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, and only a few have been reported in the last 25 years. A few cases were documented in the last century.

Many terms have been used for this malformation, including “partial transposition,” “double-outlet right ventricle,” and “origin of both great vessels from the right ventricle.”

Witham, 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. 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.

Pathologic-anatomic 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

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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).
RIGHT VENTRICULAR ORIGIN OF GREAT VESSELS

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.). Externally, 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

Figure 2
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.). Externally, 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.).

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Figure 3
Case 3. Relationships of ventricular septal defect and mitral valve. a. Interior of left ventricle. The only outlet for the left ventricle is the ventricular septal defect (D.). The anterior leaflet of the mitral valve (A.M.) forms part of the dorsal wall of the ventricular septal defect. b. Section through heart parallel to that shown in figure 2d but on a more dorsal plane that lies dorsal to the semilunar valves. The anterior leaflet of the mitral valve has continuity with the septal leaflet of the tricuspid valve (S.T.) behind the ventricular septal defect. The anterior leaflet of the mitral valve attaches anteriorly with the muscular edge of the ventricular septal defect and does not have continuity with the aortic valve. This is in striking contrast to the normal situation. L.A. = left atrium.
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 the
RIGHT VENTRICULAR ORIGIN OF GREAT VESSELS

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 posterosinferior 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.

Figure 6

Figure 7

Case 6. Low-power photomicrograph showing continuity between aortic valve and anterior leaflet of mitral valve. The relationship between the two valves is profoundly different from that in the first sub-group (fig. 5). Although the relationship in this case bears some similarity to the normal, the zone of continuity between the two valves has a close epicardial relationship. Under normal conditions, this zone is entirely in an intracardiac position. The recess between the aortic wall and the left atrial wall is the pericardial cavity. Contrast this with the normal in figure 5b.

mitral valve not because of a normal position but because an elongated anterior mitral leaflet extends upward and to the right to join the aortic valve. The aortic valve is not caudal to the pulmonary valve but lies at the same level. The ventricular septal defect does not extend caudally dorsal to the septal tricuspid leaflet but rather extends cephalad ventral to the anterior tricuspid leaflet.

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>History</th>
<th>Physical findings*</th>
<th>Blood pressure, mm. of Hg</th>
<th>Femoral pulses</th>
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<td></td>
<td></td>
<td></td>
<td>Systolic murmur, grade and location†</td>
<td>Systolic thrill, location</td>
<td>Other murmurs; type, grade and location</td>
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<td>14</td>
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<td>LICS (2-4)</td>
<td>III (3-4)</td>
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<td>III (3-4)</td>
<td>Mid diastolic</td>
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<td>15</td>
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<td>II (2-4)</td>
<td>++</td>
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<tr>
<td>4</td>
<td>6</td>
<td>M</td>
<td>When crying</td>
<td>Suprasternal notch</td>
<td>III (3-4)</td>
<td>SM, I, apex</td>
</tr>
<tr>
<td>5</td>
<td>2</td>
<td>M</td>
<td>—</td>
<td>LICS (3-4)</td>
<td>IV (3-4)</td>
<td>DM, I, apex</td>
</tr>
<tr>
<td>6</td>
<td>9</td>
<td>M</td>
<td>—</td>
<td>—</td>
<td>III (2-4)</td>
<td>DM, I, LICS (2-4)</td>
</tr>
<tr>
<td>7</td>
<td>3</td>
<td>F</td>
<td>—</td>
<td>—</td>
<td>II (4)</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>
</tr>
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</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 Stell 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

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*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 V₄, mm.</th>
<th>R in aVₑ, mm.</th>
<th>R:S ratio in aVₑ</th>
<th>QRS in lead V₄, mm.</th>
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<td>14</td>
<td>90</td>
<td>-160</td>
<td>CCL</td>
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<td>0.20</td>
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<td>r R'</td>
<td>R' = 22</td>
<td>10</td>
<td>&gt;1</td>
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<tr>
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<td>CCL</td>
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<td>0.16</td>
<td>0.08</td>
<td>R = 25</td>
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<td>112</td>
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<td>CCL; figure-of-eight</td>
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*CCL, counterclockwise; CL, clockwise.
Figure 9

a-h. Posteroanterior thoracic roentgenograms in cases 1 through 8, respectively. See text for details.
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<th>Case*</th>
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<th>PA</th>
<th>Radial artery</th>
<th>Aorta</th>
<th>LA</th>
<th>IVC</th>
<th>MRA</th>
<th>SVC</th>
<th>RV</th>
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* 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.

