

THE SURGICAL TREATMENT OF THE TETRALOGY OF FALLOT

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Maldevelopment of the bulbus cordis may be associated with right ventricular outflow stenosis and a ventricular septal defect. The pathological anatomy in this group of anomalies varies from severe obstruction to the right ventricular outflow, with a small ventricular communication, to a large ventricular septal defect dominating the picture with mild obstruction.

When the pulmonary stenosis is mild, whether the ventricular septal defect is small or large, a left-to-right shunt is always present, giving the clinical picture of a ventricular septal defect. On the other hand, if the stenosis is severe and the ventricular septal defect is small, anatomically or functionally (McCord, Van Elk, and Blount, 1958; Vogelpoel and Schrire, 1960a; Hoffman, Rudolph, Nadas, and Gross, 1960), the clinical picture is that of severe pulmonary stenosis with an intact ventricular septum.

When the septal defect is large and the stenosis severe, a haemodynamic spectrum develops, dependent on whether the pulmonary stenosis or the systemic resistance is the greater. If the systemic resistance is greater, a dominant left-to-right shunt is present, i.e., ventricular septal defect with pulmonary stenosis; if the pulmonary resistance is greater, a right-to-left shunt is present, i.e., Fallot's tetralogy; if the resistances are balanced, acyanotic tetralogy is present.

The differentiation of ventricular septal defect with severe pulmonary stenosis and Fallot's tetralogy thus becomes a matter of semantics. Moreover, the haemodynamic state is not static. An infant may present with the dynamics of ventricular septal defect with a large left-to-right shunt. As the crista supraventricularis hypertrophies, progressive infundibular narrowing develops, so that the left-to-right shunt diminishes, the heart becomes smaller, right ventricular hypertrophy develops and, finally, a right-to-left shunt

is established, viz., Fallot's tetralogy (Gasul, Dillon, Vrla, and Hait, 1957; Fyler, Rudolph, Wittenborg, and Nadas, 1958; Lynfield, Gasul, Arcilla, and Luan, 1961; Becu, Ikkos, Ljungqvist, and Rudhe, 1961).

In this paper, by Fallot's tetralogy we mean severe right ventricular outflow stenosis, i.e., stenosis of the infundibulum of the right ventricle, pulmonary valve area, or pulmonary arteries, with a large ventricular septal defect, and right and left ventricular pressures of the same order. Even in acyanotic patients, the stenosis is relatively severe, permitting bidirectional ventricular shunt or, at most, a small left-to-right shunt at rest (<30%).

PATHOLOGY

By the time the heart is examined at operation, the pathological features observed are not only the result of the primary maldevelopment but are also consequent on changes secondary to the abnormal haemodynamics. The important defects, from the surgical point of view, are the right ventricular outflow stenosis and the incomplete development of the ventricular septum.

RIGHT VENTRICULAR OUTFLOW STENOSIS.—The normal development of the infundibulum depends on the inclusion of the bulbus cordis into the embryonic right ventricle (Peacock, 1866; Brock, 1957). The dilating force of normal blood flow further develops this part of the heart. It follows that when the bulbus cordis is maldeveloped, the surgeon encounters at operation not only the changes due to embryonic derangement but also those acquired as a result of diminished and turbulent blood flow through the abnormal channels.

Since the bulbus cordis is involved in the formation of the outflow tract of the right ventricle, from the infundibular ostium to the pulmonary valves, it is not surprising that stenosis

is found anywhere between these two points, following its malformation (Brock, 1957). The changes which result from interference with normal blood flow from the sinus of the right ventricle depend on the severity and extent of the obstruction. As in any hollow muscular system, these changes may occur proximal to the obstruction, at the site of the obstruction, or distal to the obstruction.

Proximal to the Obstruction.—Work hypertrophy of the muscle of the sinus of the right ventricle (inflow tract) occurs. This process may include the crista supraventricularis and its parietal and septal bands, resulting in further narrowing of the ostium of the infundibulum.

The persistence of the septal defect and the dextroposition of the aorta may also be secondary to this obstruction and not primary. The right ventricle pumps its blood along the line of least resistance, viz., through the opening in the unformed ventricular septum, into the aorta. The septal defect therefore persists and the aorta enlarges (Hunter, 1812; Meckel, 1827).

At the Site of the Obstruction.—Turbulent blood flow through a narrowed zone results in deposition of fibrin and a gradual silting up of the obstructed area. Gradually increasing stenosis results.

Distal to the Obstruction.—When the stenosis is localized and flow distal to it is unobstructed, blood will be ejected through the narrowed area at high velocity, causing post-stenotic dilatation. When the stenosis is tubular or the flow distal to it is obstructed, the velocity of flow is reduced; there is no dilating force, so the outflow tract distal to the obstruction does not develop normally. This is of fundamental importance and probably accounts for the great variability in the anatomical findings distal to the stenosis. In some patients removal of the stenosis alone relieves the obstruction sufficiently. In others, hypoplasia of the valve ring and main pulmonary artery acts as a new area of stenosis, once the primary obstruction has been removed. The pulmonary arteries are derived from the sixth aortic arch, not from the truncus, and do not usually share in the general hypoplasia of the outflow tract. In rare cases, the whole pulmonary arterial tree fails to develop; obstruction is then generalized.

