Common Pulmonary Vein Atresia

Premortem Diagnosis in Two Infants

By Richard E. Hawker, M.B., M.R.A.C.P.,
John M. Celermajer, M.B., F.R.A.C.P., Don C. Gengos, M.B., F.R.A.C.S.,
Timothy B. Cartmill, M.B., F.R.A.C.S.,
and J. Denby Bowdler, M.B., D.D.R.

SUMMARY
Two cases of common pulmonary vein atresia (CPVA) are presented. In one, the diagnosis was suspected on clinical grounds, and in both it was confirmed by cardiac catheterization and angiocardiography. Surgical correction was attempted unsuccessfully in one. Prompt identification of the defect followed by immediate operation, using a technic of profound hypothermia and circulatory arrest, offers hope of successful correction in this rare anomaly.

Additional Indexing Words:
Congenital heart disease Cardiac catheterization Hypothermia

The aggressive approach to diagnostic cardiac catheterization, which is essential for successful palliation of cyanosed neonates with transposition of the great arteries1 or with malformations resulting in low pulmonary blood flow,2,3 results in the early diagnosis of various other rare congenital heart lesions, some of which are also amenable to surgical palliation or correction. In the last 2½ years, two neonates with the rare condition of common pulmonary vein atresia (CPVA) have been studied at the Royal Alexandra Hospital for Children, in Sydney, Australia. The correct diagnosis was established by cardiac catheterization in both, and surgical correction was attempted in one.

Report of Cases

Case 1
This 3100-g baby boy was seen at Royal Alexandra Hospital for Children at the age of 22 hours, with cyanosis and tachypnea present since birth. Examination revealed diminished peripheral pulses, palpable right ventricular overactivity, and a soft systolic ejection murmur at the left sternal edge with a widely split second heart sound. The liver was distended 3 cm below the costal margin. Chest X-ray (fig. 1) showed a heart of normal size and gross pulmonary venous congestion. ECG showed right-axis deviation and slight right ventricular hypertrophy, but was considered within normal limits for age (fig. 2). A clinical diagnosis of the obstructed form of total anomalous pulmonary venous return was made.

Cardiac Catheterization
This was performed at 24 hours of age (table 1). The catheter was passed to the right heart chambers and main pulmonary artery, and to the descending aorta via a persistent ductus arteriosus. The left atrium and left ventricle were entered via a patent foramen ovale. No pulmonary veins could be probed. Oximetry showed that the blood oxygen saturation was similar (from 40 to 48%) in all cardiac chambers and in the superior and inferior vena cava, pulmonary artery, and descending aorta. The pressure in the right ventricle and pulmonary artery was slightly

From the Adolph Basser Institute of Cardiology, Royal Alexandra Hospital for Children, Sydney, Australia.

Address for reprints: Dr. J. M. Celermajer, Adolph Basser Institute of Cardiology, Royal Alexandra Hospital for Children, Pyrmont Bridge Road, Camperdown, 2050, New South Wales, Australia.

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Figure 1

Case 1. Heart size normal. There is increased pulmonary vascularity, venous in type, giving the lungs an almost miliary pattern. No evidence of pleural fluid or Kerley B lines.

Figure 2

Electrocardiogram of case 1.

higher than that in the left ventricle and descending aorta. Biplane angiocardiogram with pulmonary artery injection showed normal right and left pulmonary arteries and a large persistent ductus arteriosus with shunting from pulmonary artery into the descending aorta (fig. 3, top). Although the angiocardiogram was programmed for 7 sec, pulmonary veins were not visualized. Angiocardiogram with left atrial injection showed normal left atrium, left ventricle, and ascending aorta. A diagnosis of CPVA was made, but since the baby’s condition was extremely poor and pulmonary veins were not seen on the angiocardiogram surgery was not considered practicable. The baby died 32 hours after birth.

Autopsy Findings

The right and left heart chambers and great arteries were normal in size and position. There was no aortic valve stenosis or coarctation. A patent foramen ovale was present. The persistent ductus arteriosus was 6 mm in diameter and was equal in size to the aortic arch. No pulmonary veins were found entering the left atrium. Dissection showed that slightly small and fibrotic pulmonary veins joined to form a common channel 3 mm in external diameter across the back of the heart (fig. 4). An atretic fibrous strand was identified connecting the CPV to the left innominate vein. Microscopically the lungs
Table 1

Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Site</th>
<th>Case 1‡</th>
<th>Case 2‡</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Pressure (mm Hg)</td>
<td>Blood O₂ sat (%)</td>
</tr>
<tr>
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<td>48</td>
<td></td>
</tr>
<tr>
<td>IVC</td>
<td>40</td>
<td></td>
</tr>
<tr>
<td>RA</td>
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<td>48</td>
</tr>
<tr>
<td>RV</td>
<td>80/0/8</td>
<td>43</td>
</tr>
<tr>
<td>PA</td>
<td>73/45*; 59</td>
<td>43</td>
</tr>
<tr>
<td>LA</td>
<td>4</td>
<td>45</td>
</tr>
<tr>
<td>LV</td>
<td>60/0/6†</td>
<td>45</td>
</tr>
<tr>
<td>DA</td>
<td>65/45*†; 52</td>
<td>45</td>
</tr>
</tbody>
</table>

*Withdrawal curve.
†Not simultaneous.
‡The saturations in all chambers and great vessels are similar and pressure in pulmonary artery is higher than in descending aorta.

