Origin of Both Great Vessels from the Right Ventricle
II. With Pulmonary Stenosis

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The first paper of this series dealt with the origin of both great vessels from the right ventricle in the absence of pulmonary stenosis. It was indicated that the clinical picture in that condition might be confused with that of a large ventricular septal defect. Electrocardiographically, such an anomaly might be confused with either a persistent common atrioventricular canal or the “AV commune” type of ventricular septal defect.

The condition forming the basis for the present communication anatomically has in common with the foregoing anomaly the origin of both great vessels from the right ventricle, but pulmonary stenosis is also present. The hemodynamic, clinical, and electrocardiographic data resemble those in the tetralogy of Fallot.

This part of the study includes five cases in which both great vessels originated from the right ventricle associated with ventricular septal defect and pulmonic stenosis, and a sixth case, in which both great vessels took origin from the right ventricle and in which a common atrioventricular canal and pulmonic stenosis were associated.

The purpose of this paper is to summarize the clinical, hemodynamic, and anatomic findings in these cases. The differentiation of this anomaly from the tetralogy of Fallot with cyanosis is important because of the more complicated surgical approach in the former.

This anomaly has been described before in articles devoted to pathology, but only in recent years have a few cases been reported clinically.

Witham,10 in 1957, summarized reports of three cases from the literature and added two of his own, calling them “double outlet right ventricle, Fallot type.”

Pathologico-anatomic Features

As in the cases in which both great vessels took origin from the right ventricle without pulmonary stenosis, the external view of the relationship of the great vessels in cases with pulmonary stenosis is very similar to the normal appearance, but important differences exist at lower levels within the heart (figs. 1-4).

The origin of the aorta lies to the right of the origin of the pulmonary artery, both
Table 1

Clinical Data in Five Cases of Origin of Both Great Vessels from Right Ventricle, with Pulmonary Stenosis

<table>
<thead>
<tr>
<th>Case</th>
<th>Age, yrs.</th>
<th>Sex</th>
<th>Cyanosis, age at onset</th>
<th>Shortness of breath</th>
<th>Palpitation</th>
<th>Cyanosis</th>
<th>Clubbing</th>
<th>Systolic thrill</th>
<th>Systolic murmur, grade and location*</th>
<th>Other murmurs</th>
<th>Second pulmonary sound</th>
<th>Blood pressure, mm. Hg</th>
<th>Femoral Gm./100 mL</th>
<th>Hemoglobin</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>7/12</td>
<td>M</td>
<td>2 mos.</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>(3)</td>
<td>-</td>
<td>-</td>
<td>115/75</td>
<td>+</td>
<td>19</td>
</tr>
<tr>
<td>2†</td>
<td>3</td>
<td>F</td>
<td>6 mos.</td>
<td>-</td>
<td>-</td>
<td>++</td>
<td>++</td>
<td>-</td>
<td>I</td>
<td>-</td>
<td>-</td>
<td>Single, not diminished</td>
<td>110/75</td>
<td>+</td>
</tr>
<tr>
<td>3†</td>
<td>13</td>
<td>M</td>
<td>2 1/2 mos.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>IV (4-5)</td>
<td>-</td>
<td>-</td>
<td>Single, not diminished</td>
<td>110/75</td>
<td>+ 15.9</td>
</tr>
<tr>
<td>4†</td>
<td>8</td>
<td>M</td>
<td>8 mos.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>III (3-4)</td>
<td>Machinery murmur; syst. ejection click</td>
<td>Diminished</td>
<td>100/80</td>
<td>+ 15.8</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>3 1/2</td>
<td>M</td>
<td>Birth</td>
<td>+</td>
<td>+</td>
<td>+++</td>
<td>+++</td>
<td>+</td>
<td>IV (3-4)</td>
<td>Diminished</td>
<td>-</td>
<td>100/75</td>
<td>+</td>
<td>+ 18.2</td>
</tr>
</tbody>
</table>

*Roman numerals indicate grade of murmur; Arabic numbers in parentheses indicate those left intercostal spaces where the murmur was heard.
†Previous Blalock’s procedure; a Potts operation also was done in case 4.
‡Previous Brock’s procedure.

