Case Reports

Levocardia with Anomalous Venous Drainage
A Case with Successful Surgical Correction

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SUMMARY
A case of levocardia with anomalous venous drainage of all right pulmonary veins to the right atrium, a sinus venosus type atrial septal defect, an anomalous inferior vena cava with azygos continuation, alterations of sinoatrial rhythm, associated with partial situs inversus viscerum was reported. The physiologic abnormalities of the heart were corrected successfully. A brief review on the other 8 reported patients of levocardia with complicated cardiac malformations treated surgically is presented.

Additional Indexing Words:
Partial anomalous pulmonary venous drainage Sinus venosus defect
Azygos continuation Coronary sinus rhythm Left atrial rhythm

When the azygos system does allow the usual rightward position of the systemic venous atrium in a patient with levocardia, the associated intracardiac defects may be simple and surgically remediable. Successful surgical correction of atrial septal defect with pulmonary venous drainage into the right atrium in levocardia has been reported very rarely, and it prompted this case report.

CASE REPORT

A 6-year-old girl was admitted to the hospital on December 16, 1968, for evaluation of a heart murmur, which was noticed 8 months prior to admission. Birth and developmental history were normal. One of her 5 siblings died of cyanotic congenital heart disease at the age of 3 months.

On admission, there were no signs of cyanosis, clubbing or dyspnea. The vital signs were within normal limits. The pertinent findings included: the cardiac dullness in a normal location without thrills. The cardiac apex beat was in the fifth intercostal space at the left midclavicular line. There was a grade 4/6 harsh
ejection systolic murmur best heard at the third left intercostal space parasternally; the second sound was split and accentuated at the second left interspace. The stomach tympany was on the right side, and the liver dullness on the left. A phonocardiogram confirmed the above auscultatory findings. The chest roentgenogram (Fig. 1) revealed a normal position of the heart, moderate cardiac enlargement, slightly increased pulmonary vascular markings, and the stomach bubble below the right diaphragm. An electrocardiogram (Fig. 2) revealed ectopic atrial rhythm with inverted P waves in leads I, II, III, aVr and V6 in the first tracing, but the subsequent ones showed coronary sinus rhythm with inverted P waves in I, II, III and aVr. In addition, there was right ventricular hypertrophy and rsR' pattern in lead V1. When cardiac catheterization was first attempted via the right saphenous vein, the catheter tip was advanced through the azygos vein into the superior vena cava, the right atrium and then to the right ventricle, but the pulmonary artery could not be entered. Venous angiograms showed a "candy cane" appearance of the anomalous vein, and right-sided venous atrium and ventricle (Fig. 3). The hemodynamic data at the second catheterization via the left arm vein are

![Fig. 1. Roentgenogram of chest. Posterior-anterior view. Note position of the stomach bubble.](image)

![Fig. 2. Preoperative electrocardiogram showing ectopic atrial rhythm, right ventricular hypertrophy and rsR' pattern in lead V1.](image)
Fig. 3. Early phase of venous angiography showing an anomalous inferior vena cava with azygos continuation and right-sided venous atrium.

Table I. Preoperative Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Observation sites</th>
<th>Blood oxygen saturation (%)</th>
<th>Intravascular pressures (mm.Hg)</th>
<th>Appearance time (sec.)</th>
<th>Dye dilution curves</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Phasic</td>
<td>Mean</td>
<td></td>
<td>left-to-right</td>
</tr>
<tr>
<td>Left pulmonary wedge</td>
<td>90.3</td>
<td>25/16</td>
<td>19</td>
<td>No</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>88.5</td>
<td>30/20</td>
<td>22</td>
<td>Small</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>96.2</td>
<td>40/2</td>
<td>16</td>
<td>Moderate</td>
</tr>
<tr>
<td>Right atrium</td>
<td>95.0</td>
<td>5/0</td>
<td>2</td>
<td>Moderate</td>
</tr>
<tr>
<td>Right pulmonary vein</td>
<td>71.0</td>
<td>50/10</td>
<td>3.0</td>
<td>No</td>
</tr>
<tr>
<td>Common hepatic vein</td>
<td>81.2</td>
<td>106/60</td>
<td></td>
<td>Moderate</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td></td>
<td></td>
<td></td>
<td>No</td>
</tr>
<tr>
<td>Left brachial vein</td>
<td></td>
<td></td>
<td></td>
<td>Moderate</td>
</tr>
<tr>
<td>Femoral artery</td>
<td></td>
<td></td>
<td></td>
<td>Moderate</td>
</tr>
</tbody>
</table>

summarized in Table I. It revealed a moderately large left to right shunt at the atrial level. The pulmonary-systemic flow ratio was 3:1. The pulmonary arterial pressure was slightly elevated. The catheter entered a right pulmonary vein. Dye curves obtained by ear piece after injection of indicator into the left pulmonary artery revealed normal appearance, suggesting normal drainage of the left pulmonary veins into the left atrium. Upper gastrointestinal series showed inversion of the stomach and duodenal loop. Barium enema revealed the entire colon to be in the
left half of the abdomen and a midline cecum (Fig. 4). A radio isotope scanning of the spleen, liver and kidneys was carried out by the intravenous injection of 50 $\mu$C of MHP ($\text{Hg}^{203}$). The spleen was located on the right side (Fig. 5) and the liver on the left. The kidneys were normal. Blood smears revealed no Heinz or Howell-Jolly bodies.

