The Two-Chambered Right Ventricle

Report of Nine Cases

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With the technical assistance of Charles E. Crawford, Jr.

The Division of the right ventricle into two chambers by aberrant, hypertrophied muscular bands is a relatively rare congenital cardiac malformation. No reports have been found that describe the details of anatomic structure and hemodynamics in this anomaly, although Keith, Rowe, and Vlad and Kjellberg et al. alluded to this malformation in their texts. Grant, Downey, and MacMahon described the architecture of the complex intrinsic bulbar musculature of the right ventricle, and stated "If the oblique component (cf. the bulbar musculature) is hypertrophied, the obstruction will lie much lower in the outflow tract, adjacent to the moderator band." The two-chambered right ventricle is usually seen in association with an interventricular septal defect, and the hypertrophied muscular bands produce an effective stenosis with an inflow high-pressure chamber immediately below the tricuspid valve and a low-pressure outflow chamber distal to the bands. This malformation should not be classified as tetralogy of Fallot, because the anatomic structure is quite different (fig. 1).

Nine patients have been found with this anomaly during the past 5 years (1956 to 1961) among 600 infants and children who had diagnostic cardiac studies at the St. Louis Children's Hospital. The purpose of this report is to show that (1) this anomaly may be mistakenly diagnosed as cyanotic tetralogy of Fallot or isolated interventricular septal defect, (2) the diagnosis can be made with the aid of selective angiocardiography, and (3) this cardiac malformation is amenable to surgical correction.

Methods and Materials

The nine patients studied were admitted to the St. Louis Children's Hospital and, in addition to the usual evaluation, were studied in the following manner.

Arterial oxygen saturation was measured by the ear-piece method with use of the ear oximeter. A standard 12-lead electrocardiogram and chest roentgenograms with and without barium swallow in posteroanterior, left lateral, right anterior oblique, and left anterior oblique positions were obtained. Right-sided cardiac catheterization was done under general anesthesia (oxygen, nitrous oxide, nitroprusside). The oxygen concentration in the gas mixture varied from 30 to 50 per cent. Side-hole catheters were used, and the heart was entered by way of a saphenous vein. The oxygen saturations were measured with the cuvette oximeter, and the electrocardiogram and intracardiac blood pressures were recorded with an Electronics for Medicine 5-channel recorder. Biplane angiocardiography, with a Schonander film changer (6 exposures per second of 24 by 30 cm. film), was performed immediately following the catheterization under general anesthesia. The contrast medium was 75 per cent sodium methylglucamine diatrizoate in a dosage of 1.0 to 1.5 ml. per Kg. of body weight. The injection was performed with a Gilford pressure syringe at 15 to 30 ml. per second, depending on the size of the patient.

Results

A summary of the clinical findings, electrocardiograms, and chest roentgenograms is presented in table 1. The catheterization data are summarized in table 2, and angiocardiographic descriptions on patients R.R. and K. S. are presented in short summaries and are accompanied by diagrams of the right ventricular cavity as well as selected angiocardiograms on each patient.

Angiocardiographic Summaries

Case 1 (figs. 2 and 3). Right ventricular hypertrophy and moderate dilatation were
Figure 1

Artist's depiction of the anatomy of a classic tetralogy of Fallot contrasted with the anatomy in a two-chambered right ventricle. In the two-chambered right ventricle the position of the ventricular septal defect in relation to the stenosis varies from case to case as indicated in the drawing (ventricular defects 1 and 2).

Figure 2

Case 1. Schematic drawings from angiocardiograms. To the left, anteroposterior view; to the right, lateral view. 1. Main pulmonary artery. 2. Low-pressure chamber. 3. Aberrant hypertrophiied muscle bands. 4. High-pressure chamber.