Origin of both great vessels seen from right ventricle.

Figure 3

Origin of both great vessels seen from right ventricle. The horizontal limb of the septal defect (D) lies anterior to the horizontal limb of the ventricular septal defect. The aortic valve lies above the horizontal limb of the ventricular septal defect. The pulmonary valve lies below the horizontal limb of the ventricular septal defect.
Origin of both great vessels from right ventricle, with infundibular stenosis. The specimen in this case resembles the one shown in figure 1 a. Right ventricular aspect. A section has been made through the infundibular zone. The ventricular septal defect lies caudal to the crista supraventricularis (C. S.). A portion of the stenotic infundibular channel is present and lies below the pulmonary valve (P. V.). The aortic valve (A. V.) lies at the same cross-sectional body plane and to the right of the pulmonary valve, being separated from the atrioventricular valvar tissue by the horizontal limb of the crista ventricularis. b. Left atrium (L. A.) and left ventricle (L. V.). The anterior leaflet of the mitral valve has been stretched to prepare this illustration; it shows a normal caudal connection with the anterolateral papillary muscle (A.) and the posteromedial papillary muscle (P.). The ventricular septal defect (D.) lies ventral to the anterior leaflet of the mitral valve. c. Sagittal section through both ventricles and anterior leaflet of mitral valve. The posterior boundary of the ventricular septal defect (D.) is formed by the tissue of the septal leaflet of the tricuspid valve (T.) and the anterior leaflet of the mitral valve (M.), which joins the crista supraventricularis (C. S.). No continuity exists between the aortic valve and the atrioventricular valvar tissue. L. V. = left ventricle; P. T. = pulmonary trunk.
defect in the aorta be anterior leaflet of the mitral valve. They emphasize that, in the usual dissection, the fact that the aorta does not communicate with the left ventricle may be obscured by an illusion, which makes it appear to originate from the left ventricle when in fact it does not. a. Right ventricular aspect. The aortic valve is separated from the ventricular septal defect (D.) by a horizontal limb of the crista supraventricularis (C.S.). A vertical limb of the crista supraventricularis creates an infundibular chamber (I.), which is stenotic. In this specimen, the incision has been carried from the left ventricle to the aorta, the crista supraventricularis being divided. The postmortem incision in the aorta has been sutured. b. Left ventricle and aortic valve. The specimen has been opened in the line of section shown in a. This figure shows apparent continuity between the aorta and the left ventricle, but this is an illusion because between the aorta, the left ventricle and the anterior leaflet of the mitral valve (A.M.) is a mass of muscle (C.S.), the crista supraventricularis, which lies above the ventricular septal defect (D.) and below the aortic valve. In normal hearts, a section through the left ventricle into the aorta in this perspective shows continuity between the aortic valve and the anterior leaflet of the mitral valve. In this heart, the crista supraventricularis separates these two valvular structures.

Table 2

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure, mm Hg</th>
<th>Oxygen saturation, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inferior vena cava</td>
<td>—</td>
<td>53</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>8/5</td>
<td>57</td>
</tr>
<tr>
<td>Right atrium</td>
<td>8/5</td>
<td>54-56</td>
</tr>
<tr>
<td>Right ventricular outflow</td>
<td>115/7</td>
<td>55</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>15/7</td>
<td>56</td>
</tr>
<tr>
<td>Right femoral artery</td>
<td>125/71</td>
<td>61</td>
</tr>
</tbody>
</table>

Clinical Findings

The clinical data in the first five cases are summarized in table 1. There were four boys and one girl; the youngest patient was 7 months of age and the oldest was 13 years. The first patient in this study was seen at the Mayo Clinic in 1936 (case 1 in table 1). All the patients had been cyanotic since the first months of life. Clubbing of the fingers was present in three instances. Three patients had a systolic thrill. The second sound at the pulmonary area was accentuated in one patient (case 4), on whom both a Blalock and a Potts operation had been done in the past. Squatting was not observed in any of the patients. None of the patients had experienced congestive cardiac failure or undue respiratory infections.

