Origin of Both Great Vessels from the Right Ventricle

I. Without Pulmonary Stenosis

By Henry N. Neufeld, M.D., James W. DuShane, M.D., Earl H. Wood, M.D., John W. Kirklin, M.D., and Jesse E. Edwards, M.D.

Among the congenital malformations in which an abnormal relationship exists between the aorta and pulmonary trunk is that in which both of these vessels arise from the right ventricle. The only outlet for the left ventricle is a ventricular septal defect. Right ventricular infundibular stenosis may or may not be associated. These cases apparently are extremely rare,\textsuperscript{1,3} and only a few have been reported in the last 25 years.\textsuperscript{4-7} A few cases were documented in the last century.\textsuperscript{1,3}

Many terms have been used for this malformation, including "partial transposition,"\textsuperscript{7}\"double-outlet right ventricle,"\textsuperscript{7} and "origin of both great vessels from the right ventricle."\textsuperscript{8}

Witham,\textsuperscript{7} in 1957, summarized six cases reported in detail in the literature and added four cases. He divided these cases into two groups and called them the "Fallot type" and the "Eisenmenger type," depending on the presence or absence of pulmonary stenosis.

One of the patients in his study was seen at the Mayo Clinic.\textsuperscript{9} This case was not included in the present analysis because additional multiple intracardiac malformations were present, and it appeared to us that different embryologic as well as hemodynamic problems were involved.

In a study of the pathologic and clinical material from the Mayo Clinic, we found 14 cases in which both great vessels originated from the right ventricle. Three main categories of this malformation occurred as follows: (1) without pulmonary stenosis (eight cases); (2) with pulmonary stenosis (five cases) and (3) with other intracardiac malformations (one case).

The purpose of this investigation is to correlate the anatomic findings (pathologic-anatomic or surgical) with the clinical and hemodynamic data and to discuss the possibilities of clinical diagnosis.

The three groups, although probably related embryologically, present different anatomic, clinical, hemodynamic, and surgical problems; therefore, we will discuss them in two separate communications.

The present paper is concerned with the eight cases of this malformation without pulmonary stenosis. The diagnosis in five instances was proved at necropsy, whereas it was established during operation in the other three. In two cases, patent ductus arteriosus, mitral insufficiency, and some degree of coarctation of the aorta were present. Pulmonary hypertension was present in all eight cases.

Pathologicoanatomic Features

The anatomic features in the five cases studied at necropsy fell into two subgroups that could be distinguished from each other by the relationship of the aortic and mitral valves. In the first subgroup (three cases), these two valves lacked the element of continuity found in normal hearts; the second subgroup (two cases) was marked by continuity between aortic and mitral valvular tissue.

In the first subgroup, the ascending aorta and the pulmonary artery appeared from the exterior to have a normal relationship, the ascending aorta lying to the right of the pulmonary trunk. The interior of the heart, however, revealed a profound abnormality of the aorta. Instead of extending inferiorly from the base of the heart to a position

From the Mayo Clinic and the Mayo Foundation, Rochester, Minnesota.

Supported in part by research grants no. H4014 and no. H3532 from the National Heart Institute, U. S. Public Health Service.
dorsal and caudal to the pulmonary valve, the aortic orifice communicated only with the right ventricle and lay to the right of and in the same coronal body plane as the pulmonary valve (figs. 1 and 2).

The interior of the right ventricle showed a prominent horizontal crista supraventricularis, with the aortic orifice lying between two branches of this structure. A ventricular septal defect was present in the basal part of the septum, caudal to the crista supraventricularis. Dorsally, this defect was bounded by the junction of the anterior mitral leaflet and the septal tricuspid leaflet. The caudal, ventral, and cephalad edges of the defect were composed of muscle; the muscle forming the cephalad boundary was the crista supraventricularis. By means of right ventricular landmarks, the position of the ventricular septal defect could be identified as lying dorsally and caudal to the crista supraventricularis and being caudal to the tricuspid valve.