Abbreviations: SVC = superior vena cava; IVC = inferior vena cava; RA = right atrium; RV = right ventricle; PA = pulmonary artery; LA = left atrium; LV = left ventricle; DA = descending aorta.

showed lymphatic dilatation in the subpleural region and in the interlobar septae. There was intimal fibrosis of the pulmonary arteries, but no obvious pulmonary venous congestion. The alveolar walls were thin and avascular.

Case 2

This 2835-g baby girl, born at 36 weeks' gestation, had been cyanotic and tachypneic since birth and had required large amounts of sodium bicarbonate for correction of acidosis. She was transferred to Royal Alexandra Hospital for Children at 22 hours of age in a moribund condition with grade IV/IV cyanosis despite oxygen administration, and with grunting respiration and poor peripheral circulation. There was right ventricular overactivity and a widely split second heart sound, but no significant murmur. The liver was distended 3 cm below the costal margin. Chest X-ray (fig. 5) showed a normally sized cardiac silhouette and signs of gross pulmonary venous congestion. The electrocardiogram was reported to be within normal limits. It was considered that this infant had pulmonary venous hypertension and, in view of the early onset and severity of symptoms in association with gross cyanosis, CPVA was considered the probable diagnosis.

Cardiac Catheterization

This was performed at 24 hours of age and the findings were similar to those in the previous case (table 1). Pulmonary artery pressure was greater than aortic pressure, and oxygen saturation was similar in all chambers and in the great arteries and veins. Pulmonary veins were not catheterized, and a hand injection of contrast medium in the left ventricle showed left ventricle and aorta of normal size. The pulmonary artery angiogram showed normal right and left pulmonary arteries and a persistent ductus arteriosus, with shunting from pulmonary artery to aorta (fig. 3, bottom). Pulmonary veins were faintly outlined, and there was a suggestion of opacity representing the common pulmonary venous trunk posterior to the left atrium. CPVA was diagnosed and surgical correction attempted.

Operation

Via a median sternotomy, the ascending aorta and right atrium were cannulated and cardiopulmonary bypass instituted under normothermic conditions. The ductus arteriosus was ligated. The heart was retracted forward, and the common pulmonary venous channel identified behind the left atrium. Matching 1-cm incisions were made in the anterior aspect of the venous channel and the posterior aspect of the left atrium, and an anastomosis was fashioned between them. After cardiopulmonary bypass was discontinued, spontaneous ventilation did not occur, the pulses and peripheral perfusion were poor, and no urine was secreted. Despite infusion of sodium bicarbonate and isoproterenol, there was no improvement and the baby died 12 hours after operation.

Autopsy Findings

The cardiac chambers and great arteries were normal in size and position. There was no aortic stenosis or coarctation. Ductus was securely ligated. The anastomosis between the common
Figure 3

Angiocardiograms of case 1 (top) and case 2 (bottom). In both examinations injection of contrast material into main pulmonary artery resulted in opacification of the pulmonary arteries and, via the patent ductus arteriosus, the aorta. The degree of retrograde opacification of the aortic arch and ascending aorta is an indication of decreased cardiac output.
Figure 4
Autopsy specimen of case 1. The heart has been retracted forward to show the thickened fibrous common pulmonary vein (CPV) running across the midline. No vessel enters the left atrium (LA) which is of normal size. The left ventricle (LV) is well developed. A pattern of fine ridges is seen over the surface of the lungs.

pulmonary venous channel and the left atrium was patent and was 6–7 mm in diameter. No other connection between the pulmonary veins and any heart chamber or systemic vein was found. Microscopically the lungs showed marked dilatation of lymphatics and congestion of pulmonary veins.

Discussion
In 1962 Lucas et al.4 reported three cases of CPVA, and we can find reference to only two other cases which conform to the original description.5, 6 However, Hastreiter7 (two cases) and Hauck8 (one case) have reported a similar condition.