Electrocardiographic tracings were obtained in four of these five cases, and they were compatible with right ventricular hypertrophy of the type seen in the tetralogy of Fallot (fig. 6).

Roentgenograms were obtained in the same four cases (fig. 7). They revealed moderate enlargement of the heart and diminished pulmonary vasculature.

Hemodynamics*

Catheterization of the right side of the heart was performed in one instance (case 2). The data are shown in table 2. Severe desaturation of the peripheral arterial blood

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*The authors gratefully acknowledge the assistance of Dr. E. H. Wood, who made available the hemodynamic data. Support for the latter was derived from grant no. H3532 of the National Heart Institute.
Figure 5

Case 5. A variant of the tetralogy of Fallot that in certain respects presents problems and anatomic relations more similar to those in origin of both great vessels from the right ventricle than to those in the classic tetralogy. The specimen shows the aortic origin (A.V.) from the right ventricle and to the right of the origin of the pulmonary trunk (P.T.). The left aspect of the aorta is at about the same plane as the ventricular septal defect (D.). The dorval wall of the ventricular septal defect is formed by the septal leaflet of the tricuspid valve and the anterior mitral leaflet (V.). This specimen shows continuity between the mitral valvular tissue and the aortic valve, but the position of the aortic valve is considerably farther to the right than it is in the classic tetralogy of Fallot.

was present. The oxygen saturation of right ventricular blood taken at the same time was 6 percentage points less than that of systemic arterial blood. The saturation of mixed venous blood ranged between 53 and 57 per cent. Pressures in the great veins and right atrium were normal. The right ventricular pressure was 115/7, expressed as millimeters of mercury. The catheter was manipulated to enter the pulmonary artery

Figure 6

Electrocardiograms in cases 2 through 5.

and was passed into the right lung field to the wedge position. The pressure in the right pulmonary artery was 10-15/0-5, and the wedge pressure was 2-4 mm. No significant difference was noted between the oxygen saturation in the main pulmonary artery and that in the outflow portion of the right ventricle. When the catheter was withdrawn across the pulmonary valve, the region of obstruction appeared to be sharply localized to a region at or near the pulmonary valve, since a high systolic ventricular pressure was recorded immediately upstream to the pulmonary valve. Further changes in pressure were not
Table 3
Cardiac Catheterization Data in Case 6

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure, mm Hg</th>
<th>Oxygen saturation, per cent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>—</td>
<td>55</td>
</tr>
<tr>
<td>Right atrium</td>
<td>8/3</td>
<td>57</td>
</tr>
<tr>
<td>Left atrium</td>
<td>11/7</td>
<td>79-85</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>116/4</td>
<td>65-67</td>
</tr>
<tr>
<td>Femoral artery</td>
<td>121/63</td>
<td>72</td>
</tr>
<tr>
<td>Pulmonary vein</td>
<td>10/5</td>
<td>99</td>
</tr>
</tbody>
</table>

observed when the catheter was withdrawn from this position to the lower part of the right ventricle.

The catheter was then advanced, and it entered the aorta. The aortic arch was located on the left.

Indicator-dilution curves were recorded at the right radial artery after injections of cardio-green dye into the main pulmonary artery, the lower part of the right ventricle, the aorta and the superior vena cava. The contours of the curves obtained after injection into the superior vena cava, right ventricle, and aorta were similar or almost identical. The similarity of the contours of curves recorded after injections into the superior vena cava and right ventricle excluded the possibility of preferential injection into the aorta from the right ventricle. The close similarity of the curves indicated an extremely large right-to-left shunt and practically an absence of flow from the right ventricle directly to the pulmonary artery.10

Figure 7
a-d. Thoracic roentgenograms in cases 2 through 5.
RIGHT VENTRICULAR ORIGIN OF GREAT VESSELS

The hemodynamic state in this anomaly is similar to that present in severe tetralogy of Fallot with cyanosis. The pressures in both ventricles are equal, and a pressure gradient exists between the right ventricle and the pulmonary artery. Peripheral oxygen desaturation is present in all cases.

