Complications Following Infundibular Resection in Fallot's Tetralogy

By Malcolm C. McCord, M.D. and S. Gilbert Blount, Jr., M.D.

Two patients are presented to demonstrate complications that may arise following infundibular resection in patients with the tetralogy of Fallot. An aneurysm of the infundibular chamber developed in one patient following removal of the infundibular stenosis. A left to right blood flow at the ventricular level occurred postoperatively in a second patient with marked cardiac enlargement and left ventricular hypertrophy.

The introduction of the Blalock-Taussig procedure in 1945 resulted in a dramatic change in the prognosis of patients with the tetralogy of Fallot. Thousands of patients with this anomaly have experienced an amelioration of symptoms and an increase in life span as a result of this operative procedure. However, this form of surgical therapy is admittedly palliative, consisting as it does in the creation of an additional cardiovascular defect. The ultimate goal in the operative therapy of patients with the tetralogy of Fallot is correction of the basic defect, namely the valvular and/or infundibular stenosis and the ventricular septal defect. A partial realization of this goal has been accomplished by the correction of the valvular or infundibular stenosis. This procedure has been carried out by Brock with a reasonable mortality rate and with clinical results at least as beneficial as those described following the shunt procedure.

The evolution of techniques permitting open heart surgery has further enhanced the possibility of correction of the basic defects in this anomaly. Experience with infundibular resection using open heart techniques has been limited to a small number of patients at this institution; however, definite limitations of the procedure have already been realized. Two patients are presented at this time to emphasize complications that may arise from infundibular resection in the tetralogy of Fallot when concomitant closure of the ventricular defect is not accomplished.

From the Cardiovascular Laboratory, University of Colorado School of Medicine, Denver, Colo.
This study was supported by a United States Public Health Service Grant H-1208.

Case Reports

Case 1. W. H., a four year-old white boy, had been cyanotic since birth, and showed a slow weight gain and delayed growth. Cyanosis became more pronounced with the onset of walking at 10 months of age and squatting was frequent. There was a progressive limitation in exercise tolerance to one half a block. Episodes of semiconsciousness with paroxysmal dyspnea had occurred with increasing frequency since one year of age.

Physical examination revealed a small boy falling in the 10th percentile for height and weight. There was cyanosis of the lips, mucous membranes and nailbeds with clubbing of the digits. A grade 4 systolic murmur of harsh quality was audible along the left sternal border with maximum intensity in the third intercostal space.

Fluoroscopic examination showed a heart at the upper limits of normal in size with a concavity in the region of the main pulmonary artery and an uplifted cardiac apex (fig. 1A). The peripheral lung fields were abnormally clear. The aorta was increased in size and descended on the left.

The electrocardiogram showed a pattern of right ventricular hypertrophy with an R wave 27 mm. in amplitude in precordial position V; and an intrinsiceud deflection time of 0.04 second.

On Jan. 28, 1954 thoracotomy was performed by Dr. Henry Swan, using hypothermia and inflow occlusion. An incision was made through the infundibular chamber and a localized stenotic ring of tissue was visualized. This structure was excised and the ventricular incision closed. The postoperative course was uneventful and the patient returned home on the fifteenth postoperative day.

The exercise tolerance increased significantly and the degree of cyanosis decreased. Seven weeks postoperatively the patient developed fever and cough productive of blood-streaked sputum. Physical examination revealed a visible and palpable heaving pulsation in the left second and third intercostal spaces. A grade 2 rough systolic murmur was audible along the left sternal border. The second heart sound in the left second intercostal space was increased in intensity. Fluoroscopic examination disclosed a
pulsating mass in the region of the main pulmonary artery (fig. 1B). The electrocardiogram showed an inversion of previously upright T waves over the right precordial leads. The patient was hospitalized and an exploratory thoracotomy was performed on March 13, 1954. A large, thin-walled aneurysm was found arising from the infundibular chamber of the right ventricle. This was not a false aneurysm arising at the site of the previous incision in the infundibular chamber. Blood flow was occluded, and the aneurysm was opened and an endoaneurysmorrhaphy was performed with removal of the major portion of the sac. Cardiac arrest occurred with temporary reinsertion of a regular rhythm but the patient died four hours later.

