Development of Right Ventricular Obstruction by Aberrant Muscular Bands

By Alexis F. Hartmann, Jr., M.D., David Goldring, M.D., and Erik Carlsson, M.D.

Attention was first called to the "two-chambered right ventricle" in recent publications. This congenital cardiac malformation is characterized by aberrant hypertrophied muscular bands that divide the right ventricular cavity into a high-pressure chamber located immediately distal to the annulus of the tricuspid valve and a low-pressure chamber distal to the hypertrophied muscle bands. The bands produce an effective obstruction to the flow of blood from the right atrium to the pulmonary artery. Thirteen of the 15 reported cases also had an interventricular septal defect. The malformation should not be confused with tetralogy of Fallot because the obstructive hypertrophic bands are usually proximal to the right ventricular infundibulum. The diagnosis may be suspected at the time of cardiac catheterization and firmly established with selective angiocardiography and this malformation is amenable to surgical correction.

The purpose of this report is to show that the aberrant muscular bands, although present, may be nonobstructive in early infancy and that the obstructive effect is developed with time as the bands become progressively more hypertrophied. This is based upon a study of three patients by catheterization and angiocardiography at intervals of 1½ to 2 years.

Methods and Materials

Three patients were admitted to the St. Louis Children's Hospital and were studied in the following manner.

A standard 12-lead electrocardiogram and chest roentgenograms with and without barium swallow in the posteroanterior, left lateral, right anterior oblique, and left anterior oblique positions were obtained. Right-sided cardiac catheterization was done with NIH side-hole catheters under general anesthesia (oxygen 30 to 50 per cent, nitrous oxide, Fluothane). Oxygen saturation was measured with the cuvette oximeter and the electrocardiogram and intracardiac blood pressures were recorded with an Electronics-for-Medicine five-channel recorder.

Biplane angiocardiography was performed under general anesthesia with a Schönder film changer (maximum exposure rate, 6 films per second) immediately following catheterization. The contrast medium was 75 per cent sodium and methylglucamine diatrizoates in a dose of 1.0 to 1.5 ml. per Kg. of body weight. The Gildlund pressure syringe was used and set at an injection rate of 15 to 25 ml. per second.

Report of Cases

Case 1

The patient, B.S., was a product of a full-term, normal pregnancy and weighed 6 lb., 4 ounces at birth. She was referred to the hospital for cardiac evaluation on October 31, 1961, at the age of 4 months. A systolic thrill was palpable at the left sternal border. A loud grade-IV/VI harsh systolic murmur was heard over the entire precordium, and was maximal at the left sternal border in the third and fourth intercostal spaces. An electrocardiogram was suggestive of biventricular enlargement. Chest roentgenograms showed pneumonia of the left lower lobe, marked enlargement of the right ventricle and left atrium, and pulmonary overcirculation. After the patient recovered from the pneumonia, right heart catheterization and angiocardiography were carried out uneventfully (table 1). It was thought that the patient had an interventricular septal defect with a large left-to-right shunt and severe pulmonary hypertension.

During the next 2 years, she suffered from repeated upper and lower respiratory tract infections and she was hospitalized on two occasions because of pneumonia. Her growth and develop-
Table 1

Results of Right Heart Catheterization

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (months)</th>
<th>Date</th>
<th>IVC</th>
<th>SVC</th>
<th>RA</th>
<th>Right ventricle</th>
<th>Right ventricle</th>
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<td>High pressure</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>MPA</td>
<td>RA</td>
</tr>
<tr>
<td>1. B.S.</td>
<td>5</td>
<td>11-10-61</td>
<td>85</td>
<td>-</td>
<td>86</td>
<td>92</td>
<td>-</td>
</tr>
<tr>
<td>2. P.Y.</td>
<td>5</td>
<td>10-14-63</td>
<td>89</td>
<td>92</td>
<td>89</td>
<td>93</td>
<td>-</td>
</tr>
<tr>
<td>3. K.M.</td>
<td>24</td>
<td>12-16-57</td>
<td>76</td>
<td>78</td>
<td>78</td>
<td>92</td>
<td>90</td>
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<td>46</td>
<td>10-12-59</td>
<td>85</td>
<td>-</td>
<td>81</td>
<td>83</td>
<td>90</td>
</tr>
</tbody>
</table>

IVC, inferior vena cava; SVC, superior vena cava; RA, right atrium; MPA, main pulmonary artery.