In some instances, relatively complete mixing of systemic venous and left ventricular blood in the right ventricle might be expected to occur; when this is present, closely similar values for oxygen saturation in the systemic and pulmonary arteries would be found. On the other hand, some of the patients will show poor mixing in the right ventricle. In such instances, a streaming effect apparently exists, 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. In these cases, the oxygen saturation of the pulmonary arterial blood will be significantly less than that of the systemic arterial blood. This situation prevailed in case 2 (table 2).

Diagnostic Clues
A clue to the correct diagnosis of the origin of both great vessels from the right ventricle with pulmonary stenosis can be obtained during cardiac catheterization or angiography or both. Utilizing a two-catheter technic (venous and aortic) and bearing in mind the anatomic relationship of the semilunar valves, one might prove that both semilunar valves are in the same cross-sectional and coronal body planes. The same applies to biplanar selective angiography. Another clue that can be obtained in angiography is the relationship of the ventricular septal defect to the crista supraventricularis. In the tetralogy of Fallot, the crista supraventricularis is ventral to the ventricular septal defect; in origin of both great vessels from the right ventricle with pulmonary stenosis, the crista supraventricularis is cephalad to the ventricular septal defect.

Report of Unusual Case
The afore-mentioned sixth case showed additional interesting pathologic features.

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Figure 8
Case 6. a. Electrocardiogram of a 3-year-old girl who had persistent common atrioventricular canal. b. Thoracic roentgenogram in same case.
Case 6. Origin of both great vessels from right ventricle, with persistent common atrioventricular canal. In this unusual heart, the relative positions of the aorta and the pulmonary trunk are contrary to those in the usual examples of origin of both great vessels from the right ventricle. In the latter condition, the aortic origin lies to the right of the pulmonary trunk; in this specimen, the aortic origin lies ventral to the pulmonary trunk. a. External view of heart and great vessels, showing prominent ventral position of aorta. b. Interior of right ventricle. The aorta (A.V.) arises from the right ventricle above the crista supraventricularis (C.S.), which creates an infundib-
RIGHT VENTRICULAR ORIGIN OF GREAT VESSELS

Case 6. A 3-year-old girl was brought to the clinic in June 1958, for evaluation of a cardiac lesion and the possibilities of surgical intervention. The child had been cyanotic since birth, and her physical and mental development was impaired.

Cyanosis and clubbing were present. The heart was quiet, and a harsh systolic murmur (grade 2) was best heard at the second and third left intercostal spaces. The second cardiac sound in the pulmonary area was single and diminished. The femoral pulsations were normal.

The electrocardiogram showed a normal sinus rhythm. The mean manifest QRS electric axis was —170 degrees, with a figure-of-eight loop in the frontal plane, the initial portion of the loop being counterclockwise. Signs of right ventricular hypertrophy were present in the thoracic leads (fig. 8a).

Roentgenologic examination revealed cardiac enlargement and decreased pulmonary vasculature (fig. 8b).

Cardiac catheterization was performed (table 3). The pulmonary artery was not entered. The left atrium was entered in a retrograde fashion from the ventricle, suggesting either an anatomically common ventricle or a persistent common atrioventricular canal type of defect. The pulmonary venous saturation was greater than the left atrial saturation, indicating a right-to-left shunt at the atrial level. The most likely diagnosis considered from the catheterization data was a common ventricle or a persistent common atrioventricular canal associated with pulmonary stenosis.

An infundibular resection was performed at operation. The patient died on the same day.