Figure 10

Case 4. a. Position of catheter in posteroanterior view. VC = venous catheter; AC = aortic catheter; A = aortic valve; V = pulmonary valve. Note that both valves are at approximately the same level. b. Lateral view. Note that the distal part of the aortic catheter is positioned anterior to the venous catheter.

Data in Case 3

The data obtained during cardiac catheterization in case 3 established the presence of an interventricular communication and an aortico pulmonary communication, the latter being interpreted as a patent ductus arteriosus. The catheter traversed the ductus on numerous occasions and was advanced into the abdominal aorta.

The oxygen saturation of right radial arterial blood (91 per cent) was 2 per cent greater than that of blood withdrawn simultaneously from the femoral artery. The oxygen saturation of femoral and pulmonary arterial blood apparently was identical. Dye-dilution curves recorded simultaneously at the right radial and femoral arteries after injection into the superior vena cava were similar, each showing a right-to-left shunt that was approximately 35 per cent at the radial artery and slightly greater at the femoral artery.

The pulmonary blood flow of 15.0 liters per minute was considerably greater than the systemic blood flow of 5.8 liters. The estimated pulmonary resistance of 380 dynes sec. cm⁻² was significantly less than the total systemic resistance of 1035 dynes sec. cm⁻².

The data were interpreted to indicate a large left-to-right shunt at the ventricular level. The identity of the oxygen saturation levels in the pulmonary and femoral arteries is compatible with the interpretation that partial or complete interruption of the aortic arch was present, with all, or a large portion, of the descending thoracic aorta originating from the ductus arteriosus. Partial interruption of the aortic arch was found at operation. The systolic pressure in the aortic arch was significantly higher than that in the descending aorta (table 4).
Table 4

<table>
<thead>
<tr>
<th>Pressure, mm. Hg</th>
<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>
<td>128/66*</td>
</tr>
<tr>
<td>Ductus closed</td>
<td>143/74</td>
<td>111/75</td>
<td>149/58*</td>
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*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 aV_{6} and V_{1} and of the S waves in V_{6}. 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 Dushe 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 17, 18 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, 17 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-

ing two catheters simultaneously in the aorta and pulmonary artery, which demonstrates that the aorta is to the right of the pulmonary artery, with both lying parallel in postero-anterior as well as lateral views. This procedure also shows that the cephalocaudal levels of the aortic and pulmonic valves are similar (fig. 10a). In the lateral view, the catheters appear to be approximately in the same coronal body plane (fig. 10b).

Some cases of origin of both great vessels from the right ventricle (as well as some cases of single ventricle) 17 show poor mixing of venous and arterial blood in the ventricle. In these instances, a streaming effect apparently exists, 14 the left ventricular blood being shunted preferentially through the ventricular septal defect directly into the aorta and the right ventricular blood being directed into the pulmonary artery. When this occurs, the oxygen saturation of the pulmonary arterial blood is significantly less than that of the systemic arterial blood. Such patients present a difficult diagnostic problem at cardiac catheterization, because the data on blood oxygen saturation obtained from the cardiac chambers and great vessels resemble those in patients who have the usual type of ventricular septal defect with a large left-to-right shunt.

A clue to the correct diagnosis of common origin of the great vessels from the right ventricle can be obtained at cardiac catheterization in such patients by recognizing the unusual positions of the aorta and pulmonary arteries in relationship to each other, as well as the anatomic relationships of the semilunar valves. Demonstration of these unusual anatomic relationships can be particularly clear if the two-catheter technic (fig. 10) or biplanar selective angiocardioography is used.

Summary and Conclusions

Clinical, hemodynamic, and pathologic-anatomic findings were studied in eight cases in which both great vessels took origin from the right ventricle in the absence of pulmonary stenosis. In each case, a ventricular septal defect constituted the only outlet for
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

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