Case 3

The patient, K.M., was a product of a full-term pregnancy and weighed 7 lb. 14 ounces at birth. He was admitted to the hospital for cardiac evaluation on December 9, 1957, at the age of 2 years. Physical examination revealed a systolic murmur and an intercostal systolic ejection murmur. Right heart catheterization was carried out on December 9, 1957.

The right heart catheterization was performed as described in Table 1 (column 1) and the diagnosis at this time was a "two-chambered right ventricle" and an interventricular septal defect.

Case 4

The patient, P.Y., was a product of a full-term pregnancy and weighed 5 lb. 14 ounces at birth. She was admitted to the hospital on April 26, 1962, at the age of 4 months for cardiac evaluation. Physical examination revealed a systolic murmur and an intercostal systolic ejection murmur. Right heart catheterization was performed on October 13, 1963, as described in Table 1 (column 1) and the diagnosis at this time was a "two-chambered right ventricle" and an interventricular septal defect.
(table 1) and it was thought that the patient had an interventricular septal defect and questionable pulmonary stenosis.

He was readmitted on October 9, 1959, at the age of 3½ years for re-evaluation. In the interim, he had been asymptomatic and his growth and development had been good. The cardiac findings by physical examination were unchanged. The electrocardiogram and chest roentgenograms revealed no interval change. Right heart catheterization and angiocardiography were carried out uneventfully (table 1) and the patient was thought to have a “two-chambered” right ventricle and an interventricular septal defect.

![Pressure recordings from the right ventricle and pulmonary artery. The studies were separated by an interval of 2 years. (Patient B.S.)](image)

**Figure 1**

**Case 1 (B.S.). A. Chest roentgenogram, posteroanterior (October 31, 1961). The heart is enlarged, the right ventricular outflow tract and the main pulmonary artery are dilated. The peripheral pulmonary vessels are engorged. B. Chest roentgenogram, posteroanterior (Sept. 27, 1962). The heart has decreased in size and the right ventricular outflow tract and main pulmonary artery are less dilated. The peripheral pulmonary vessels are less engorged.**

**Figure 2**
Open-heart surgery was carried out on September 21, 1961, with the aid of extracorporeal support. Hypertrophied muscle bundles were found, which obstructed the entrance to the infundibulum (approximately 5 mm. opening). These bundles were resected. A high interventricular septal defect, approximately 2 cm. in diameter, was closed with an Ivalon patch. His postoperative course was uneventful and he was discharged on October 4, 1961. (This child was included as one of a series of patients in a previous publication.2)

Results

Catheterization

The initial catheterization studies (table 1) on all three patients showed evidence of a large left-to-right shunt at the ventricular level and no evidence of right ventricular ob-

![Figure 3](image1)

**Figure 3**

*Case 1 (B.S.). Angiocardiography of the right ventricle (October 11, 1961). A. Anteroposterior. B. Lateral projection. Aberrant muscle bundles are seen in the sinus portion of the right ventricle.*

![Figure 4](image2)

**Figure 4**

*Case 1 (B.S.). Angiocardiography of the right ventricle (October 14, 1963). A. Anteroposterior. B. Lateral projection. Arrows point to markedly hypertrophic aberrant muscle bands creating a stenotic segment in the distal sinus part of the right ventricle.*
striction in patients B.S. and P.Y. The degree of stenosis in K.M. was minimal. The second catheterization studies, however, demonstrated an obstruction in the right ventricle in all three patients. This degree of stenosis was quite marked in patient B.S., moderate in K.M., and mild in P.Y. The change in the intracardiac hemodynamics is demonstrated in the recordings of intracardiac pressure in patient B.S. (fig. 1) taken at 4 months and 28 months of age.

**Roentgenologic Examinations**

The initial chest roentgenograms in the three patients showed changes characteristic of a left-to-right shunt at the ventricular level,

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**Figure 5**

*Case 2 (P.Y.). Angiocardiography of the right ventricle (May 30, 1962). The arrow points to a filling defect in the contrast medium in the proximal outflow tract caused by a thin abnormal muscular band extending into the sinus part of the right ventricle.*

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**Figure 6**

*Case 2 (P. Y.). Angiocardiography of the right ventricle (September 10, 1963) A. Anteroposterior. B. Lateral projection. The hypertrophy of the aberrant muscle bundles is seen in the proximal outflow tract and in the sinus part immediately below (arrow).*
i.e., biventricular and left atrial enlargement, a prominent right ventricular outflow tract, and pulmonary vascular engorgement. Subsequent chest roentgenograms obtained during the 1- to 2-year period of follow-up showed a reduction in the relative size of the heart. The right ventricular outflow tract became less prominent, the pulmonary vessels less engorged, and the dorsal bulge of the left atrium less pronounced. The change was striking in the first case (B.S.; fig. 2A, B), and less so, but still obvious in the other two cases.

