Tetralogy of Fallot with Anomalous Tricuspid Valve Simulating Pulmonary Stenosis with Intact Septum

By Henry N. Neufeld, M.D., Dwight C. McGoon, M.D., James W. DuShane, M.D., and Jesse E. Edwards, M.D.

The anatomic and clinical features and the results of hemodynamic investigations in the tetralogy of Fallot are now universally known. It has been emphasized that anatomic deviations in the degree of pulmonary stenosis or in the size of the ventricular septal defect may yield different patterns of clinical features and hemodynamics in this anomaly.

Recently, we observed 3 cases in which the clinical picture suggested severe pulmonary stenosis with an intact ventricular septum associated with a right-to-left shunt. However, cardiac catheterization showed that the shunt was at the ventricular level. It was demonstrated also that the right ventricular systolic pressure was significantly greater than the systolic pressure in systemic arteries. Therefore, it was assumed clinically that these cases were examples of severe pulmonary stenosis with a small ventricular septal defect.

Pathologic examination in one instance (case 1) and the anatomic findings at operation in the other two (cases 2 and 3) demonstrated that the ventricular septal defects were of the usual "large" size seen in the tetralogy of Fallot. In addition to these findings, however, an anomaly of the tricuspid valve was present in all 3 cases. This took the form of a flap of accessory valvular tissue that partially closed the ventricular septal defect during ventricular systole.

It is the purpose of this paper to illustrate the clinical and pathologic findings in these 3 cases. To the best of our knowledge, this anatomic entity has not been described previously in relationship to the associated alterations in hemodynamics.

Presentation of Cases

Since the findings were similar in all 3 patients, these are described together for the group. The clinical findings are summarized in table 1 and the hemodynamic data in table 2.

Clinical Data

The patients were 2 boys and a girl, aged 12, 5, and 7 years, respectively. Two had been cyanotic since early infancy, whereas cyanosis had been observed in the 7-year-old patient since the age of 5 years. There was no history of "squatting" in any of the patients. The main complaints were shortness of breath and changes related to reduced cardiac functional capacity. Clubbing of the digits and distended cervical veins were noted in each patient. One patient (case 1) had signs of severe congestive failure of the right side of the heart.

All 3 patients had a typical murmur of stenosis, with maximal intensity at the pulmonary zone, and a diminished nonduplicated (pure) second pulmonic sound. In case 1, a febrile illness had been present for several weeks in the immediate period before our observation; as judged from later studies, this probably represented an episode of bacterial endocarditis, although this specific diagnosis had not been made. Congestive cardiac failure began at the time of the febrile illness and was the cause of death in this patient. In cases 2 and 3, corrective operations were done with the aid of the bypass machine and extracorporeal circulation. In both of these cases, the postoperative course and follow-up studies indicated satisfactory results.

Radiologic and Electrocardiographic Findings (Fig. 1-3)

The radiologic examination showed slight to great enlargement of the heart. The pulmonary vasculature was diminished in each case. Evidence of poststenotic dilatation of the pulmonary artery was absent. In case 1, a right aortic arch was observed radiologically.

Electrocardiographically, each patient had a normal sinus rhythm. The P waves were peaked in lead II in all instances. Right axis deviation ranging from +100 to +150 degrees was observed. Signs of pronounced right ventricular hypertrophy were present in each.
Table 1
Clinical Data in Tetralogy of Fallot with Anomalous Tricuspid Valve

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yr.) and sex</th>
<th>Cardiac functional capacity, N. Y. Ht. Class.</th>
<th>Right vent. impulse</th>
<th>Onset of cyanosis</th>
<th>Clubbing</th>
<th>Distended neck veins</th>
<th>Congest. heart failure</th>
<th>Systolic murmur</th>
<th>Pulmonic second sound</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>12 M</td>
<td>IV</td>
<td>Normal</td>
<td>Early infancy</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Grade 2</td>
<td>Diminished, pure</td>
</tr>
<tr>
<td>2</td>
<td>5 M</td>
<td>IV</td>
<td>Forceful</td>
<td>Early infancy</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>Grade 2</td>
<td>Diminished, pure</td>
</tr>
<tr>
<td>3</td>
<td>7 F</td>
<td>III</td>
<td>Forceful</td>
<td>At 5 years</td>
<td>+</td>
<td>+</td>
<td>–</td>
<td>Grade 4</td>
<td>Diminished, pure</td>
</tr>
</tbody>
</table>