present. The supraventricular crista was well visualized but not prominent. The parietal band was displaced caudally and to the left and formed part of the severe low infundibular stenosis. The left part of the stenosis was formed by the septal band. The stenosis divided the right ventricle into two approximately equal-sized chambers. The diameter of the orifice of the stenosis was about 0.5 cm. Above the stenosis, the outflow tract had a normal appearance. The pulmonary valves were slightly thickened and deformed, but no stenosis could be demonstrated. The main pulmonary artery and peripheral arteries of the lungs were normal in appearance. The left atrium was slightly enlarged. The left ventricle emptied almost completely at the end of the ventricular systole. The aortic valves were somewhat thickened and opened normally during ventricular systole. The ascending aorta was wide. The diagnoses were made of hypertrophied muscular bands below the infundibulum, producing severe stenosis and a two-chambered right ventricle, and pulmonary and aortic valve deformities without significant stenosis. This patient was operated on successfully with the use of extracorporeal circulation. The anatomic structure of the right ventricle was found to be as described in the angiocardiographic report.

Case 3 (figs. 4, 5, and 6). The right ventricle was hypertrophic and showed coarse
TWO-CHAMBERED RIGHT VENTRICLE

Figure 3

Case 1. Angiocardiography in anteroposterior and lateral projections. Low right ventricular injection. Low stenosis is formed by hypertrophic bands. No ventricular septal defect is present (cf. fig. 2).

Discussion

Clinical Findings

The clinical findings in patients with a two-chambered right ventricle, with or without a ventricular septal defect, are not diagnostic and are quite similar to the findings in patients with isolated low pressure ventricular septal defects or acyanotic "tetralogy of Fallot."

The auscultatory findings were similar in all patients. A loud, harsh systolic murmur was heard best at the left sternal border, and a systolic thrill was palpable in this area. The pulmonary second sound was diminished in all patients.

trabeculations. The crista supraventricularis was not prominent, but the parietal and septal bands were hypertrophied and displaced so that a low stenosis was formed. The high-pressure chamber of the right ventricle was smaller than the low-pressure one. The main pulmonary artery was moderately widened, but there were no signs of poststenotic dilatation. The peripheral pulmonary arteries were somewhat wider than normal. The pulmonary veins were slightly widened and irregular. A ventricular septal defect was demonstrated by right-to-left flow of the contrast medium during the injection. The left atrium was enlarged. The left ventricle was slightly dilated. No appreciable increase in residual blood was present at the end of ventricular systole. During the opacification of the left ventricle, the right ventricle was re-opacified, indicating a left-to-right shunt at the ventricular level. The aorta was right-sided but otherwise unremarkable. The diagnoses were hypertrophied muscular bands below the infundibulum, which produced a stenosis and a two-chambered right ventricle, and a ventricular septal defect located approximately at the level of the stenosis. This patient was operated upon successfully with the use of extracorporeal circulation, and the anatomic structure of the right ventricle was the same as described above.
### Clinical Findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr.)</th>
<th>Height (cm.)</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Physical findings</th>
<th>Electrocardiogram</th>
<th>Chest roentgenograms</th>
<th>Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>12</td>
<td>143.5</td>
<td>M</td>
<td>Frequent upper and lower respiratory infections; easy fatigability; mild cyanosis on exertion</td>
<td>Acyanotic; systolic thrill at 2 to 5 LIs; grade 4/4 harsh systolic murmur over the precordium, loudest at the LSB; P; diminished</td>
<td>N</td>
<td>Moderate cardiomegaly; suggestion of 3rd ventricle; pulmonary vessels are normal</td>
<td>Yes</td>
</tr>
<tr>
<td>2</td>
<td>11</td>
<td>156</td>
<td>F</td>
<td>Easy fatigability and cyanosis</td>
<td>Slightly cyanotic; systolic thrill at 3 and 4 LIs; grade 4/4 harsh systolic murmur over the entire precordium, loudest at the LSB; P; diminished</td>
<td>RVE, IRBBB</td>
<td>RVE; 3rd ventricle observed; pulmonary vessels unremarkable; right-sided aortic arch, normal in width</td>
<td>Yes</td>
</tr>
<tr>
<td>3</td>
<td>2½</td>
<td>91.5</td>
<td>M</td>
<td>Frequent upper respiratory tract infections</td>
<td>Acyanotic; systolic thrill at 3 to 5 LIs; grade 4/4 harsh systolic murmur over the entire precordium, loudest at the LSB; P; diminished</td>
<td>N</td>
<td>Cardiomegaly; RVE, LVE, LAE; pulmonary vessels wide; arch and descending aorta right-sided</td>
<td>Yes</td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>89</td>
<td>M</td>
<td>Cyanosis with crying</td>
<td>Acyanotic; systolic thrill at 3 and 4 LIs; grade 3/4 harsh systolic murmur over the entire precordium, loudest at the LSB; P; diminished</td>
<td>N</td>
<td>Cardiomegaly; biventricular enlargement; LAE; main and peripheral pulmonary vessels wide</td>
<td>No</td>
</tr>
<tr>
<td>5</td>
<td>12</td>
<td>145</td>
<td>F</td>
<td>Frequent respiratory infections; easy fatigability</td>
<td>Acyanotic; systolic thrill at 2 to 5 LIs; grade 4/4 harsh systolic murmur over the entire precordium, loudest at the LSB; P; diminished</td>
<td>IRBBB, prolonged P-R interval</td>
<td>RVE; 3rd ventricle present; pulmonary vessels wide</td>
<td>No</td>
</tr>
</tbody>
</table>
### Table 1 (Continued)

