Infradiaphragmatic Anomalous Pulmonary Venous Drainage

Normal Hemodynamics Following Operation in Infancy

By W. Jegier, M.D., E. Charrette, M.D., and A. R. C. Dobell, M.D.

SUMMARY

The pathological anatomy and hemodynamic effects of total anomalous pulmonary venous drainage of the infradiaphragmatic type are reviewed in nine infants examined from 1940 to 1965 in the pathology department. Two infants underwent corrective surgery in the period; one operation was unsuccessful. The other infant had normal hemodynamics 6 months postoperatively and continues to do well 2 years after operation. Surgical principles are discussed. Some patients undergoing an anastomosis between the common pulmonary venous trunk and the left atrium will not require a second operation either to ligate the venous trunk or to close the atrial septal defect.

ADDITIONAL INDEXING WORDS:
Foramen ovale communication
Anastomosis of common pulmonary venous trunk and left atrium
Oxygen saturation
Blood pressure

In approximately 10% of cases of total anomalous pulmonary venous drainage, the common pulmonary venous trunk runs caudad anterior to the vertebral bodies, pierces the diaphragm, and drains into the ductus venosus, the inferior vena cava, or the portal system.1 The left atrium is small since it is filled only by way of a patent foramen ovale. Such infants are typically cyanotic from birth and die in the first few weeks of life.2 The embryological maldevelopment is similar to other types of total anomalous venous drainage. The pulmonary veins begin as a primordial bud from the superior-posterior wall of the left atrium. This does not communicate with the pulmonary venous plexus which at this period forms a part of the splanchnic plexus. The pulmonary venous bud becomes canalized and unites with the pulmonary venous plexus. As development progresses, the connection between the venous plexus and the splanchnic plexus obliterates and the common pulmonary trunk becomes absorbed into the left atrial wall resulting in separate entry of all four pulmonary veins into the left atrium.

Nine infants with infradiaphragmatic total anomalous pulmonary venous drainage (IDAPVD) were examined by the Pathology Department of the Montreal Children’s Hospital from 1940 to 1965. Certain clinical and pathological details concerning these infants are shown in table 1. One of these nine infants had undergone unsuccessful operative correction. Another infant survived operation and is alive and well 2 years later with normal hemodynamics. This experience is reported in detail to emphasize the possibility of complete correction and to add it to the three previously reported successful operations.4 5

Report of Case

The child was the product of a normal full-term pregnancy with a birth weight of 3,145 gm. She appeared normal at birth, but at 6 hours of age had a transient episode of generalized cyanosis requiring oxygen with rapid improvement in

From the Department of Cardiovascular Surgery, The Montreal Children’s Hospital, and the Joint Cardiorespiratory Service, The Royal Victoria Hospital, and The Montreal Children’s Hospital, McGill University, Montreal, Canada.
ANOMALOUS PULMONARY VENOUS DRAINAGE

Table 1

Autopsy Experience with Infradiaphragmatic Total Anomalous Pulmonary Venous Drainage

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age at death</th>
<th>Type of pulmonary venous drainage</th>
<th>Interatrial communication</th>
<th>Additional features</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3 wk</td>
<td>Portal vein</td>
<td>Flap foramen ovale</td>
<td>Patent ductus venosus and umbilical vein</td>
</tr>
<tr>
<td>2</td>
<td>6 days</td>
<td>Lt. gastric &amp; portal vein</td>
<td>Flap foramen ovale</td>
<td>Patchy pneumonia</td>
</tr>
<tr>
<td>3</td>
<td>9 days</td>
<td>Ductus venosus</td>
<td>Flap foramen ovale (9 mm)</td>
<td>Operative death</td>
</tr>
<tr>
<td>4</td>
<td>12 days</td>
<td>Ductus venosus</td>
<td>Flap foramen ovale (4 mm)</td>
<td>Fibrous cord between</td>
</tr>
<tr>
<td>5</td>
<td>23 days</td>
<td>Portal vein</td>
<td>Flap foramen ovale (&quot;large&quot;)</td>
<td>LUL pulmonary vein and left atrium</td>
</tr>
<tr>
<td>6</td>
<td>3 wk</td>
<td>—</td>
<td>Flap foramen ovale (5 mm)</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>2 mo</td>
<td>Portal vein</td>
<td>Flap foramen ovale (4 mm)</td>
<td>Bronchiolitis</td>
</tr>
<tr>
<td>8</td>
<td>2 mo</td>
<td>—</td>
<td>Flap foramen ovale</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>33 days</td>
<td>Portal vein</td>
<td>Flap foramen ovale (8 mm)</td>
<td>Marked stenosis of venous trunk at diaphragm</td>
</tr>
</tbody>
</table>

LUL = left upper lobe.

Figure 1

Roentgenograms of the chest. (Left) Before operation. There is significant overdistension of the lung with a heart of almost normal size and marked pulmonary venous congestion. Fluid can be seen in the major fissure on the right. Small amounts of fluid in both pleural cavities. (Right) Postoperative roentgenogram of the same child. There is normal aeration of pulmonary tissue, no significant cardiac enlargement, and normal pulmonary vascularity.

color. She did not deteriorate until the fifth day of life when respiratory distress occurred and cyanosis deepened.

On the sixth day of life, she was dyspneic with a respiratory rate of 70 per minute. There was minimal peripheral cyanosis. The precordium was hyperactive with a right ventricular heave. A soft II/VI (Levine scale) ejection systolic murmur was heard at the left sternal border. A systolic ejection click was present in the pulmonary area. The liver was enlarged to 3 cm below the right costal margin in the midclavicular line.