Table 2
Physiologic Data in Tetralogy of Fallot with Anomalous Tricuspid Valve

<table>
<thead>
<tr>
<th>Cases</th>
<th>Average pressures, mm. Hg</th>
<th>Average blood oxygen saturation, %</th>
<th>Pulmonary flow, L./min.⁶</th>
<th>Pulmonary flow index, L./min./M.²</th>
<th>Systemic flow, L./min.</th>
<th>Right-to-left shunt, % of systemic flow</th>
<th>Systemic flow index, L./min./M.²</th>
</tr>
</thead>
<tbody>
<tr>
<td>1*</td>
<td>17/11</td>
<td>22</td>
<td>0.3</td>
<td>0.4</td>
<td>3.8</td>
<td>88</td>
<td>5</td>
</tr>
<tr>
<td>2*†</td>
<td>138/5-15</td>
<td>21</td>
<td>160/1-20</td>
<td>208/12</td>
<td>287</td>
<td>119/2-20</td>
<td>37/8</td>
</tr>
<tr>
<td>3‡‡</td>
<td>97/73</td>
<td>27</td>
<td>118/2-20</td>
<td>119/69</td>
<td>97/73</td>
<td>115/75</td>
<td>95/70</td>
</tr>
</tbody>
</table>

*Dye-dilution curves showed a right-to-left shunt at the ventricular level.
†Pressures obtained during operation.
‡Cardiac catheterization performed elsewhere.
§Calculation of pulmonary flow was based on the assumption that pulmonary venous blood is fully saturated; since the child was studied under balanced amnesia and analgesia, some degree of pulmonary venous desaturation may have been present.

Physiologic Findings

Cardiac catheterization was done in each case, and dye-dilution studies were made in 2. The results showed remarkable similarity among the 3 cases (table 2). We are indebted to Dr. C. R. Cumming, of Winnipeg, Manitoba, who kindly supplied the data obtained at cardiac catheterization in case 3.

The striking feature in all 3 cases was the increased right ventricular systolic pressure, which was significantly greater than the systemic pressure (fig. 4). The gradient of systolic pressure between the two systems ranged from 41 to 97 mm. Hg. In each case, the pressure tracings showed evidence for both valvular and infundibular stenosis. In case 1, this was established by measurements of infundibular and pulmonary arterial pressures. In the other 2 cases, the catheter did not enter the pulmonary trunk, but an infundibular chamber was assumed; here, the pressure...
Figure 1
Case 1. Posteroanterior thoracic roentgenogram and electrocardiogram (see text for details).

Figure 2
Case 2. Posteroanterior thoracic roentgenogram and electrocardiogram (see text for details).

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Figure 3

Case 3. Posteroanterior thoracic roentgenogram and electrocardiogram (see text for details).

Figure 4

Case 2. Pressures obtained simultaneously within right ventricle and femoral artery, showing gradient in pressure between the two circulations, with the right ventricular pressure exceeding the systemic pressure.

was high but it was less than that in the main right ventricle. These observations were interpreted as reflecting stenosis at a point leading into the infundibular chamber and indicating severer stenosis at the exit of the chamber, the latter point most likely being the pulmonary valve.

Right atrial pressures were greatly increased in all 3 cases, and prominent “a” waves were noted. Pronounced peripheral oxygen desaturation was present in all cases. In the 2 patients (cases 1 and 2) in whom dye-dilution studies were performed, a right-to-left shunt was demonstrated at the ventricular level.

Pathologic Data

Case 1. The interior of the heart showed a large ventricular septal defect 1.6 cm. in diameter in the uppermost portion of the ventricular septum behind a hypertrophied and vertical crista supraventricularis. The crista divided the outflow tract of the right ventricle into a posterior subaortic channel and an anterior subpulmonary channel (fig. 5). The entrance to the latter was only 3 mm. in diameter. Above this, the infundibular subpulmonary chamber was dilated to a diameter of about 1.5 cm. The pulmonary valve was hi-
cuspid and stenotic. The aorta arose from both ventricles above the ventricular septal defect.