Although patients with levocardia have usually multiple physiologic cardiovascular abnormalities, preoperative diagnosis of this patient was only partial anomalous pulmonary venous drainage into the right atrium with atrial septal defect, which are amenable to surgery.

At operation on February 4, 1969, the cardiac chambers and great vessels had a normal solitus position. The pulmonary artery was dilated, being 3.0 cm.
in diameter, but the aorta was 1.0 cm. Under deep hypothermia induced by surface cooling, occlusion of the superior vena cava, the right superior pulmonary vein and the common hepatic vein were done, as well as the aorta and the pulmonary artery trunk. The right atrium was opened, and a sinus venosus type atrial septal defect (1.5 × 1.5 cm.) was found. Both (superior and inferior) right pulmonary veins were seen to enter the right atrium. A large common hepatic vein drained into the right atrium simulating the inferior vena cava. The atrial septal defect was enlarged caudally and then closed by means of a Teflon patch which diverted the entrance of the right pulmonary veins to the left side, and the physiologic abnormalities of the heart were completely corrected (Fig. 6). Duration of occlusion was 31 min. at 24°C. Blood loss during the operation was 450 ml. Recovery was uneventful.

Postoperatively normal intracardiac pressures and oxygen saturation values were obtained. A postoperative electrocardiogram showed marked inversion of the T waves over the left side of the hemithorax with inverted P waves in leads II, III and aV_F (Fig. 7).

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**Fig. 6.** Left, showing anomalous pulmonary venous drainage into the right atrium with a sinus venosus defect. Right, complete surgical correction by means of Teflon patch (5 × 3 cm.). SVC = Superior vena cava; Az = Azygos vein; ASD = Atrial septal defect; PV = Pulmonary veins; CS = Coronary sinus; CHV = Common hepatic vein.

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**Fig. 7.** Postoperative electrocardiogram showing coronary sinus rhythm and giant negative T waves in leads V_4-6.
Levocardia with total or partial inversion of abdominal viscera is a rare condition commonly associated with complicated cardiac malformations, and has been estimated to occur in approximately 1 per cent of all cases of congenital heart disease. Although most patients with levocardia show cyanosis and early fatal outcome, the prognosis is obviously better in the absence of cyanosis. Only 25 per cent of patients with levocardia survive the first year of life, and only 6 per cent of them live beyond 5 years.

This case of levocardia was associated with several cardiovascular abnormalities and ectopic atrial rhythm. Very similar cases had been reported rarely in literature. These defects are relatively uncommon but have been reported singly or associated with other cardiovascular malformations, and asplenia or polysplenia. In asplenia, wherein pulmonary stenosis and defects of the ventricular septum are common, cyanosis and diminished pulmonary blood flow are usually present. In polysplenia, the presence of septal defects and acyanosis with increased pulmonary blood flow form the usual clinical pattern. We could find the presence of the spleen by means of angiography and radioactive isotopic visualization. Considering the cardiovascular abnormality, the present case probably belongs to the polysplenic ones advocated by Moller et al., but no surgical exploration was performed.

The triad of anomalous inferior vena cava draining into the superior vena cava by way of azygos continuation, high atrial septal defect and alterations in sinoatrial rhythm was described by Hastreiter et al. as a syndrome and probably represent a mild form of cardiovascular malformation associated with visceral heterotaxia. On the other hand, the prevailing elements of the complex cardiac anomalies with a similar pattern in levocardia advocated by Ongley et al. are (1) either total or partial situs inversus, (2) multiple spleens, and (3) abnormalities of veins in the form of (a) pulmonary venous drainage to the venous atrium, and (b) the inferior vena caval system joining the azygos system. An atrial septal defect is usually present. Ventricular septal defects, pulmonic stenosis and transposition of the great vessels may be associated. The prognosis for patients with these anomalies is poor.