The anatomic relationships of the mitral valve were abnormal because of the aforementioned lack of continuity between the aortic and mitral valves. Although the basal attachments of the posterior mitral leaflet were normal, those of the anterior leaflet were abnormal. The main basal attachment of the anterior mitral leaflet was to the septal tricuspid leaflet, with which, as already noted, it was continuous behind the septal defect. Anteriorly, the base of the anterior mitral leaflet attached to the muscle forming the superior boundary of the ventricular septal defect (fig. 3).

Two of the three hearts in this first subgroup corresponded to the above description. A second type in this first subgroup of cases was represented by one case (fig. 4). In this instance, the ventricular septal defect was cephalad to the crista supraventricularis and beneath the pulmonary valve. The aorta arose entirely from the right ventricle, being to the right of and slightly dorsal to the pulmonary valve. The aortic and pulmonary valves were in the same cross-sectional body plane. The ventricular septal defect was separated from the tricuspid valve by the crista supraventricularis, which lay in a horizontal position. The septal defect was surrounded on all sides by muscle. Dorsally, this muscle made up the left extremity of the crista supraventricularis.

The posterior mitral leaflet had normal attachments. The anterior mitral leaflet was attached to the posterior edge of the ventricular septal defect, being separated from the aortic valve and the tricuspid valve by the aforementioned muscle, which formed the posterior wall of the ventricular septal defect.

From the viewpoint of anatomic classification, it is of significance in these cases that, if an incision is made through the left ventricular wall into the aorta, one initially gains the impression that the aorta is in direct continuity with the left ventricle. However, this is an illusion since, in order to reach the aorta from the left ventricle, one must pass through the ventricular septal defect (fig. 5).
Case 3. First subgroup of origin of both great vessels from right ventricle without pulmonary stenosis and with ventricular septal defect. a. Outflow portion of right ventricle and great vessels. The ascending aorta (A.) lies beside the pulmonary trunk (P.T.). Exteriorly, the relationship between the two great vessels did not appear abnormal. A probe lies in a patent ductus arteriosus. b. Interior of right ventricle and origin of pulmonary trunk (P.T.). Between the two limbs of the crista supraventricularis (C.S.) is a channel leading to the origin of the aorta. The ventricular septal defect lies below the lower limb of the crista. c. Interior of right ventricle in similar perspective to that in b. A sagittal section has been made through the valves and great vessels. The ventral limb of the crista supraventricularis has been removed, leaving the dorsal limb (C.S.). The aortic valve (A.V.) lies to the right of and in the same body plane as the pulmonary valve (P.V.). The ventricular septal defect (D.) lies dorsal to both the aortic and pulmonary valves and below the crista supraventricularis. d. Section made in right ventricle in c carried across ventricular septum and through lateral wall of left ventricle, exposing left ventricular cavity (L.V.). The ventricular septal defect lies between the arrows. It is the only outlet for the left ventricle and it leads directly into the right ventricle (R.V.).

As already noted, the second subgroup of these five cases studied at necropsy comprises the two cases in which the aortic and mitral valves were continuous. This normal relationship existed not by virtue of a normal position of the aortic valve but as the result of an unusually long anterior mitral leaflet. The latter ascended through the ventricular septal defect to join the aortic valve. The latter lay entirely over the right ventricle and to the right of the ventricular septal defect.

From the exterior, the relationships of the aorta and pulmonary trunk appeared to be normal (fig. 6). The interior of the right ventricle revealed a large ventricular septal defect dorsal to the horizontal crista supraventricularis. In the usual instance in which a ventricular septal defect lies dorsal to the crista, the defect also is caudal to the septal leaflet of the tricuspid valve. In these two cases, the relationship of the defect to the tricuspid valve was unusual. It was not related closely to the septal leaflet of the tricuspid valve but instead lay cephalad and medial to the commissure between the anterior...
and septal tricuspid leaflets. The defect in one case extended upward toward the right to lie cephalad to the major portion of the anterior tricuspid leaflet (fig. 6a).

Forming the lower dorsal boundary of the ventricular septal defect was the anterior leaflet of the mitral valve where it was continuous with the septal tricuspid leaflet.

Cephalodorsally, the anterior mitral leaflet extended upward to form the cephalodorsal boundary of the ventricular septal defect, and then it joined the root of the aorta and the aortic valve.