Postmortem examination revealed a heart weight of 120 Gm., as compared with a body weight of 12 Kg. The right ventricle was hypertrophied, the wall measuring 9 mm. in thickness. A fibrous base was present in the area of the excised infundibular stenosis. The infundibular chamber was thin walled and showed the recent incision. The original infundibular incision was well healed. A 1.7 by 1.5 cm. ventricular septal defect was present with the aorta overriding the defect. The aorta measured 4.5 cm. in circumference while the pulmonary artery was 1.5 cm. in circumference.

Comment. The development of an aneurysm of the infundibular chamber following resection of an infundibular stenosis constitutes an unusual complication of this surgical approach to the tetralogy of Fallot. In the absence of evidence of a false aneurysm at the site of the incision or of infarction of the myocardium in this area it seems justified to suggest that this aneurysm developed as a result of the sudden increase in pressure and increase in blood flow through the previously existing infundibular chamber. At the time of the initial surgery the infundibular chamber was a thin walled structure protected from the high right ventricular systolic pressure and from a high volume of blood flow by the presence of the infundibular stenosis. Following extirpation of the infundibular stenosis this chamber was suddenly exposed to the stress of the elevated right ventricular systolic pressure and to an increased blood flow. It is likely that an element of functional valvular pulmonic stenosis complicated the hemodynamic derangement as the small pulmonic valve ring would not accommodate the increase in blood flow. The critical factor in the production of this aneurysm is considered to be the abrupt change in the hemodynamic pattern more than the magnitude of the changes in themselves. Therefore, it is proposed that the formation of the aneurysm developed as a response to this abrupt change in hemodynamics before compensatory hypertrophy of
the myocardium in the wall of the infundibular chamber could become established.

**Case 2.** P. E., a 12-year-old boy, was cyanotic and presented cardiac murmurs at birth. Development was retarded with an onset of walking at 20 months of age and talking at 36 months of age. Cyanosis became more marked with the onset of walking and severe ease of fatigue, exertional dyspnea and squatting occurred. Episodes of paroxysmal dyspnea developed at 7 years of age and were associated with loss of consciousness and deep cyanosis. The patient's exercise tolerance was limited to walking one-half block prior to surgery.

Physical examination revealed cyanosis of the lips and nailbeds with severe clubbing of the digits. The heart was normal in size and no thrills or shocks were present. A harsh systolic murmur of grade 3 intensity was audible along the left sternal border with maximum intensity in the third intercostal space. The second heart sound in the second left intercostal space was normal in intensity and was pure in quality.

The fluoroscopic examination showed a decreased vascularity of the lung fields (fig. 2A). The right pulmonary artery was small and showed a reduced amplitude of pulsations. The aorta was increased in size and descended on the left. A slight concavity was present in the area of the main pulmonary artery. A small bulge was apparent high on the ventricular segment considered to represent an infundibular chamber. The cardiac apex was uplifted suggesting right ventricular hypertrophy. The electrocardiogram (fig. 3) presented a pattern suggesting right ventricular hypertrophy.

Cardiac catheterization was performed on March 16, 1953. The significant results are summarized in table 1. The catheter was introduced into the right ventricle where a pressure of 120/38 mm. Hg was recorded, and was advanced into the aorta and into the pulmonary artery. The pressure tracing recorded during the withdrawal of the catheter from the pulmonary artery to the right ventricular area is shown in figure 4. An intermediate pressure is present representing an infundibular chamber. Open heart surgery was performed by Dr. Henry Swan on April 21, 1953 utilizing hypothermia and inflow occlusion. The infundibular chamber was incised and a localized diaphragm-like infundibular stenosis excised under direct vision. The infundibular orifice was increased from an initial 3 mm. diameter to an estimated 10 mm. diameter. The incision was closed and circulation re-established after a total period of eight minutes of occlusion. The postoperative course was uneventful and the patient returned home May 11, 1953.