Up to this point, the description of this heart is that of the classic anatomic tetralogy of Fallot. However, an additional malformation related to the tricuspid valve was present that evidently had modified the functional state from that in the usual tetralogy of Fallot. The tricuspid valve possessed the usual 3 leaflets but, in addition, an accessory flap extended from the ventricular aspect of the tricuspid annulus into the right ventricular cavity (fig. 6a). The lower edge of the accessory flap received a set of chordae that inserted into the papillary muscle of the conus of the right ventricle. The upper edge of the flap was attached to the lower end of the ventricular septal defect. The long axis of the accessory flap was parallel to the long axis of the ventricular septal defect. An attempt was made to simulate the effect of ventricular systole on this anomalous flap. When pressure was exerted against the lower aspect of the flap, it was possible to have it engage and almost close the ventricular septal defect (fig. 6b).

An additional remnant of accessory valvular tissue extended upward from the upper edge of the ventricular septal defect to the right ventricular wall. The posterior leaflet of the tricuspid valve revealed an accessory orifice about which chordae were inserted (fig. 7a).

In addition to the afore-mentioned changes, both the tricuspid valve and the upper edges of the stenotic zone in the subpulmonary tract showed partly calcified vegetations that histologically were consistent with the appearance of healed bacterial endocarditis.

The left ventricular cavity contained a mural thrombus undergoing organization at its apical portion (fig. 7b). This lesion was interpreted as the site of healed mural endocarditis.

The aortic arch was anomalous. The arch passed over the right main bronchus, and the descending aorta lay to the right of the esophagus in its greater course through the thorax. In the lower portion of the thorax, the aorta deviated to the left and entered the abdomen through a normally positioned aortic hiatus of the diaphragm.

The branches of the aortic arch were mirror images of the normal. The following branches arose from before backward: the left innominate artery, the right common carotid artery, and the right subclavian artery. The left innominate ar-
tery was long, and a segment was present between the origin of the left common carotid and left subclavian arteries that showed an infolding of the superior aspect like that seen in coarctation of the aorta. The ligamentum arteriosum was left-sided and extended from the left pulmonary artery to the left innominate artery, inserting opposite the origin of the left subclavian artery.

Study of the interior of the right atrium showed that the oriﬁces of the superior and inferior venae cavae were in normal positions. The foramen ovale was closed. The right atrial ostium of the coronary sinus was closed by a delicate flap, presumably representing adhesion of the valve of the coronary sinus to the right atrial wall. The only recognized outlet for coronary venous blood was a series of dilated thebesian veins in the left atrium. A persistent left superior vena cava was not present; such an absence also is noted in other cases of atresia of the right atrial ostium of the coronary sinus.  

Cases 2 and 3. In these 2 cases, operations were performed after necropsy had been done in the ﬁrst case. The basic malformation, as identiﬁed by the surgeon, in cases 2 and 3 was the tetralogy of Fallot, including the presence of a large ventricular septal defect. In each instance, an accessory flap of tissue also lay inferior to the tricuspid valve and in close relationship to the ventricular septal defect. The flap in each case was in such a position and of such nature that it was considered possible that it had been able to close the ventricular septal defect partially during ventricular systole.

Discussion

Although the wide functional range represented by the tetralogy of Fallot is now well known, 1,10,11 the 3 cases reported here must be considered as a special group from pathologic and hemodynamic points of view.

The patients presented a general clinical picture of severe obstruction to outflow from the right ventricle, associated with cyanosis. The clinical history did not aid in the differential diagnosis. The physical ﬁndings were most suggestive of severe pulmonic stenosis with an intact septum. A forceful impulse of

Figure 6

Case 1. a. Outﬂow tract of right ventricle and tricuspid valve. Behind the crista supraventricularis (C.S.) is a large ventricular septal defect. The arrow points to an accessory flap of tissue attached to the ventricular aspect of the tricuspid valve. In this view, the ﬂap is in a simulated diastolic position. b. Same specimen. A probe has been placed under the accessory ﬂap of the tricuspid valve (arrow) and pushed upward, simulating the effect of systole on the accessory ﬂap. This simulated systolic position of the ﬂap shows that the ventricular septal defect is all but closed by the ﬂap when it is pressed into the defect.