For cases of cyanotic levocardia a shunt operation has been performed for the relief of inadequate pulmonary blood flow, but the operative mortality is relatively high. However, simple or mild malformations may be surgically remediable in patients with levocardia. From the literature successful surgical correction of defects in patients with levocardia were reported by Leachman et al., Creech et al., Ongley et al., Hastreiter et al., and Niihori et al. (Table II). All these patients have atrial septal defect as-
Table II. Successfully Corrected Patients with Levocardia

<table>
<thead>
<tr>
<th>Author</th>
<th>Data</th>
<th>Age at operation</th>
<th>Corrected cardiovascular anomalies</th>
<th>IVG drainage and other systemic venous anomalies</th>
<th>Presence of spleed</th>
<th>Cyanosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leachman4)</td>
<td>1964</td>
<td>Adult</td>
<td>ASD (Ostium II).</td>
<td>Azygos continuation</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>2y</td>
<td>ASD (Ostium II).</td>
<td>Azygos continuation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Creech17)</td>
<td>1964</td>
<td>13y</td>
<td>Common atrium. TAPVD.</td>
<td>left-sided IVG. right-sided hepatic veins. (right SVC.) TAPVD to left SVC.</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Ongley4)</td>
<td>1965</td>
<td>8½y</td>
<td>Common atrium. MI (a cleft in the anterior leaflet).</td>
<td>left-sided IVG. left SVC. (right SVC.)</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Hastreiter2)</td>
<td>1968</td>
<td>9y</td>
<td>ASD (Fossa ovalis type). PAPVD.</td>
<td>Normal drainage of left-sided IVG. left SVC. PAPVD to right atrium.</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Niihori8)</td>
<td>1969</td>
<td>13y</td>
<td>ASD (Ostium I).</td>
<td>Azygos continuation</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>12y</td>
<td>Common atrium.</td>
<td>Azygos continuation</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>3y</td>
<td>ASD (Patent foramen ovale).</td>
<td>Azygos continuation. Hemiazygos continuation left SVC.</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Iwabuchi</td>
<td>1970</td>
<td>6y</td>
<td>ASD (Sinus venosus type). PAPVD.</td>
<td>Azygos continuation. PAPVD to right atrium.</td>
<td>Yes</td>
<td>No</td>
</tr>
</tbody>
</table>

TAPVD: Total anomalous pulmonary venous drainage; PAPVD: Partial anomalous pulmonary venous drainage; MI: Mitral insufficiency; IVG: Inferior vena cava.

associated with azygos or hemiazygos continuation to the right (venous) or common atrium, and may have spleen or not, with or without cyanosis. In patients with levocardia, if an anomalous inferior vena cava with azygos continuation occurs as the principal or only avenue of venous return from below the diaphragm, the situs of the venous atrium is positioned to the right,1),4) and the associated intracardiac defects may be simple and surgically remediable,1),8) or isolated and not requiring correction.

Angiocardiography is the most reliable method short of surgical exploration or necropsy for identifying the situs of the atria; and electrocardiography is useful, too. The normal position of the atria is suggested by the finding of an upright P wave in lead I of the electrocardiogram, since there is usually inversion of the P wave in this lead in cases in which the superior vena cava and the venous atrium lie on the left side of the heart.18) Such P wave abnormalities, however, are not a consistently reliable criteria of atrial orientation.3) Intermittent inverted P wave in lead I as seen in Padmavati's case19) and
ours without transposition of the atria, or nodal rhythm, which is common in levocardia with corrected transposition, may occur. The electrocardiogram (Fig. 2) suggests left atrial rhythm advocated by Mirowski\(^{20}\) based on inverted P wave in leads I and V\(_6\). But the diagnosis of left atrial rhythm by the electrocardiogram is still troublesome.\(^{21},^{22}\) The postoperative electrocardiogram (Fig. 6) showed upright P\(_1\) and inverted P\(_{II}\) and P\(_{III}\), which is a common pattern in isolated levocardia, and looks like coronary sinus rhythm advocated by Scherf et al.\(^{23}\) Coronary sinus rhythm was found frequently in patients with sinus venous defect, persistent left superior vena cava or azygos continuation.\(^{24}-^{26}\) In addition, the presence of an rsR' pattern in lead V\(_1\) is most frequently associated with drainage of the anomalous pulmonary veins into the superior vena cava or right atrium.\(^{21}\) Considering these electrocardiographic findings, our case might represent typical electrocardiograms of levocardia with partial anomalous pulmonary venous drainage and sinus venous defect without transposition of the atria.

Dye dilution technique is very useful for identifying whether the catheter in the pulmonary vein entered from the right atrium through the atrial septal defect or directly. In patients with partial anomalous pulmonary venous connection and sinus venous defect, normal dye curves are usually obtained when the dye is injected into the left pulmonary artery as the cases of Braunwald et al.\(^{27}\) and ours. Nevertheless, preoperatively we could suspect that our patient had atrial septal defect because of a minimal right to left shunt curve recorded after injection of the dye into the right atrium.

Anomalies of the systemic venous return may impose some restrictions on their cannulation when cardiopulmonary bypass is used, that is of no concern when simple deep hypothermia is applied. In any event, accurate preoperative anatomic as well as hemodynamic diagnosis is of prime importance for adequate management of these patients.

References