Paradoxical Results of Infundibular Resection in Tetralogy of Fallot

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The ultimate aim of surgical treatment of Fallot's tetralogy is complete anatomic repair by removal of the obstruction to pulmonary blood flow and closure of the ventricular septal defect. This ideal has been achieved by a small number of surgeons using a pump oxygenator and cardiac arrest. Until it can be realized by all, less complete operations will be performed that will improve the condition without correcting it. Considerable clinical improvement follows infundibular resection by open cardiotomy with or without pulmonary valvotomy. Paradoxically some of these patients may develop a syndrome of heart failure due to the development of a left-to-right shunt through the ventricular septal defect.

The 7 patients discussed in this paper had severe disability due to Fallot's tetralogy. By means of hypothermia infundibular resection and pulmonary valvotomy were carried out under direct vision in 5 patients and infundibular resection alone in the other 2. Except for 1 instance, the operations were successful in that the disability and cyanosis virtually disappeared. At the same time, however, all the patients developed signs of cardiac failure, namely raised venous pressure and cardiac and hepatic enlargement. The purpose of this paper is to explain how these seemingly paradoxical results came about.

There were 7 patients in this series, 2 male and 5 female (table 1). Five of them were aged 6 to 13 and the other 2 were 17 and 26 years. The diagnosis of Fallot's tetralogy had been confirmed by angiocardiography. A Blalock-Taussig operation had been performed on 4 of them 4 to 8 years previously; in 2 of these patients, the operation was repeated on the opposite side. All these operations had been unsuccessful; from the absence of the characteristic murmur, the anastomosis was considered to have closed in all except 1 child who retained a faint continuous murmur. All were cyanotic at rest and showed clubbing of fingers and toes. They became a deeper blue and very breathless after a quick walk of 50 yards or less.

Methods

The operations were performed by Mr. Holmes Sellors under hypothermia at 30 C. After the circulation had been occluded, a small incision was made into the right ventricular outflow tract. The infundibular stenosis was resected by knife, scissors, or punch on the anterior and lateral aspects so that the obstruction between the ventricle proper and the infundibulum was relieved.

Five of the 7 patients had valvular stenosis in addition to the infundibular obstruction. Here the incision in the ventricle was prolonged upwards or a separate incision was made in the anterior wall of the pulmonary artery. The valve, which was cone-shaped, was divided in 2 places from the narrowed orifice toward the valve ring so as to leave an adequate channel.

In the course of the ventriculotomy the ventricular septal defect was well visualized in 2 cases (J.B. and S.S.). Two or 3 sutures were inserted, and the defect appeared to be closed completely in J.B. and partially in S.S. Postoperative catheterization however showed that the defect remained patent in both patients.

Clinical Course

After 6 to 12 months, all 7 patients admitted to little or no disability and were not restricting themselves in any way. Five of them showed no trace of cyanosis at rest (arterial oxygen satura-

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tion 95 per cent or over). Two had slight cyanosis (85 and 88 per cent saturation). The patients were not observed after strenuous effort, but they or their parents said that their color did not change after exercise. The hemoglobin content of the blood returned to normal in all cases (table 1).

The following features of the early postoperative course were common to all patients: cyanosis was replaced by a pink skin color, the jugular venous pressure rose to the level of the angle of the jaw with the patient at 45°, and radiologically the heart size and pulmonary vascular markings increased (fig. 1 and table 1). In the patient J.R., the clinical and radiologic picture of left ventricular failure and pulmonary edema followed the development of right ventricular failure after 1 week but responded promptly to digitalis, mersalyl, and sodium restriction.

On auscultation (fig. 2) a long systolic murmur loudest in the third left interspace replaced the short pulmonary systolic murmur; a pulmonary diastolic murmur appeared in all but 1 patient, and pulmonary valve closure (P₂) became audible in 2 patients with infundibular stenosis only. The murmur appeared to extend through systole and it continued up to or past aortic valve closure (A₂), which was audible but ended before P₂, which was either faint or inaudible.

In the electrocardiogram (fig. 3), the signs of right ventricular hypertrophy changed to right bundle-branch block in 2 cases with T inversion extending as far as V₂ 1 year after operation. Inverted T waves in the chest leads were the only changes after 1 year in 2 cases, and in the remainder, the tracings were unaltered.