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr.)</th>
<th>Height (Cm./Kg.)</th>
<th>Symptoms</th>
<th>Physical findings</th>
<th>Electrocardiogram</th>
<th>Chest roentgenograms</th>
<th>Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>6 V.B.</td>
<td>5</td>
<td>113</td>
<td>Frequent respiratory infections</td>
<td>Slightly cyanotic; systolic thrill at 3 and 4 LIS; grade 4/4 harsh systolic murmur over the entire precordium, loudest at the LSB; P₂ diminished</td>
<td>Suggestive of RVE</td>
<td>RVE and slight elevation of the apex of the heart; suggestion of 3rd ventricle; pulmonary vessels narrow</td>
<td>Yes</td>
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<tr>
<td>7 D.M.</td>
<td>11</td>
<td>132</td>
<td>Asymptomatic except for retarded physical growth and development and enuresis*</td>
<td>Acyanotic; systolic thrill at 3 and 4 LIS; grade 4/4 harsh systolic murmur over the entire precordium, loudest at the LSB; P₂ diminished</td>
<td>N</td>
<td>RVE; central pulmonary vessels wide; peripheral vessels narrow; dilatation of the ascending aorta</td>
<td>No</td>
</tr>
<tr>
<td>8 K.M.</td>
<td>3½</td>
<td>86</td>
<td>Asymptomatic</td>
<td>Acyanotic; systolic thrill at 3 and 4 LIS; grade 4/4 harsh systolic murmur over the entire precordium, loudest at the LSB; P₂ diminished</td>
<td>Suggestive of RVE</td>
<td>RVE, LVE, and LAE; Suggestion of 3rd ventricle; pulmonary vessels wide</td>
<td>Yes</td>
</tr>
<tr>
<td>9 S.S.</td>
<td>14</td>
<td>160</td>
<td>Asymptomatic</td>
<td>Acyanotic; systolic thrill at 3 and 4 LIS; grade 3/4 harsh systolic murmur over the entire precordium, loudest at the LSB; P₂ diminished; decescendo diastolic murmur at pulmonic area; P₂ diminished</td>
<td>N</td>
<td>Cardiomegaly; LVE, LAE; pulmonary vessels slightly wide; aorta slightly increased in width</td>
<td>No</td>
</tr>
</tbody>
</table>