When the left ventricular incision was extended into the aorta during dissection of these hearts, the incision cut through the elongated anterior mitral valve and aorta. Muscular tissue did not intervene between these two valvular structures (fig. 7), as was present in the second type of the first subgroup of hearts.

It might be asked justifiably how the hearts of the second subgroup differ from those with the common variety of ventricular septal defect. The following differences may be pointed out: In the hearts of this subgroup, the aorta does not connect directly with the left ventricle or straddle the ventricular septal defect; rather, it lies to the right of the defect. The aortic valve connects with

Circulation, Volume XXIII, March 1961
Case 1. Second subgroup of origin of both great vessels from right ventricle without pulmonary stenosis. a. Interior of right ventricle. The ventricular septal defect (D.) is separated from the pulmonary valve (P.V.) by the horizontal crista supraventricularis (C.S.). The defect, although lying posteroinferior to the crista, has an abnormal relationship to the tricuspid valve, lying ventral to the anterior tricuspid leaflet (A.T.). The aortic valve lies immediately above the defect and communicates entirely with the right ventricle, although it has continuity with the anterior leaflet of the mitral valve. b. Left ventricle and aortic valve (A.V.). The dorsal wall of the ventricular septal defect (D.) is formed by the anterior leaflet of the mitral valve (A.M.) as it becomes continuous with the septal leaflet of the tricuspid valve below and with the aortic valve above. Continuity is present between aortic valvular tissue and tissue of the anterior leaflet of the mitral valve, but it does not result from a normal position of the aorta but rather because of an abnormally long anterior mitral leaflet, which ascends through the ventricular septal defect to become continuous with the aortic valve. An additional abnormality is that the portion of the heart in which the aortic and mitral valves are continuous has an epicardial relationship; in normal hearts, on the contrary, the zone of continuity between the mitral and aortic valves is entirely intracardiac.

Clinical Findings
The clinical findings (table 1) were similar to those in cases of ventricular septal defect with associated pulmonary hypertension.
Table 1

Clinical Data in Eight Cases of Origin of Both Great Vessels from Right Ventricle

<table>
<thead>
<tr>
<th>Case</th>
<th>Age, yrs.</th>
<th>Sex</th>
<th>Cyanosis</th>
<th>Shortness of breath</th>
<th>Congestive failure</th>
<th>Respiratory infections</th>
<th>Systolic thrill, location</th>
<th>Systolic murmur, grade and location†</th>
<th>Other murmurs; type, grade and location</th>
<th>Second pulmonic sound</th>
<th>Blood pressure, mm. of Hg</th>
<th>Femoral pulses</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>14</td>
<td>F</td>
<td>—</td>
<td>+</td>
<td>—</td>
<td>+</td>
<td>LICS (2-4)</td>
<td>III (2-4)</td>
<td>—</td>
<td>++ + Split</td>
<td>98/68</td>
<td>+</td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>F</td>
<td>—</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Left sternal border</td>
<td>III (3-4)</td>
<td>Mid diastolic</td>
<td>++ + Split</td>
<td>92/60</td>
<td>+</td>
</tr>
<tr>
<td>3</td>
<td>15</td>
<td>F</td>
<td>—</td>
<td>+</td>
<td>—</td>
<td>+</td>
<td>LICS (2-3)</td>
<td>II (2-4)</td>
<td>SM, I, apex; DM, I, apex</td>
<td>++</td>
<td>110/60</td>
<td>+</td>
</tr>
<tr>
<td>4</td>
<td>6</td>
<td>M</td>
<td>When crying</td>
<td>+</td>
<td>—</td>
<td>+</td>
<td>Suprasternal notch</td>
<td>III (3-4)</td>
<td>SM, I, apex</td>
<td>+++</td>
<td>94/60</td>
<td>±</td>
</tr>
<tr>
<td>5</td>
<td>2</td>
<td>M</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>LICS (3-4)</td>
<td>IV (3-4)</td>
<td>DM, I, LICS</td>
<td>+++</td>
<td>90/50</td>
<td>+</td>
</tr>
<tr>
<td>6</td>
<td>9</td>
<td>M</td>
<td>—</td>
<td>+</td>
<td>—</td>
<td>—</td>
<td>III (2-4)</td>
<td>DM, I, LICS (2-4)</td>
<td>+++</td>
<td>88/60</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>3</td>
<td>F</td>
<td>—</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>II (4)</td>
<td>—</td>
<td>+</td>
<td>94/50</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>2</td>
<td>M</td>
<td>Since age 3 mo. when crying</td>
<td>+</td>
<td>+</td>
<td>++</td>
<td>Lower sternum</td>
<td>III (4)</td>
<td>DM, I, LICS</td>
<td>+ Split</td>
<td>95/45</td>
<td>+</td>
</tr>
</tbody>
</table>