The first angiocardiographic examination on B.S. showed an abnormal muscular band in the distal sinus part of the right ventricle (fig. 3). The second examination performed 2 years later demonstrated again the abnormal muscular band now markedly enlarged creating a subinfundibular obstruction. The outflow tract of the ventricle distal to the obstruction was dilated (fig. 4).

The first angiocardiographic examination on P.Y. showed a thin abnormal muscular band in the subinfundibular region (fig. 5).

The second examination, which was performed 18 months later, showed marked hypertrophy of the muscle in the subinfundibular region with apparent prolongation of the outflow tract as well as obstructive bands in the outflow tract (fig. 6).

Only one angiocardiographic examination was performed on patient K.M. and two abnormal muscular bands were seen in the septal portion of the outflow tract. One of the bands made a deep indentation in the ventricular lumen immediately proximal to the outflow tract and inserted in the anterior ventricular wall (fig. 7).

Discussion

The catheterization data (table 1) reveal definite evidence that an obstruction developed in the right ventricle in the interval between the two studies. This was confirmed by the angiocardiographic examinations, which demonstrated progressive hypertrophy of the aberrant muscular bands, and the plain chest roentgenograms, which showed a decrease in degree of pulmonary vascular en-

Figure 7

Case 3 (K.M.). Angiocardiography of the right ventricle (October 12, 1959). A. Anteroposterior projection. Aberrant muscle bands in the proximal part of the outflow tract and in the sinus part of the ventricle (arrows). B. Lateral projection. The bands are seen anteriorly in the outflow tract and as a filling defect below the outflow tract (arrows).
gorgement at the time of the second examination.

The logical sequence of events in these patients would be that the bands were congenital but produced little or no obstruction in early infancy so that their presence was not shown by catheterization but was suggested by the angiocardiograms. The muscular bands became progressively more hypertrophied, so that at the time of the second examination the complete picture of obstructive aberrant muscle bands was shown both by catheterization and angiocardiography. (A mild right ventricular pressure gradient was present in patient K.M. at the time of the first examination but this study was done at the age of 2½ years and presumably the bands already had enough time to develop sufficient hypertrophy to be slightly obstructive.) We do not suggest that this course is followed by all patients born with aberrant right ventricular muscle bundles. In some patients the aberrant muscle bundles may be obstructive at birth. In others, the degree of hypertrophy may never progress sufficiently to produce right ventricular obstruction. As with other congenital cardiac malformations there is a spectrum of severity. The stimulus for the progression of hypertrophy of the bands is not known. The effect, if any, of the left-to-right flow through the ventricular septal defect upon the rate of hypertrophy of the aberrant bands in the three patients of this series cannot be ascertained. It must be remembered, however, that two patients in our original series and a number of patients reported by Lucas had obstructive aberrant bands without a ventricular septal defect.

The progressive hypertrophy of these aberrant muscle bands is analogous to the progressive hypertrophy of the infundibular muscle in a group of patients described by Gasul. In infancy these patients had the findings of a ventricular septal defect and the shunt was left to right. When these same patients were studied at a later time the picture was that of tetralogy of Fallot. There was a pronounced degree of infundibular hypertrophy so as to be obstructive, and the shunt through the ventricular septal defect was now right to left. Angiocardiographic studies were not reported so that there is doubt about the exact site of obstruction in the right ventricle. It is conceivable that the patients reported by Gasul might well have had aberrant muscular bands rather than infundibular obstruction.

Our studies suggest that the muscle bands are congenital in origin and, in some patients, undergo progressive hypertrophy in time so that the fully developed picture of the “two-chambered” right ventricle emerges beyond the age of 2 years. Early recognition of this congenital malformation is important because surgical intervention might be carried out with less risk when the hypertrophied bands are smaller rather than at a later age when the hypertrophy would be much greater.

Conclusion

The aberrant hypertrophic muscular bands which divide the right ventricle into chambers are congenital in origin and undergo progressive hypertrophy with time. The fully developed anatomic and hemodynamic picture is usually seen after the age of 2. This sequence of events is proposed after a study of the data on three patients who had catheterization and angiocardiographic examinations at intervals of 1½ to 2 years.

References


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Circulation. 1964;30:679-685
doi: 10.1161/01.CIR.30.5.679

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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