An immediate and striking increase in exercise tolerance occurred, so that the patient was able to ride a bicycle, play baseball, walk a distance of at least a mile. and was able to "keep up" with friends in all respects. Re-examination Feb. 5, 1954 revealed no cyanosis and marked regression in the degree of clubbing of the digits. The heart was enlarged to percussion, a vigorous apical thrust was visible and palpable, and a systolic thrill was present along the sternal border. A harsh systolic murmur, grade 5 in intensity, was audible along the
The preoperative, A, and postoperative, B, electrocardiogram of case 2 demonstrating a change from a pattern of right ventricular hypertrophy to a pattern of left ventricular hypertrophy and left bundle branch block.

Fluoroscopic examination showed a normal vascularity of the lung fields. The right pulmonary artery was small but exhibited a normal amplitude of pulsation. There was a marked increase in the heart size (fig. 2 B) with a configuration suggesting left ventricular enlargement.

The electrocardiogram showed a striking change with the development of a pattern suggesting left bundle branch block with left ventricular hypertrophy (fig. 3).

Cardiac catheterization studies (table 1) demonstrated an increase in the pulmonary artery pressure.
TABLE 1.—The Pre- and Postoperative Catheterization Data Determined in Case 2

<table>
<thead>
<tr>
<th></th>
<th>Hemoglobin</th>
<th>Pressure, mm. Hg</th>
<th>Blood Oxygenation, Volumes Per Cent,</th>
<th>Cardiac Index, L/min./m²</th>
<th>Blood Shunt, L/min./m²</th>
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<tbody>
<tr>
<td></td>
<td>Grams/Per</td>
<td>Interior vena cava</td>
<td>Right atrium</td>
<td>Pulmonary artery</td>
<td>Aorta</td>
</tr>
<tr>
<td>Preop., 16 March 1953</td>
<td>19.8</td>
<td>17.85</td>
<td>7.86</td>
<td>12.90</td>
<td>60.4</td>
</tr>
<tr>
<td>Postop., 5 Feb. 1954</td>
<td>14.4</td>
<td>12.66</td>
<td>63.5</td>
<td>13.90</td>
<td>75</td>
</tr>
</tbody>
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Fig. 4. The preoperative pressure tracing in case 2 showing a low pulmonary artery pressure, P.A., and intermediate infundibular chamber pressure, I.C., and a high right ventricular pressure, R.V. A short segment of the infundibular pressure tracing was removed to facilitate graphic presentation.

Fig. 5. The postoperative pressure tracing in case 2 showing an increase in pulmonary artery pressure, P.A., and a decrease in right ventricular pressure, R.V. There is no intermediate pressure area indicating an infundibular chamber as seen preoperatively.

Fig. 6. The postoperative pressure tracing in case 2 on withdrawal of the catheter from the aorta into the right ventricle. A significant systolic pressure gradient is present between the aorta and right ventricle.

Intermediate pressure levels representing an infundibular chamber as noted preoperatively (fig. 4). The catheter was advanced into the aorta and a pressure of 75/60 mm. Hg determined. On withdrawing the catheter from the aorta into the right ventricle a right ventricular pressure of 57/8 mm. Hg was recorded. The continuous pressure tracing during this drawback is shown in figure 6 and demonstrates the systolic pressure gradient from the aorta to the right ventricle.

Re-examination in September, 1954 revealed maintenance of normal exercise tolerance. A brachial arterial oxygen saturation of 98 per cent at rest and 87 per cent following vigorous exercise was determined. Auscultation revealed the appearance of a high pitched blowing diastolic murmur in the second left intercostal space. This murmur was interpreted as indicating the development of functional pulmonic insufficiency.