Cardiac catheterization demonstrated a left-to-right shunt at ventricular level in all patients. A small right-to-left shunt remained in 2. The degree of shunt is reflected by the ratio of pulmonary to systemic flow: this was 2 to 1 or more in 4 of the 7 patients. The pulmonary systolic pressure was 40 to 65 mm. Hg in 5 cases, but the pulmonary resistance was less than 4 units (320 dynes sec. em.⁻²) in all. The right ventricular diastolic pressure was significantly raised in 3 cases and the right atrial pressure was high in 6 cases with a dominant “a” wave. "Wedge" pulmonary capillary pressure was raised in the 3 patients in whom it was recorded. The catheterization data are detailed in table 2.

**DISCUSSION**

In Fallot’s tetralogy, the right ventricle pumps a considerable amount of its output into the aorta where the resistance is lower
FIG. 1. Chest x-rays before (above) and after (below) operation. The heart size has increased and pulmonary plethora has developed. Left. Male, L.S., age 26. Cardiac-thoracic ratio before operation 48; 1 year later, 57. Right. Female, J.R., age 6. Cardiac-thoracic ratio before operation 39; 9 months later, 58.
than that due to the infundibular or pulmonary valve stenosis. Brock first advocated the removal of the obstruction to the right ventricular output so that the pulmonary blood flow would increase and the amount of blood shunted into the aorta would be reduced or abolished. The object of removing the infundibular or valve obstruction under direct vision in the present cases was to achieve such a redirection of flow. This has occurred but other, perhaps less desirable, sequelae have resulted, similar to those described in a single case by McCord and Blount.

Interventricular Shunt. Although the pressure in both ventricles remained identical in 5 of the 7 patients catheterized after operation, the shunt from right ventricle to aorta disappeared in all except 2 and in these cases it was greatly reduced. However, a left-to-right shunt developed in all 7 patients. The radiologic appearance of pulmonary plethora and of cardiac enlargement were the first signs of this shunt, and it was subsequently confirmed by catheterization. The redirection of blood flow resulted from the removal of the obstruction to the pulmonary flow. The pulmonary vascular resistance was consequently lower than the systemic and the left-to-right shunt developed. The increase of heart size may be partially attributed to the rapid onset of this shunt. Wood has suggested that the disproportionate degree of right ventricular enlargement results from the need for the right ventricle to pump a greater volume at systemic pressure. This increase in right ventricular work was followed in all cases by the development of right ventricular failure within 3 days of operation. The clinical rise of jugular venous present...
sure reflected the rise of mean right atrial pressure and a dominant "a" wave was due to the increased resistance to filling of the ventricle. Radiologic enlargement of the atrium also occurred.

Pulmonary Circulation. The pulmonary circulation was suddenly exposed to a greatly increased blood flow. The normal pulmonary vasculature could accept such an increase with little or no rise in pressure but in Fallot's tetralogy, the pulmonary vessels are abnormal and a two- to four-fold increase in pulmonary systolic pressure resulted from an increase in flow 1½ to 5 times the normal (table 2). Pulmonary capillary venous pressures were recorded in only 3 patients. In 2 children, these values were a little above normal (8 and 13 mm. Hg) and in the third, a man aged 25, there was a considerable increase (28, 22, 25, and 19 mm. Hg for "a," "x," "v," and "y" respectively). The total pulmonary resistance did not exceed 4 units (or 320 dynes sec. cm.⁻²) in any case, and the pulmonary arteriolar resistance was considerably less. The Eisenmenger syndrome was not seen to develop. It would seem that the pressure increase was the result of several factors: transmission of the high right ventricular systolic pressure, increased pulmonary blood flow, raised left atrial pressure, and limitation of distensibility of the pulmonary arterioles.

Pulmonary incompetence was created at operation in 6 of the 7 cases, including 1 with normal pulmonary valves. Keith⁵ proposed that the competence of the pulmonary valves depended on the support of the ventricular muscle that extends to the valve ring, a view confirmed experimentally by Brock.⁶ Although the valvotomy in the present cases was designed to produce a competent valve as far as possible, regurgitation resulted, partly because the newly fashioned flaps failed as a valve, and partly because the supporting musculature of the valve ring had been damaged.