*Study showed marked hydronephrosis and hydroureters bilaterally, enlarged bladder and bladder neck obstruction. Uretero-ileostomy was performed. LIS, left intercostal space; LSB, left sternal border; N, within normal limits; RVE, right ventricular enlargement; IRBBB, incomplete right bundle-branch block; LVE, left ventricular enlargement; LAE, left atrial enlargement.*
**Catheterization Data**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Oxygen saturation per cent, cuvette oximeter</th>
<th>Pressures, mm. Hg</th>
<th>Associated defects</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>O₂ Sat. per cent ear piece</td>
<td>High-pressure chamber</td>
<td>Low-pressure chamber</td>
</tr>
<tr>
<td>1</td>
<td>R.R. 92 60 62 57 57 58 57 .. ..</td>
<td>8 4</td>
<td>77/0 38/0</td>
</tr>
<tr>
<td>2</td>
<td>R.L. 98 80 74 75 74 80 72 74 .. 87</td>
<td>6 0</td>
<td>133/0 ..</td>
</tr>
<tr>
<td>3</td>
<td>K.S. 98 80 74 73 81 81 81 .. ..</td>
<td>8</td>
<td>72/0 49/0</td>
</tr>
<tr>
<td>4</td>
<td>C.E. 92 83 .. 82 96* 92 91 91 .. ..</td>
<td>14 4</td>
<td>72/0 47/0</td>
</tr>
<tr>
<td>5</td>
<td>C.B. 98 84 81 82 84 82 82 81 .. 81</td>
<td>2</td>
<td>97/0 28/0</td>
</tr>
<tr>
<td>6</td>
<td>V.B. .. 82 80 82 89 87 88 88 .. 88</td>
<td>11</td>
<td>72/0 27/0</td>
</tr>
<tr>
<td>7</td>
<td>D.M. 98 89 83 87 87 87 87 .. ..</td>
<td>7</td>
<td>58/0 24/0</td>
</tr>
<tr>
<td>8</td>
<td>K.M. 98 85 .. 81 84 89 90 88 .. ..</td>
<td>6</td>
<td>70/0 33/0</td>
</tr>
<tr>
<td>9</td>
<td>S.S. .. 88 85 86 91 .. 92 92 .. ..</td>
<td>2</td>
<td>48/0 25/0</td>
</tr>
</tbody>
</table>

*Probably close to the VSD.

IVC, inferior vena cava; SVC, superior vena cava; RA, right atrium; RV, right ventricle; MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery; VSD, ventricular septal defect; PDA, patent ductus arteriosus; +, yes; .., no.
Case 3. Schematic drawings from angiocardograms. To the left, anteroposterior view; to the right, lateral view. Arrows represent the location of the ventricular septal defect. 1. Main pulmonary artery. 2. Low-pressure chamber. 3. Aberrant hypertrophied muscle bands. 4. High-pressure chamber.

The electrocardiogram was within normal limits in five cases. The other four patients showed varying degrees of right ventricular enlargement or incomplete right bundle-branch block.

Seven of the nine children had normal physical growth and development, and two were subnormal. Two of the nine manifested intermittent cyanosis upon exertion (patients 2 and 4).

Chest Roentgenograms

In the classic tetralogy of Fallot with marked infundibular stenosis, the plain chest roentgenograms are fairly typical and the diagnosis can usually be suspected on the basis of the plain films. The appearance on the plain films of the two-chambered right ventricle with or without a ventricular septal defect, on the other hand, is rather kaleidoscopic. The appearance of the plain chest films is determined by the degree of stenosis and the pressure in the right ventricle. If a ventricular septal defect is present, the location and size govern the appearance of the pulmonary vasculature.

The right ventricle was enlarged and hypertrophic in most patients, and was seen as a continuity of the anterior heart border with the sternum in the lateral view. In one case there was a suggestion of a "coeur en sabot." with an upturn of the apex of the heart. A third ventricle was seen in five patients (1, 2, 5, 6, and 8) as a localized bulge of the left cardiac contour below the pulmonary artery. Kjellberg et al.2 observed a third ventricle in 42 of 63 cases of tetralogy of Fallot.

The appearance of the pulmonary vasculature varied from patient to patient. In case 1, which did not have a ventricular septal defect or patent ductus arteriosus, the pulmonary vasculature was normal. The appearance of the pulmonary vasculature in the other cases was determined by the degree of left-to-right shunt, except in case 6 (V.B.), which had narrow pulmonary vessels in spite of significant left-to-right shunts (ventricular septal defect and a patent ductus arteriosus.)

The left atrium showed enlargement in four cases (3, 4, 8, and 9). The left atrial enlargement implies a significant left-to-right shunt.