*LICS, left intercostal space; DM, diastolic murmur; SM, systolic murmur.
†Roman numerals indicate grade of murmur; arabic numbers in parentheses indicate those left intercostal spaces where the murmur was heard.
There were four boys and four girls, with ages ranging from 2 to 15 years.

Slight cyanosis had been noted after effort in two patients. All but one had experienced dyspnea after effort. In three patients, congestive heart failure had been present in the past or was noted at the time of observation. Six patients had a history of recurrent respiratory infection.

Examination at the clinic did not show cyanosis or clubbing in any of the patients. A systolic thrill was present in all but one patient. A systolic murmur was present in all eight cases, varying from grade II to grade IV (on the basis of grades I to VI established by the New York Heart Association).

A grade I apical diastolic murmur was heard in three patients, and two had a Graham Steell murmur. An apical systolic murmur suggesting mitral insufficiency was noted in two patients (cases 3 and 4); this lesion subsequently was found at necropsy. The second pulmonic sound was accentuated in all eight patients. Femoral arterial pulsations were palpable in all instances, being somewhat diminished in one patient (case 4) because of an associated coarctation of the aorta. The blood pressure was in the normal range in all patients.

Electrocardiographic Findings

All eight patients had normal sinus rhythm (fig. 8). The P-R interval was greatly prolonged in case 3 and slightly prolonged in all the other cases, as based on the assumed P-R interval for age and heart rate (Ziegler).10

The P waves were high and notched in leads I, II, and aVf, suggesting left atrial enlargement, in the two patients (cases 3 and 4) in whom mitral insufficiency was found at necropsy.

The most striking electrocardiographic features in these eight cases were the manifest mean electric axes and the vectors of the QRS complexes in the frontal plane as projected from the scalar electrocardiograms (table 2). The manifest mean electric axes of the QRS complexes in seven of the patients

Figure 8

Electrocardiograms in two cases. a. Case 1. b. Case 7. See table 2 for details.
Table 2
Electrocardiographic Findings in Eight Cases of Origin of Both Great Vessels from Right Ventricle

