extraterine life.

An interesting description exists on a case of total anomalous pulmonary venous connection into a left gastric vein. The clinical picture was quite different from the usual total anomalous pulmonary venous connection and consisted of severe episodes of hematemesis that lead to death on the second day of life. Autopsy revealed prominent esophageal and gastric varices.

In the supradiaphragmatic group, evidence of pulmonary venous obstruction is usually readily apparent at autopsy. It consists of a constriction of the anomalous vertical pulmonary vein, or termination of the main pulmonary veins in a "blind sac," with total or partial drainage of the pulmonary venous return through an accessory pulmonary vein. This accessory vein drains anomalously into the superior vena cava, the azygos vein, or the portal vein. In at least one case, however (case 6 of this series), the hypoplastic pulmonary veins, including the accessory vein, were obviously too small to carry the total pulmonary venous drainage, and another explanation must be sought. Zuckerkandl (1881) demonstrated with postmortem injections of the pulmonary veins that anastomoses occur between the pulmonary and the systemic venous systems, the latter represented mainly by derivatives of the cardinal veins. The main sites for these anastomoses are the connections between the pulmonary veins and the bronchial or mediastinal veins, and, to a lesser extent, the aortic venous plexus and azygos veins via the esophageal and posterior mediastinal veins. The pulmonary venous plexus also
communicates with the derivatives of the umbilico-vitelline system by small vessels, the venae comitantes of the vagus nerve, which pass through the diaphragm with the vagus and enter the portal vein. This alternate pathway via the systemic veins is possibly a significant factor in the cases under study.

**Total Anomalous Pulmonary Venous Connection with Absent Interatrial Communication**

The presence of an interatrial communication is considered an integral part of the complex of total anomalous pulmonary venous connection. One of our cases of infradiaphragmatic type (case 3) had a completely obliterated foramen ovale at the time of death. The right-to-left shunt must have taken place via a patent ductus arteriosus, which was very large (0.8 cm in diameter). Another interesting anatomic feature in this case was the presence of extremely small left chambers of the heart. The ratio of the volumetric capacities of the right and left chambers was approximately 5:1. The coronary arterial blood supply could have been maintained only through retrograde filling of the initial portion of the aorta via the patent ductus arteriosus. On histologic examination, multiple areas of healing myocardial infarctions were found, involving predominantly the left ventricle. No other living infant with obstructed total anomalous pulmonary venous connection and a completely intact atrial septum has been reported. None of our cases in the infradiaphragmatic group have had a surgical procedure; this has been unsuccessful in the cases so far reported. In one of our obstructed supradiaphragmatic cases (case 5) an anastomosis was performed between the common pulmonary venous trunk and the left atrium. The anastomosis was too small, and the baby died 11 days later. If surgery is contemplated it should be performed early, probably during the first 2 to 3 weeks of life, since clinical deterioration proceeds very rapidly.

**Summary**

The clinical, physiologic, and anatomic features of a special group of cases of total anomalous pulmonary venous connection are reviewed.

The occurrence of severe pulmonary venous obstruction in total anomalous pulmonary venous connection produces a characteristic syndrome:

1. Very early onset of dyspnea and heart failure.
2. Considerably more cyanosis than the usual case without obstruction.
3. Typical x-ray showing diffuse hazy lung fields with reticulated appearance without cardiac enlargement.
4. Electrocardiographic evidence of severe right ventricular hypertrophy with a tall R or qR pattern in lead V1 and reversal of the R/S ratio over the precordium.
5. Physiologic findings of practically normal pulmonary flow, right ventricular and pulmonary arterial pressures greater than systemic, and a marked gradient between the pulmonary arterial "wedge" and right atrial pressures.
6. Rapid deterioration and death in the first weeks or months of life.

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ALOIS R. HASTREITER, MILTON H. PAUL, MARIAN E. MOLTHAN and ROBERT A. MILLER

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