Right Axis Deviation, Clockwise QRS Loop, and Signs of Left Ventricular Underdevelopment in a Child with Complete Type of Persistent Common Atrioventricular Canal

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The electrocardiogram has proved to supply reliable information for clinical recognition of cardiac defects of the type known as complete form of persistent common atrioventricular canal. The electrocardiographic findings consistently associated with these defects have been left axis deviation and QRS vectors in the frontal plane which describe a counterclockwise loop above the isoelectric point, or a figure-of-eight loop along the horizontal line. These patterns are thought to be a result of an abnormality involving the ventricular conduction pathways. These electrocardiographic findings have been described with varying frequency in other cardiovascular anomalies, including instances of the ostium secundum type of atrial defect. However, anatomically proved cases of complete persistent common atrioventricular canal unattended by the characteristic electrocardiogram are rare. The case to be presented here is such a one, but includes two unusual features, namely, left ventricular hypoplasia and obstruction to left ventricular outflow. The electrocardiogram revealed right axis deviation and a clockwise inscription of the QRS vector loop in the frontal plane. Of additional interest in this case were features that suggested underdevelopment of the left ventricle.

Case Report

A 2-year-old girl was admitted to the Children's Orthopedic Hospital at the age of 6 months. A heart murmur was first detected in

Figure 1

Left. Posteroanterior roentgenogram of the chest revealing cardiomegaly, prominence of the pulmonary artery segment, and increased pulmonary vascularity. Right. Frontal view of the heart and great vessels at autopsy. The arrows point to the interventricular sulcus, which is displaced superiorly and laterally. R.V., right ventricle; L.V., left ventricle.
early infancy and chronic congestive heart failure became manifest at 5 months. Physical examination revealed a relatively well-nourished child who had tachycardia and mild respiratory distress. She had a left precordial bulge, hyperactive precordium, loud second heart sound, systolic thrill, and a grade-IV/VI long, harsh systolic murmur best heard along the left sternal border, and a mid-diastolic rumbling murmur localized between the apex and the lower left

Table 1
Data Obtained at Cardiac Catheterization Demonstrating Left-to-Right Shunting at Atrial and Ventricular Levels, Pulmonary Hypertension, and Mild Systemic Arterial Desaturation

<table>
<thead>
<tr>
<th>Location</th>
<th>O₂ Content (vol. %)</th>
<th>% O₂ saturation</th>
<th>Pressure, mm Hg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>5.4</td>
<td>34</td>
<td>—</td>
</tr>
<tr>
<td>Right atrium, mid</td>
<td>9.5</td>
<td>56</td>
<td>—</td>
</tr>
<tr>
<td>Right atrium, low</td>
<td>11.4</td>
<td>62</td>
<td>—</td>
</tr>
<tr>
<td>Right ventricle, inflow</td>
<td>11.6</td>
<td>63</td>
<td>—</td>
</tr>
<tr>
<td>Right ventricle, outflow</td>
<td>13.6</td>
<td>80</td>
<td>62/8</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>13.3</td>
<td>78</td>
<td>50/20</td>
</tr>
<tr>
<td>Pulmonary artery wedge</td>
<td>15.3</td>
<td>90</td>
<td>72/50</td>
</tr>
</tbody>
</table>

Pulmonary blood flow—6.2 L/min/M²
Systemic blood flow—1.9 L/min/M²

sternal border. The chest roentgenograms provided evidence (fig. 1, left) of cardiomegaly and increased pulmonary vascularity. Cardiac catheterization data (table 1) indicated the presence of left-to-right shunting at atrial and ventricular levels, and pulmonary hypertension. Mild systemic arterial desaturation was thought to be due to respiratory depression. Because of the character of the electrocardiographic evidence, a diagnosis was made of ventricular septal defect and coexistent ostium secundum-type atrial septal defect rather than of persistent common atrioventricular canal (fig. 2). Because the child's heart failure could not be controlled by medical measures, surgical treatment was undertaken when the patient was 2 years of age. The patient died early in the postoperative period.

Postmortem examination revealed hypoplasia of the left ventricle, fibrous bands obstructing the left ventricular outflow tract, and complete persistent common atrioventricular canal (fig. 1, right, and fig. 3). Moderate hepatomegaly was present. The lungs on gross examination were relatively normal. Microscopic examination, however, revealed mild hemorrhage and edema and early pulmonary vascular disease.

Discussion

In the case presented here complete persistent common atrioventricular canal existed

Figure 2
Twelve-lead scalar electrocardiogram demonstrating right axis deviation and right ventricular overload. Note the apparent clockwise direction of the QRS inscription in the frontal plane.
unattended by electrocardiographic findings typical of such a lesion. Awareness of this possibility is of importance when considering surgical treatment for a patient who has left-to-right shunting at both atrial and ventricular levels. Of additional interest was the existence of left ventricular hypoplasia and dominant left-to-right shunting at ventricular level in the absence of electrocardiographic evidence of left ventricular overload. Finally, as a result of underdevelopment of the left ventricle, evidence of posterior, superior, and leftward displacement of the interventricular sulcus was observed on the chest roentgenogram. Recognition of the latter two circumstances in a patient with congenital heart disease should alert the physician to the need for angiocardiographic study of the left ventricle.

Summary

Clinical data and findings at necropsy are presented on a 2-year-old girl who had persistent complete common atrioventricular canal, hypoplastic left ventricle, and obstruction to the left ventricular outflow tract. Electrocardiographic findings were atypical of common atrioventricular canal in that right axis deviation and a clockwise inscription of the QRS loop in the frontal plane were present. Findings in this case suggest that underdevelopment of the left ventricle should be suspected when (1) displacement of the interventricular sulcus is noted on roentgenographic examinations and (2) predominant left-to-right shunting at ventricular level is found in the absence of electrocardiographic evidence of left ventricular overload.

Acknowledgment

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References

Pathology of Angina Pectoris

The association of coronary disease with angina was first recognized by Edward Jenner from post-mortem examination, though it is possible that John Hunter, on whose account, as his anginal symptoms dated from 1773, Jenner kept silence, knew or suspected it in 1776 when John Fothergill published a fatal case of angina in which at the post-mortem Hunter found that "the two coronary arteries from origin to many of their ramifications on the heart were become one piece of bone." Jenner, who is said to have diagnosed angina in Hunter in 1777, never directly published anything on this subject, but he communicated his opinions to C. H. Parry, who in 1788 read a paper, "An Inquiry into the Symptoms and Causes of the Syncope Anginoso, Commonly called Angina Pectoris; illustrated by Dissections," to a small medical society in Gloucestershire of which Jenner was a member, and came to the conclusion that coronary disease was the cause. In this paper, not published until eleven years later, he quoted the case of ossification of the coronary arteries published by Black of Newry in 1795 and pointed out that he and Jenner had independently come to the same opinion in 1788.—Sir Humphry Davy Rolleston. The Harveian Oration. Great Britain, Cambridge University Press, 1928, p. 88.
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