Pathologic Findings. Necropsy revealed that the great vessels were transposed; both vessels originated from the right ventricle, but the pulmonary trunk lay dorsal to and to the right of the ascending aorta (fig. 9). The ascending aorta was considerably wider than normal, while the pulmonary trunk was thin-walled and narrow, measuring only about 8 mm. in diameter.

Significant venous anomalies were present. Bilateral superior vena cavae were present; the bridge between the two that is present under normal conditions also existed in this case. The right superior vena cava entered the right atrium in the normal position, while the left superior vena cava entered the left atrium at a position comparable to that at which the right one entered the right atrium. Two inferior vena cavae were present that perforated the diaphragm on either side of the esophagus. The right one measured about 1.5 cm. in diameter, while the left measured 1 cm. The right inferior vena cava entered the right atrium in a normal position, while the left inferior vena cava entered the left atrium in the posterosuperior aspect, inferior to the left inferior pulmonary vein. The pulmonary veins entered the left atrium normally.

The interior of the heart showed an atrial septal defect measuring about 1 by 0.5 cm. involving the fossa ovalis. No lower limbus of the fossa ovalis could be identified. The lowermost portion of the atrial septum contained a large defect characteristic of that associated with a persistent common atrioventricular canal. A common atrioventricular valve served both sides of the heart. The lowermost part of the defect in this region was formed by the crest of the underlying ventricular septum. Thus, the defect presented a picture of the complete variety of persistent common atrioventricular canal. Beneath the posterior leaflet were many interchordal spaces that allowed interventricular communication. A small number of interchordal spaces were found beneath the anterior leaflet of the common atrioventricular valve. The coronary sinus was absent. Two ventricles were present, each being about the same size, although the right ventricular muscle appeared somewhat thicker than the left. The left ventricle did not have any vessel of egress. This chamber communicated with the right side by way of the subvalvular communications in relation to the persistent common atrioventricular canal that already has been described; in addition, a defect about 8 mm. in diameter was present in the muscular part of the most superior anterior portion of the ventricular septum.
The aorta arose entirely from the right ventricle. Posterior to the origin of the aorta was a narrow muscle-walled tract measuring only about 3 mm in diameter that led to the pulmonary artery beyond. Evidence of recent surgical intervention in relationship to the subpulmonary stenosis was present. The aortic arch was on the left side. The ductus arteriosus was closed. The brachial arteries appeared to be enlarged.

The pathologic diagnoses included origin of both great vessels from the right ventricle, with pulmonary stenosis; persistent atrioventricular canal (complete variety); anomalous connection of a persistent left superior vena cava with the left atrium; bilateral inferior vena cavae, the left one communicating with the left atrium; absence of the coronary sinus.

Comment. Despite the pathologic-anatomic findings of significant pulmonary stenosis, the initial vectors in the electrocardiogram in the frontal plane were directed to the left, as they are in persistent common atrioventricular canal, which was present in this patient.

Summary and Conclusions
Clinical, hemodynamic, and pathologic-anatomic findings were studied in six cases in which both great vessels took origin from the right ventricle in the presence of pulmonary stenosis. This condition is indistinguishable from the tetralogy of Fallot, with cyanosis on the basis of clinical, electrocardiographic, and radiologic findings. The hemodynamics are also similar in both conditions.

The more complicated surgical approach in the condition under consideration emphasizes the importance of its differentiation from the usual case of tetralogy of Fallot with cyanosis.

Only angiocardiography and cardiac catheterization might be of diagnostic help, if one bears in mind the anatomic characteristics of this condition. The fact that both the aortic and pulmonary valves are at the same horizontal body level in this anomaly must be considered at angiocardiography. Also, the fact that the crista supraventricularis is cranial and more dorsal to the ventricular septal defect may be of some diagnostic help at angiocardiography. Careful analysis of the exact position of the semilunar valves during combined catheterization of the right side of the heart and the aorta is of some diagnostic importance.

References

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