<table>
<thead>
<tr>
<th>Case</th>
<th>Age, yrs.</th>
<th>Rate, beats per min.</th>
<th>QRS axis, degrees</th>
<th>QRS vector loop, frontal plane*</th>
<th>P waves, mm.</th>
<th>P–R interval, sec.</th>
<th>QRS complex, sec.</th>
<th>QRS in lead V₆, mm.</th>
<th>R in aV₆, mm.</th>
<th>R:S ratio in aV₆</th>
<th>Q in lead V₆, mm.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>14</td>
<td>90</td>
<td>-160</td>
<td>CCL</td>
<td>High, notched; 3 mm. in I; 4 mm. in II</td>
<td>0.20</td>
<td>0.14</td>
<td>R' = 22</td>
<td>10</td>
<td>&gt;1</td>
<td>Q = 2</td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>120</td>
<td>-70</td>
<td>CCL</td>
<td>High; 3 mm. in II</td>
<td>0.16</td>
<td>0.08</td>
<td>R = 25</td>
<td>9.5</td>
<td>&gt;1</td>
<td>Q = 2</td>
</tr>
<tr>
<td>3</td>
<td>15</td>
<td>83</td>
<td>-120</td>
<td>CCL</td>
<td>Notched in I, II and aV₆</td>
<td>0.24</td>
<td>0.12</td>
<td>R' = 23</td>
<td>12</td>
<td>&gt;1</td>
<td>Q = 2</td>
</tr>
<tr>
<td>4</td>
<td>6</td>
<td>112</td>
<td>-170</td>
<td>CCL; figure-of-eight</td>
<td>High and notched in I, II and aV₆</td>
<td>0.17</td>
<td>0.14</td>
<td>R = 25</td>
<td>8</td>
<td>&gt;1</td>
<td>Q = 10</td>
</tr>
<tr>
<td>5</td>
<td>2</td>
<td>142</td>
<td>-30</td>
<td>CCL</td>
<td>Notched in I and aV₆</td>
<td>0.16</td>
<td>0.07</td>
<td>RS</td>
<td>2.5</td>
<td>&lt;1</td>
<td>Q = 6</td>
</tr>
<tr>
<td>6</td>
<td>9</td>
<td>100</td>
<td>-130</td>
<td>CCL</td>
<td>2 mm. in II</td>
<td>0.16</td>
<td>0.10</td>
<td>RS</td>
<td>16</td>
<td>&gt;1</td>
<td>Q = 4</td>
</tr>
<tr>
<td>7</td>
<td>3</td>
<td>120</td>
<td>-90</td>
<td>CCL</td>
<td>Normal</td>
<td>0.16</td>
<td>0.08</td>
<td>RS</td>
<td>12</td>
<td>&gt;1</td>
<td>Q = 5</td>
</tr>
<tr>
<td>8</td>
<td>2</td>
<td>136</td>
<td>+120</td>
<td>CL</td>
<td>3 mm. in II</td>
<td>0.14</td>
<td>0.08</td>
<td>RS</td>
<td>9</td>
<td>=1</td>
<td>Q = 12</td>
</tr>
</tbody>
</table>

*CCL, counterclockwise; CL, clockwise.
Figure 9

a-h. Posteroanterior thoracic roentgenograms in cases 1 through 8, respectively. See text for details.
### Table 3

**Hemodynamic Data in Eight Cases of Origin of Both Great Vessels from Right Ventricle**

<table>
<thead>
<tr>
<th>Case</th>
<th>Pressure, mm. Hg†</th>
<th>Blood oxygen saturation, per cent‡</th>
<th>O₂ capacity, vol. per cent</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>RA</td>
<td>RV</td>
<td>PA</td>
</tr>
<tr>
<td>1</td>
<td>A</td>
<td>14/8</td>
<td>103/16</td>
</tr>
<tr>
<td>B</td>
<td>15/9</td>
<td>82/8</td>
<td>—</td>
</tr>
<tr>
<td>2 (B)</td>
<td>8/5</td>
<td>85/6</td>
<td>—</td>
</tr>
<tr>
<td>3 (A)</td>
<td>9/3</td>
<td>110/15</td>
<td>110/68</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4 (A)</td>
<td>20/14</td>
<td>124/10</td>
<td>129/60</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5 (A)</td>
<td>4</td>
<td>70/5</td>
<td>60/15</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>A</td>
<td>7</td>
<td>92/0</td>
</tr>
<tr>
<td>B</td>
<td>9/6</td>
<td>75/6</td>
<td>74/23</td>
</tr>
<tr>
<td>7</td>
<td>A</td>
<td>10/0</td>
<td>80/0</td>
</tr>
<tr>
<td>B</td>
<td>7</td>
<td>77/4</td>
<td>—</td>
</tr>
<tr>
<td>8 (B)</td>
<td>9/5</td>
<td>86/6</td>
<td>73/19</td>
</tr>
</tbody>
</table>

* A, catheterization performed elsewhere; B, data obtained during operation.
† RA, right atrium; RV, right ventricle; PA, pulmonary artery; LA, left atrium; LV, left ventricle; FA, femoral artery.
‡ IVC, inferior vena cava; MRA, mid right atrium; SVC, superior vena cava.
lay between -30 and -170 degrees; the frontal QRS vector described a counterclockwise loop in seven of these patients, and its major portion was superiorly oriented. Only one patient (case 8) had a different vector and axis. All patients showed a certain degree of right ventricular hypertrophy. The R waves in lead V1 were high in all instances. The R:S ratio in lead aVR was 1 or more in seven patients (fig. 8).