Comment. The sequence of events occurring in this patient represents a second form of cardiovascular response following resection of infundibular stenosis. The infundibular resection
removed the high resistance to blood flow into the pulmonary artery and introduced into the hemodynamic pattern the relatively low resistance of the pulmonary vascular bed. The result of the removal of the obstruction to pulmonary flow was a decrease in right ventricular pressure, the establishment of a systolic pressure gradient between the left and right ventricles, and a left to right blood flow through the ventricular defect. The hemodynamic pattern was therefore essentially that of an isolated large ventricular septal defect. The residual systolic pressure gradient noted postoperatively between the right ventricle and pulmonary artery (fig. 5) suggests a mild degree of valvular pulmonic stenosis, a mild degree of residual infundibular stenosis, or functional pulmonic stenosis resulting from the relatively small pulmonic valve ring. The relative lack of significance of the anatomic position of the aortic root in the hemodynamic pattern associated with a ventricular defect is well illustrated in this patient. Despite the overriding of the aorta in relation to the ventricular septal defect the right ventricular pressure was significantly less than the left (fig. 6) and the previously existing right to left shunt was completely abolished at rest as demonstrated by the full arterial oxygen saturation. It has been proposed that the presence of a ventricular defect and an overriding aorta results in an identical systolic pressure in the left and right ventricles so that infundibulec- tomy could not reduce the right ventricular pressure in the tetralogy of Fallot. The data presented at this time and studies on patients with isolated ventricular defects demonstrate that this view is not necessarily valid. The presence of a pressure gradient between the two ventricles in patients with a ventricular defect depends upon the relative resistance offered by the pulmonary and systemic circulations and upon the size of the ventricular defect, with the position of the aortic root being of secondary importance.

Following surgery, the dominant physiologic abnormality consisted of the abrupt establishment of a left to right shunt through the ventricular defect. This was reflected dynamically by the dilatation and hypertrophy of the left ventricle, and by the increase in pulmonary blood flow with progressive dilatation of the pulmonic valve ring resulting in functional pulmonic insufficiency. The immediate clinical manifestation of these changes was a striking improvement in symptomatology exceeding that usually observed following a shunt procedure. However, the long term prognosis remains in doubt as it is anticipated that this patient will pursue the natural history of a patient with a large ventricular defect. Closure of the ventricular defect is therefore planned in this patient at such time as this operative procedure is feasible.

**Discussion**

The patients presented in this communication illustrate complications that may arise as a result of surgical relief of the infundibular stenosis without the concomitant closure of the ventricular defect in patients with the tetralogy of Fallot. The course in these two patients emphasizes the inherent balance of dynamic factors in this congenital anomaly. The pulmonic stenosis controls the magnitude of the left to right shunt through the ventricular defect while the ventricular defect limits the increase in right ventricular systolic pressure. One application of this concept has been the operative production of partial pulmonary occlusion in patients with large ventricular defects and high magnitude pulmonary blood flow. Thus, sole correction of either the pulmonic stenosis or the ventricular defect is not a desirable definitive measure in patients with the tetralogy of Fallot. Obviously, considerable nicety of judgement is demanded at the time of surgery to determine the precise extent of infundibular resection that will result in a beneficial degree of increased pulmonary blood flow.

It is recommended, therefore, that the direct surgical approach to the pulmonic stenosis with infundibular stenosis be considered as merely an interim procedure pending the development of techniques permitting simultaneous correction of both the valvular stenosis and/or the infundibular stenosis and the ventricular defect. It is anticipated that techniques per-
mitting such procedure on a wide scale with a reasonable mortality will soon be realized.

**Conclusion**

Two patients with a tetralogy of Fallot are presented who demonstrate complications that may occur following infundibular resection. An aneurysm of the infundibular chamber developed in one patient and a significant left to right blood flow developed in the second patient with resulting marked enlargement of the left ventricle.

**SUMMARIO IN INTERLINGUA**

Es presentate duo casos pro demonstrar complicationes que pote occurrer post resection infundibular in patientes con tetralogia de Fallot. Un aneurysma del camera infundibular se disveloppava in un del pacientes post le elimination del stenosis infundibular. Un fluxo ab le sinistra verso le dextera se disveloppava al nivello ventricular post le operation in un secunde patiente qui habeva un marcate allargamento cardiac e hypertrophia sinistro-ventricular.

**REFERENCES**