*Circulation, Volume XXXV, February 1967*
Peripheral arterial pulses were of normal volume. The chest x-ray (fig. 1 left) showed marked overdistension of the thorax with low flat diaphragms. The heart was only slightly enlarged. The most striking features were marked pulmonary congestion and edema. There was a small amount of fluid in both pleural cavities and in the major fissure on the right.

The electrocardiogram (fig. 2) demonstrated an electrical QRS axis in the frontal plane of +135°. The precordial pattern was that of marked right ventricular hypertrophy with 2 mm q waves recorded in lead 4 V6, and 9 mm S waves in lead V6. The size and shape of P waves were not remarkable.

The child underwent limited cardiac catheterization, and a right ventricular angiogram was obtained. The left atrium, which seemed to be unusually small, was catheterized by way of the right atrium. The right ventricular cavity and the main pulmonary artery were moderately enlarged. During the venous phase, two main pulmonary venous trunks were demonstrated uniting into a common trunk in the left paravertebral gutter and continuing infradiaphragmatically (fig. 3). At no time was the left atrium visualized.

An emergency operation was performed on the sixth day of her life. The heart was exposed through a median sternotomy. The left atrium and ventricle appeared small. The right and left pulmonary venous trunks were joined behind the left atrium to form a single common trunk which ran caudal to penetrate the diaphragm just anterior to the vertebral bodies. Total cardiopulmonary bypass was instituted and the apex of the heart was tilted up. The pulmonary venous trunk was visualized and a side-to-side anastomosis 1.5 cm in diameter was constructed between it and the posterior surface of the left atrium. The common trunk was not narrowed or ligated below the anastomosis.

A selective angiogram of the pulmonary artery performed 6 months postoperatively showed a normal-sized left atrium and ventricle (fig. 4) with no suggestion of the previously present pulmonary venous trunk. Pressures and oxygen saturations obtained from the preoperative and postoperative cardiac catheterizations are shown in table 2.

Two years after operation, the patient is a thriving child, weighing 14.8 kg. There is no heart murmur and the second heart sound is normal in intensity. Her postoperative chest roentgenogram is shown in figure 1 b.

Her electrocardiogram (fig. 5) shows a significant decrease in the pattern of right ventricular hypertrophy with an rsR' pattern in V1, reduction in amplitude of R waves in the right chest leads, and S waves in the left chest leads.

**Discussion**

Burroughs and Edwards collected 14 cases of anomalous pulmonary venous drainage of the infradiaphragmatic type. No infant survived longer than 3 months. The hemodynamics of these cases were characterized by high resistance to flow determined not only

**Table 2**

<table>
<thead>
<tr>
<th>Site</th>
<th>Oxygen saturation (%)</th>
<th>Pressure (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preop 6-Mo. P.O.*</td>
<td>Preop 6-Mo. P.O.</td>
</tr>
<tr>
<td>Right atrium</td>
<td>— 78</td>
<td>2 0</td>
</tr>
<tr>
<td>Left atrium</td>
<td>— 100</td>
<td>3 5</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>— 80 80/2</td>
<td>37/4</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>— 76</td>
<td>— 37/13</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>— 74</td>
<td>— —</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>— 76</td>
<td>— —</td>
</tr>
</tbody>
</table>

*Ascorbic acid indicator-dilution curves showed no evidence of a left-to-right shunt postoperatively.*

*Circulation, Volume XXXV, February 1967*
ANOMALOUS PULMONARY VENOUS DRAINAGE

Figure 3

A single frame of the preoperative angiocardiogram is shown on the left. The infradiaphragmatic drainage of the common pulmonary venous trunk is visualized as indicated in the line drawing on the right.

by the long narrow venous trunk but often by the presence of a hepatic capillary bed between the pulmonary venous trunk and the inferior vena cava. The pulmonary venous hypertension leads to the characteristic clinical features of this condition.1, 6

Pulmonary venous congestion is related not only to venous outflow tract obstruction but also to pulmonary blood flow. This flow will depend on the size of the atrial septal defect which provides the only escape route from the lesser circulation. That this defect may be small is indicated by our successful operation in which the atrial septal defect was not closed, and yet no shunt is currently detectable. Furthermore, it is noteworthy that in none of the nine cases reviewed from our autopsy files was there a bona fide atrial septal defect. The communication in each heart was via the foramen ovale as it was in the experience of Burroughs and Edwards.2

Surgical technique has varied in the successful operations reported by others. Sloan, as quoted by Cooley and Balas5 found it necessary to enlarge the left atrium with a Teflon prosthesis. This was not necessary in our patient in whom the postoperative angiocardiogram showed a normal-sized left atrium. Cooley himself divided the common venous trunk, anastamosed its proximal end to the left atrium, and closed the atrial defect producing a one-stage correction of the lesion. In contrast, we followed the dictates of Woodwark and Mustard and their associates4, 7 who suggested a staged procedure leaving the ligation of the venous trunk and possible closure of the defect until a later operation. Indeed this second operation may
never be required as appears to be true of our patient. In our case the anastomosis between common pulmonary venous trunk and left atrium is adequate and the resistance to infradiaphragmatic flow is sufficient to direct all pulmonary venous blood into the left atrium. Furthermore, the septal defect was presumably a foramen ovale which now partitions the atria. It appears to us that infants who require operation can be expected to have foramen ovale communications not requiring closure.

Acknowledgment
We gratefully acknowledge the assistance of Dr. J. S. Fawcett in reviewing the pathological material.

References
Infradiaphragmatic Anomalous Pulmonary Venous Drainage: Normal Hemodynamics Following Operation in Infancy
W. JEGIER, E. CHARRETTE and A. R. C. DOBELL

Circulation. 1967;35:396-400
doi: 10.1161/01.CIR.35.2.396
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1967 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/35/2/396

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/