Figure 4

Case 3. Anteroposterior view. Low right ventricular injection. The band-like stenotic area is well visualized. Note coarse trabeculation in low-pressure chamber (cf. fig. 4).
Catheterization

The right-sided cardiac catheterization may strongly suggest, but not establish, the diagnosis of this anomaly when the pressures and oxygen saturations are measured in both chambers of the right ventricle and the anatomic position of the stenosis is observed by fluoroscopy. However, a number of pitfalls exist. The high-pressure chamber may be bypassed during the catheterization of the right ventricle, and pressures and oxygen saturations may be obtained only in the low-pressure chamber. An erroneous diagnosis may then be made of an interventricular septal defect with normal pressures. This happened during a previous catheterization at another hospital in case 2 (R.L.). If no rise in oxygen saturation is found, and the pressure in the high-pressure right ventricular chamber is not recorded, an erroneous diagnosis of a normal right-sided catheterization will be made.

The position of the interventricular septal defect in relation to the site of stenosis can sometimes be ascertained by right-sided cardiac catheterization. In case 8 (K.M.) the rise in oxygen saturation occurred in the low-pressure chamber of the right ventricle, indicating that the ventricular septal defect was located above the area of stenosis. In cases 3, 4, 6, and 9, the rise in oxygen saturation was noted in the high-pressure chamber of the right ventricle, and indicated that the interventricular septal defect opened into this chamber (confirmed at time of operation in case 3). In case 5, no significant rise in oxygen saturation was encountered and the absence or presence of an interventricular septal defect could not be determined until selective angiocardiography was performed.

Angiocardiography

Angiocardiography is the ideal diagnostic technic in cases of low infundibular stenosis of the right ventricle. The contrast medium should be injected into the high-pressure chamber of the right ventricle. Biplane angiocardiography is essential for the correct interpretation, since the stenosis sometimes is seen only in one projection. The anteroposterior and the lateral are the best projections. The stenosis is usually demonstrated as oblique bands in the lateral view, originating in the region of the supraventricular crista and proceeding caudally and ventrally. In the anteroposterior view, the stenosis is usually demonstrated as oblique, irregular bands separating the right ventricle into two chambers about equal in size. The stenosis is always best demonstrated in systole and may not be seen at all in diastole. Therefore,
it is of importance to have a relatively high film frequency so that every phase of the heart cycle is depicted. If the pressure is high in the right ventricle, it is possible to demonstrate the ventricular septal defect directly during the injection. Contrast medium will pass from the right ventricle to the left and the location of the defect can be observed. It is frequently impossible to show the direct relationship between the stenosis and the septal defect because the ventricular septal defect is demonstrated only in the lateral view. The septal defect, however, is usually located quite close to the stenosis, as observed in two of the operated cases (2 and 3). When the pressure in the right ventricle is relatively low, the septal defect cannot be demonstrated directly during the injection. During the opacification of the left ventricle, however, the right ventricle is re-opacified as evidence of the left-to-right shunt. Occasionally, a dilution effect can be seen in the right ventricle in the early phase of the examination as a result of the left-to-right shunt.

The pulmonary vasculature and the left side of the heart including the aorta are visualized with the aid of angiocardiography. Finally, angiocardiography will reveal the presence of additional defects, such as patent ductus arteriosus, in patients 6, 7, and 9, and deformities of the aortic valves, as shown in patients 1 and 7.

Summary

A relatively rare congenital cardiac malformation is described in nine patients and is characterized by aberrant hypertrophied muscular bands that divide the right ventricular cavity into two chambers. These hypertrophied muscular bands produce an effective stenosis and obstruct the outflow of blood from the right ventricle. An interventricular septal defect is usually seen in association with this abnormality. This cardiac malformation is anatomically distinct from the classic tetralogy of Fallot but may be mistakenly diagnosed as an acyanotic tetralogy of Fallot or an interventricular septal defect. The use of selective angiocardiography in conjunction with right-sided cardiac catheterization will establish the correct diagnosis. This congenital lesion is amenable to surgical correction, and five of the patients in this series were successfully operated upon with the aid of extracorporeal circulation. Surgery has been recommended for the remaining four patients because of the encouraging results.

References

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* Circulation. 1962;26:279-287
  doi: 10.1161/01.CIR.26.2.279

* Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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