The hemodynamic effect of pulmonary incompetence in this situation is difficult to assess. While it would appear to add further to the right ventricular stroke volume, it is probably of lesser importance. This point is illustrated by case J.B. in whom considerable
Table 2.—Postoperative Cardiac Catheterization

<table>
<thead>
<tr>
<th>Name</th>
<th>SVC* Sat. (%)</th>
<th>RA Sat. (%)</th>
<th>RV Sat. (%)</th>
<th>PA Sat. (%)</th>
<th>PCV Pressure†</th>
<th>Syst. art. Sat. (%)</th>
<th>Blood Flow (L/min.)</th>
<th>Interval after operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>J.B.</td>
<td>65</td>
<td>63, 63</td>
<td>80, 6 71</td>
<td>N.E.</td>
<td>75/0</td>
<td>95</td>
<td>3.0, 2.3</td>
<td>6 mo.</td>
</tr>
<tr>
<td>R.G.</td>
<td>62</td>
<td>58, 0</td>
<td>90, 0 78</td>
<td>65, 85</td>
<td>100/65</td>
<td>96</td>
<td>10.4, 3.3</td>
<td>11 mo.</td>
</tr>
<tr>
<td>S.G.</td>
<td>61, 15</td>
<td>61</td>
<td>75, 10 72</td>
<td>72</td>
<td>7</td>
<td>5</td>
<td>5.4/16</td>
<td>7 weeks</td>
</tr>
<tr>
<td>J.R.</td>
<td>61</td>
<td>2, 0</td>
<td>96, 2 87</td>
<td>40, 69</td>
<td>7</td>
<td>13</td>
<td>16</td>
<td>13 mo.</td>
</tr>
<tr>
<td>S.S.</td>
<td>60</td>
<td>12</td>
<td>90, 15 69</td>
<td>50, 10 70</td>
<td>90/50</td>
<td>Cuff</td>
<td>1.3/1</td>
<td>3 mo.</td>
</tr>
<tr>
<td>L.S.</td>
<td>50</td>
<td>19, 5</td>
<td>110, 20 65</td>
<td>60, 12 77</td>
<td>28</td>
<td>25</td>
<td>110/65</td>
<td>6.0, 2.8</td>
</tr>
<tr>
<td>D.W.</td>
<td>67</td>
<td>2/0, 69</td>
<td>100, 0 79</td>
<td>25, 2</td>
<td>76</td>
<td>125/62</td>
<td>5.7, 3.0</td>
<td>8 mo.</td>
</tr>
</tbody>
</table>

*SVC, superior vena cava; RA, right atrium; RV, right ventricle; PA, pulmonary artery; PCV, pulmonary capillary venous area; Syst. art., systemic artery; Pul., pulmonary blood flow; Sys., systemic blood flow.
†Levels of atrial waves, a, x, v, and y. All pressures in millimeters of mercury.

postoperative cardiac enlargement developed in the absence of clinical pulmonary incompetence. Furthermore, pulmonary incompetence after successful valvotomy for pure pulmonary stenosis is fairly common. Blount et al. have observed 3 such patients for 3½ years without noting any ill effects. The evidence from animals with intact septa is equivocal; when the pulmonary valve is completely excised in dogs, heart failure does not follow at rest, but right ventricular hypertrophy always develops. Diminution or regression of the pulmonary incompetence has not been noted in the present cases over a period of a year, as far as can be judged from auscultation or phonocardiography.

Left Ventricular Function. In Fallot’s tetralogy, the presence of a longstanding right-to-left shunt and an underdeveloped left ventricle may appear incompatible. It should be remembered, however, that the upper margin of the ventricular septal defect is closely related to the dextroposed aortic root. Consequently the shunt traverses only a small part of the left ventricular outflow tract and the body of the ventricle bears no extra load.

There appear to be 2 possible causes of left
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ventricular failure in these cases. The first is transmission of the raised right ventricular diastolic pressure to the left ventricle when right ventricular failure appears. The left atrial pressure would then rise as it did in 3 of our patients in whom it was measured. The second is the inability of the underdeveloped left ventricle to deal with the increased pulmonary venous return consequent on the newly acquired left-to-right interventricular shunt. It is impossible to be sure which of these 2 factors is principally responsible for initiating left ventricular failure. It seems likely that both play a part and are closely interrelated.