A QRS or QR pattern was present in lead V3 in all cases, with the Q waves ranging from 2 to 12 mm. (0.2 to 1.2 mv.), the R waves from 13 to 33 mm. (1.3 to 3.3 mv.), and the S waves from 4.5 to 22 mm. (0.45 to 2.2 mv.).

Roentgenologic Appearance
Roentgenologic examination revealed enlargement of the cardiac shadow and increased pulmonary vasculature as evidence of pulmonary arterial hypertension in all patients (fig. 9). In two instances (cases 1 and 8), the supracardiac shadow was widened.

Hemodynamics
The cardiac catheterization data are shown in table 3. In two of the patients (cases 3 and 4), complete data were obtained in our laboratory; case 3 will be discussed in greater detail. Most important is the fact that the pulmonary arterial pressure was equal to the systemic arterial pressure in all of the patients.

The peripheral systemic arterial oxygen saturation varied from slightly below normal (88 per cent) to normal (96.7 per cent) in four of the five patients in whom it was obtained, while the fifth patient (case 4) showed severe arterial hypoxemia. This patient had congestive heart failure and was studied while he was under anesthesia. Values for the oxygen saturation of pulmonary arterial blood were obtained in the same five patients; in two (cases 3 and 6), the values in the pulmonary artery and the aorta were closely similar.

Sufficient data were obtained in four patients to exclude the presence of a large left-to-right shunt at the atrial level.

Circulation, Volume XXIII, March 1961
Table 4  
Effect of Temporary Closure of Patent Ductus Arteriosus on Central Arterial Pressures (Case 3)  

<table>
<thead>
<tr>
<th>Aortic arch</th>
<th>Descending aorta</th>
<th>Pulmonary artery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ductus open</td>
<td>137/57</td>
<td>124/58</td>
</tr>
<tr>
<td>Ductus closed</td>
<td>143/74</td>
<td>111/75</td>
</tr>
</tbody>
</table>

*Pressures in pulmonary artery not recorded simultaneously with those in aorta.

Temporary closure of the ductus during operation resulted in a decrease in systolic pressure in the descending aorta and an increase in systolic pressure in the pulmonary artery, indicating the presence of a significant flow of blood via the ductus into the descending aorta (table 4).

This patient had practically uniform mixing of systemic venous and pulmonary venous blood in the outflow tract of the right ventricle and in the pulmonary artery, since the oxygen saturation of blood in the pulmonary, right radial, and femoral arteries was closely similar.

Discussion

The clinical picture in these patients was that of a large ventricular septal defect with pulmonary hypertension. Electrocardiography showed features that could be of some diagnostic help. To our knowledge, the electrocardiographic changes in this anomaly have not been previously described in detail. Witham included an electrocardiographic tracing in one case of his "Eisenmenger group," but it is difficult from the reproductions to judge the exact portion of the electric axis and the vectorial QRS loop in the frontal plane; the axis was described as left and the P-R interval as being prolonged.

As already noted, all of our patients had prolonged P-R intervals, and the manifest mean electric QRS axis ranged between -30 and -170 degrees in all but one instance. The QRS loop as projected on the frontal plane rotated in a counterclockwise direction in six patients, and its major portion was superiorly oriented. The universal presence of right ventricular hypertrophy was indicated by the magnitude of the R waves in leads aVR and V1 and of the S waves in V6. In cases 1, 3, and 4, a typical pattern of right bundle-branch block was observed.

The combination of the QRS electric axis lying above the superior point and a counterclockwise rotation of the QRS vector in the presence of signs of right ventricular hypertrophy has been described as an important electrocardiographic sign in persistent common atrioventricular canal by Toscano-Barboza and associates. Toscano-Barboza and DuShane found such electrocardiographic findings in nine of 59 anatomically proved cases of ventricular septal defect. According to Keith and co-workers, and Char and associates, the combination of left axis deviation in the standard leads and right ventricular hypertrophy in the precordial leads in cases with ventricular septal defect is rare.