Figure 7

Case 1. a. Right atrium and tricuspid valve. The posterior leaflet of the tricuspid valve has an accessory congenital orifice (point of arrow). The valve of the inferior vena cava is represented by a network of delicate ﬁbrous strands. Inferior to the orifice of the inferior vena cava, the orifice of the coronary sinus is closed. Vegetations of bacterial endocarditis are seen on the atrial aspect of the tricuspid valve. b. Left ventricle and aortic valve. Inferior to the aortic valve is a large ventricular septal defect. The aorta straddles the defect. An organizing mural thrombus is present in the apical portion of the left ventricle.
the right ventricle is unusual in the tetralogy of Fallot, but it was observed in 2 of these 3 patients. Distended cervical veins as a clinical sign of increased right atrial pressure are unusual in the tetralogy of Fallot but are often seen in severe pulmonary stenosis with an intact ventricular septum.

Congestive right heart failure is extremely unusual in the tetralogy of Fallot; it was present in 1 of our cases.

The electrocardiogram showed pronounced right ventricular hypertrophy in all cases, especially case 2, in which the deep, sharply inverted T waves in leads from the right side of the precordium suggested that the right ventricular pressure was greater than the systemic pressure.

The radiologic findings of cardiac enlargement in cases 1 and 2 were also unusual for the tetralogy of Fallot, although the presence of a right aortic arch in case 1 was more in favor of this anomaly than it was of pulmonary stenosis with an intact ventricular septum.6

The findings at cardiac catheterization, as already mentioned, showed a significantly higher systolic pressure in the right ventricle than in the systemic circulation. Since it is usually assumed that the right ventricular pressure in the tetralogy of Fallot is about equal to the systemic pressure,6,8 finding of a significant differential immediately places doubt on the diagnosis of a classic tetralogy.

We think that the additional flap on the tricuspid valve was responsible for the increase of right ventricular pressure, as this flap partially closed the ventricular septal defect during systole. This partial closure of a large defect during ventricular systole changes the dynamics from those of the usual tetralogy of Fallot into those representative of cases with severe pulmonary stenosis and a small ventricular septal defect.

The clinical entity of pulmonary stenosis with an anatomically small ventricular septal defect apparently is extremely rare.1, 11, 12 In reported cases of such an anomaly, the hemodynamic and clinical data have been similar to those seen in the present 3 cases.

Summary
An anatomicopathologic entity of the tetralogy of Fallot with an additional finding of an accessory flap of tricuspid valvular tissue is described. In the 3 cases reported in this paper, the outstanding hemodynamic finding was a right ventricular systolic pressure that was significantly greater than the systemic systolic pressure.

The hemodynamic findings were explained by the fact that the accessory tricuspid flap partially closed the large ventricular septal defect.

The clinical picture was similar to that of either severe pulmonary stenosis with an intact ventricular septum or severe pulmonary stenosis with a small ventricular septal defect and a right-to-left shunt at the ventricular level.

Sumario in Interlingua
Es describite un entitate anatomico-pathologic de tetralogia de Fallot con le constatation additional de un lobo accessorii de tissui de valvula tricuspid. In le 3 casos reportate in le presente communication, le plus frapante observation hemodynamic eseva un tension systolic dextero-ventricular significativemente plus grande que le systemic tension systolic.

Le caracteristicas hemodynamic se explicave per le facto que le lobo accessorii tricuspid claudeva partialmente le grande defecto ventriculo-septal.

Le tableau clinic eseva simile a illo de (1) sever stenosis pulmonar con intacte septo ventricular o (2) sever stenosis pulmonar con un miere defecto ventriculo-septal e un shunting dextero-sinistre al nivello ventricular.

References
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We may say on the grounds of incontestible experience, that, in their early stages, they are, in a large proportion of instances, susceptible of a perfect cure; and that, when not, they may, in general, be so far counteracted as not materially, and sometimes not at all, to curtail the existence of the patient. We may, accordingly, predict that the term "disease of the heart," which at present sounds like a death-knell when uttered by the physician, will hereafter become by familiarity not more alarming than the term asthma, under which it is frequently disguised.—J. Hope, M.D. Diseases of the Heart and Great Vessels. London, William Kidd, 1832, p. 20.
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