This syndrome is not peculiar to Fallot’s tetralogy after infundibular resection, for it has been observed after a Blalock or Pott’s anastomosis. The extent to which it develops after infundibular resection depends on how freely the stenosis is removed. Partial resection, leaving a large systolic gradient between pulmonary artery and right ventricle, is often followed by great improvement, but an inadequate resection may be fatal. These results are presented as only an intermediate step in the complete anatomic correction of Fallot’s tetralogy, the final one being the closure of the ventricular septal defect. In the first 2 patients in this series, Mr. Holmes Selors attempted to close the defect after dealing with the stenosis, but the time available with hypothermic circulatory occlusion was too short and the defects were only partially closed. The future policy is to complete the correction when an efficient bypass is available. It is not clear at present whether Fallot’s tetralogy is best treated by excision of the stenosis and closure of the ventricular septal defect in 2 stages as envisaged here, or in 1 stage as reported by Lillehei and his co-workers.9 Although the former course has been followed faute de mieux, it would appear to have the advantage of allowing the right ventricular outflow tract and pulmonary vasculature to become adjusted to an increased flow while an emergency escape valve remains in the form of the ventricular septal defect.

Summary

After a more or less complete excision of the infundibular stenosis, with or without pulmonary valvotomy in 7 patients with Fallot’s tetralogy, a syndrome emerged with the following features:

Immediate changes: the disappearance of cyanosis, signs of right heart failure with cardiac enlargement, radiologic and, in 1 case, clinical evidence of left heart failure, increase in the duration and intensity of the systolic murmur, appearance of a pulmonary diastolic murmur, increased pulmonary blood flow.

Subsequent changes: regression of clubbing, improved effort tolerance, continued elevation of jugular venous pressure. The possible mechanism of these changes is discussed.

SUMMARIO IN INTERLINGUA

Post le excision plus o minus complete del stenosis infundibular (con o sin valvotomia pulmonar) in 7 patients con tetralogia de Fallot, un syndrome se manifestava con le sequente caracteristicas:

Alterationes immediate. Disparition de cyanose, signos de disfallimento dextero-cardiac con allargamento del corde, indicios radiologic (in 1 caso) clinic de disfallimento sinistro-cardiac, augmento del intensitate e del duration del murmurae systolica, apparition de un murmurae pulmo-diastolic, e augmento del fluxo de sanguine pulmonar.

Alterationes subsequente. Regression del phenomeno de digitos hippocracic, meliorate toleration de effortio, e continue elevation del pression in le venas jugular.

Le mechanismo possibile de iste alterationes es discutite.

REFERENCES


The credit of pointing out the analogy between this condition and angina pectoris, which is ascribed usually to Potain (1870), but which is maintained by Weber to be due to Brodie (1846), belongs in reality to Allan Burns, whose Observations on Some of the Most Frequent and Important Diseases of the Heart (1809) is a well-known storehouse of interesting facts. Since, so far as I know, this distinguished writer's connection with this supposed new theory has not been pointed out (except in the second edition of my Practice), I will read to you in full what he says on the subject: "Such a state of the arteries of the heart (referring to atheroma) must impair the function of that organ. It has been long known, that although the heart is always full of blood, yet it can not appropriate to its own wants a single particle of fluid contained in its cavities. On the contrary, like every other part, it has peculiar vessels set apart for its nourishment. In health, when we excite the muscular system to more energetic action than usual, we increase the circulation in every part, so that to support this increased action the heart and every other part has its power augmented. If, however, we call into vigorous action a limb round which we have with a moderate degree of tightness applied a ligature, we find that then the member can only support its action for a very short time, for now its supply of energy and its expenditure do not balance each other; consequently, it soon, from a deficiency of nervous influence and arterial blood, fails and sinks into a state of quiescence. A heart, the coronary vessels of which are cartilaginous or ossified, is in nearly a similar condition; it can, like the limb begin with a moderately tight ligature, discharge its functions so long as its action is moderate and equal. Increase, however, the action of the whole body, and along with the rest that of the heart, and you will soon see exemplified the truth of what has been said, with this difference, that as there is no interruption to the action of the cardiac nerves, the heart will be able to hold out a little longer than the limb.

"If a person walks fast, ascends a steep, or mounts a pair of stairs, the circulation in a state of health is hurried, and the heart is felt beating more frequently against the ribs than usual. If, however, a person, with the nutrient arteries of the heart diseased in such a way as to impede the progress of the blood along them, attempt to do the same, he finds that the heart is sooner fatigued than the other parts are, which remain healthy. When, therefore, the coronary arteries are ossified, every agent capable of increasing the action of the heart, such as exercise, passion, and ardent spirits, must be a source of danger."—William Osler, M.D. Lectures on Angina Pectoris and Allied States, 1897.
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