Toscano-Barboza and associates, in their explanation of the electrocardiographic changes in common atrioventricular canal, suggested that the defect at the top of the ventricular septum causes a fundamental alteration in the excitation pathway into the ventricles.

Neufeld and co-workers found similar electrocardiographic alterations in excitation with ventricular septal defects of the persistent common atrioventricular canal type. Histologic studies showed that the conduction tissue lay dorsal to and skirted the ventricular septal defect. Similar changes were found by Lev in cases of common atrioventricular canal.

Since the defect in cases in which both great vessels originate from the right ventricle is positioned dorsally, the same changes in the position of the conduction system might be expected. At present, these changes in conduction are thought to be on the same embryologic basis as the defect and are a possible cause of the electrocardiographic alterations.

In the presence of clinical signs of ventricular septal defect with pulmonary hypertension and the electrocardiographic changes already described, the possibility of both great vessels originating from the right ventricle should be considered, as well as the possibility of a ventricular septal defect of
the persistent common atrioventricular canal type.

Hemodynamics

Measurements of pressure obtained during catheterization or operation or both disclosed closely similar systolic pressures in the right and left ventricles. In case 3, the oxygen saturation in the pulmonary artery was almost equal to that in the radial artery, which may be attributed to nearly complete mixing of systemic venous and left ventricular blood in the right ventricle. The pulmonary blood flow depends on the relative levels of pulmonary and systemic vascular resistance. With a lower pulmonary resistance, the pulmonary flow will be greater than the systemic flow, as in case 3.

In the presence of both an extremely high pulmonary flow and good mixing in the right ventricle, the oxygen saturation of pulmonary arterial blood can be equal to that in the systemic arterial system, and the systemic arterial blood oxygen saturation can approach normal values. If, during cardiac catheterization, a left-to-right shunt at the ventricular level is found and the oxygen saturation in the pulmonary artery is equal to that in one of the systemic arteries, the diagnosis of a single ventricle or the origin of both great vessels from the right ventricle is suggested. Determination of the anatomic positions of the semilunar valves and the great vessels may help to differentiate these two conditions.

In a single ventricle without pulmonary stenosis, the relative positions of the aorta and pulmonary artery frequently are reversed. The ascending aorta lies ventral and parallel to the pulmonary artery, with the aortic valve occupying a position that is ventral and cephalad to and to the left of the pulmonary valve.

When both great vessels take origin from the right ventricle, the ascending aorta lies to the right of the pulmonary artery and the two semilunar valves lie in approximately the same coronal and cross-sectional body planes.

The relationship of the great vessels to each other may be demonstrated by position-
The left ventricle. The aortic and pulmonary valves were in approximately the same cross-sectional and coronal body planes, which contrasts to the situation in the usual ventricular septal defect, in which the aortic valve lies in a normal position, being caudal to the pulmonary valve.

The clinical features simulated those of a large ventricular septal defect associated with pulmonary hypertension. The importance of distinguishing these two anomalies is emphasized, since the surgical methods for repair are different.

The following findings aid in the differential diagnosis: (1) electrocardiography shows a manifest mean electric axis above the zero line, with the frontal-plane vectorial loop of the QRS directed in a counterclockwise direction and its main portion lying above the zero line; (2) cardiac catheterization or angiocardiography or both demonstrate that the aortic valve lies at the same cross-sectional body level as the pulmonary valve in the anteroposterior view and at the same coronal plane in the lateral view; (3) in some cases, the oxygen saturation of blood in the pulmonary artery approaches or equals that in the aorta, indicating that relatively complete mixing of pulmonary venous and systemic venous blood has occurred in the outflow tract of the right ventricle.

References

3. Peacock, T. B.: Quoted by Witham, A. C.
Origin of Both Great Vessels from the Right Ventricle: I. Without Pulmonary Stenosis

HENRY N. NEUFELD, JAMES W. DUSHANE, EARL H. WOOD, JOHN W. KIRKLIN and JESSE E. EDWARDS

Circulation. 1961;23:399-412
doi: 10.1161/01.CIR.23.3.399

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1961 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/23/3/399.citation

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/