The common pulmonary vein forms as an outgrowth from the left atrium to meet the splanchnic plexus which drains the primitive lung to the cardinal and umbilicovitelline system of veins. The connections with the systemic circulation then regress and the common pulmonary vein is absorbed so that four pulmonary veins come to enter the left atrium separately. Atresia of the common pulmonary vein results from a failure of development of the common pulmonary vein at a relatively late stage, after the primitive connections between the splanchnic plexus and the systemic veins have regressed. Failure to develop at an earlier stage, while the connections are still present, leads to total anomalous pulmonary venous drainage.9

Patients with CPVA have survived for up to 28 days in the absence of a gross communication between the pulmonary veins and the heart or systemic circulation. Obviously, minute collateral vessels must have been present, and in one patient aged 3 days4 dilated bronchial veins and a collateral vessel anastomosing with varicose esophageal veins were demonstrated. No significant blood flow is possible through the fibrous strands connecting the confluence of pulmonary veins with right atrium,4 left atrium,7 or innominate vein.
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(our case 1). These strands, however, demonstrate the relationship between CPVA and total anomalous pulmonary venous return. It then becomes only a matter of degree whether the cases of Hastreiter⁷ and Hauck,⁸ which showed a confluence of pulmonary veins and a small accessory pulmonary vein draining one of the lobes of the lung to the portal vein or superior vena cava, are classified as atresia of the common pulmonary vein or obstructed form of anomalous pulmonary venous return.

The large lymphatics which are present in the fetus persist and help to remove the interstitial pulmonary edema.¹⁰ It is now recognized that CPVA is one of the causes of the pathologic condition congenital pulmonary lymphangiectasis.⁸

Neonates with CPVA display cyanosis and respiratory distress from the first day of life. With the exception of the wide splitting of the second sound the findings on physical examination are not helpful. The electrocardiogram was done in three of the five published cases and was reported to show abnormal right ventricular preponderance. In all, however, it was performed between 3 and 4 weeks of age. In one of these it was reported to be within normal limits at the age of 6 days. The chest X-ray showed gross pulmonary venous congestion with a diffuse pattern of pulmonary vascular markings that was either reticular or miliary in type in the four reported infants in whom it was performed, as well as in our two cases. There was no cardiac enlargement except in one infant surviving 28 days in whom it was slight, probably because pulmonary blood flow is very low and therefore volume load on the heart is not increased.

Cardiac catheterization reveals severe pulmonary hypertension with right-to-left shunting through the ductus arteriosus and foramen ovale. No site of entry for pulmonary venous blood into the circulation can be identified, so oxygen saturations in the great veins, left and right heart chambers, and great arteries are all similar.

Angiocardiography is the most useful diagnostic procedure. Pulmonary arteriogram shows persistence of contrast medium in the pulmonary arteries, and neither the left atrium nor the great veins (nor the right heart chambers) are opacified. Because of the very low flow through the lungs, pulmonary veins are opacified poorly or not at all. A hand injection of contrast medium in the left atrium is sufficient to exclude a lesion of the left heart and establish that the left heart chambers and aorta are normally developed. Our cases, as well as the only two previously reported in whom cardiac catheterization was carried out, showed right-to-left shunting through the ductus. In all infants the ductus arteriosus was widely patent at autopsy.

A diagnosis of CPVA should be considered in any infant who presents soon after birth with severe cyanosis, tachypnea, and radiologic signs of severe pulmonary congestion without cardiomegaly. Preoperative diagnosis can be suggested by cardiac catheterization although it may not be possible to demonstrate the pulmonary venous channel angiographically.

Surgical correction of this lesion is theoretically possible. One infant⁷ had a thoracotomy, but the chest was closed after the pulmonary venous drainage was misinterpreted as being normal. Case 2 of this paper is the only infant in whom anastomosis of the common pulmonary venous trunk and left atrium has been performed.

If the pulmonary venous channel is found at exploration it is relatively easy to anastomose it to the left atrium since the two structures lie close together. However, infants with severe pulmonary venous obstruction such as that due to infradiaphragmatic anomalous pulmonary venous return are usually acidemic and have such poor pulmonary function that they rarely survive operation, despite satisfactory anatomic correction.

In retrospect, ligation of the ductus arteriosus in the second case may have been an error. This, or the patent foramen ovale, might be left as a means of decompressing the right heart until the pulmonary arteriolar resistance which is elevated in response to the obstruction to the pulmonary veins begins to fall.
Because of the need to define the anatomic details of the anomaly at operation a median sternotomy incision is essential. If repair is judged to be possible, complete circulatory arrest with profound hypothermia should afford optimal exposure and operating conditions.

Our present opinion is that hypothermia can be safely achieved by whole-body perfusion using a pump oxygenator and an extracorporeal heat exchanger with early venting of the common pulmonary venous trunk. Alternatively, surface cooling could be utilized.11

Thus, successful correction of this rare lesion will depend upon prompt and correct diagnosis achieved by cardiac catheterization and angiography as described in these two cases, followed by immediate operation utilizing profound hypothermia with complete circulatory arrest.

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References

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