VENTRICULAR SEPTAL DEFECT.—The defect is always large in Fallot's tetralogy and is generally situated beneath the aortic valve, behind the supraventricular crest, involving the posterior part of the membranous septum (Fig. 1). It is oval, bordered antero-superiorly by the aortic ring and postero-inferiorly by a muscular margin,

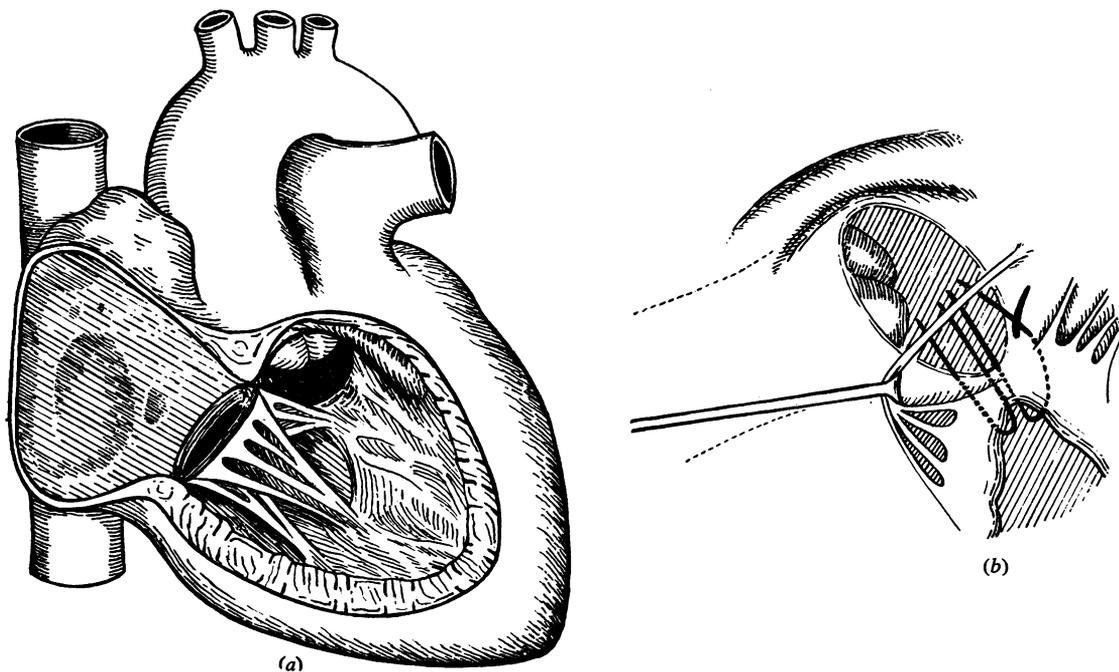


FIG. 1.—(a) Diagrammatic illustration of the anatomy of a defect in the posterior part of the membranous ventricular septum. (b) Method of insertion of silk sutures to avoid the production of heart block.

which is the upper border of the muscular septum. Cephalad, these two borders are joined by the crista, which may overhang them in part, and inferiorly the oval is bordered by the atrioventricular ring, between the septal and anterior leaflets of the tricuspid valve.

The relation of the atrioventricular bundle to this defect is important. At the postero-inferior angle, the bundle divides into right and left branches; the right branch continues on the right of the septum while the left swings around on the left ventricular side of the septum. The proximity of the conducting system to the postero-inferior angle of the defect must be carefully remembered during surgical closure.

Rarely, the defect is in the anterior part of the septum (Warden, DeWall, Cohen, Varco, and Lillehei, 1957; Senning, 1959). The opening extends just beneath the pulmonary valve cusp, well back, replacing the crista entirely. The pulmonary and aortic valve leaflets are separated

by a narrow band, and closure of the defect may distort the aortic annulus, resulting in aortic incompetence.

MATERIAL AND METHODS

Forty-four patients with Fallot's tetralogy were submitted to open heart surgery with extracorporeal circulation, using the bubble oxygenator. In 42, complete correction was attempted and in two the ventricular septal defect was not closed. All patients were thoroughly investigated and assessed pre-operatively in the Cardiac Clinic. The pre-operative diagnosis was established by cardiac catheterization, or angiocardiology, using methods previously described (Vogel, Schrire, Nellen, and Goetz, 1957), and more recently with dye dilution techniques and biplane angiocardiology. By definition, only patients with severe right ventricular outflow stenosis without large ventricular septal defects and right and left ventricular pressures of the same order were

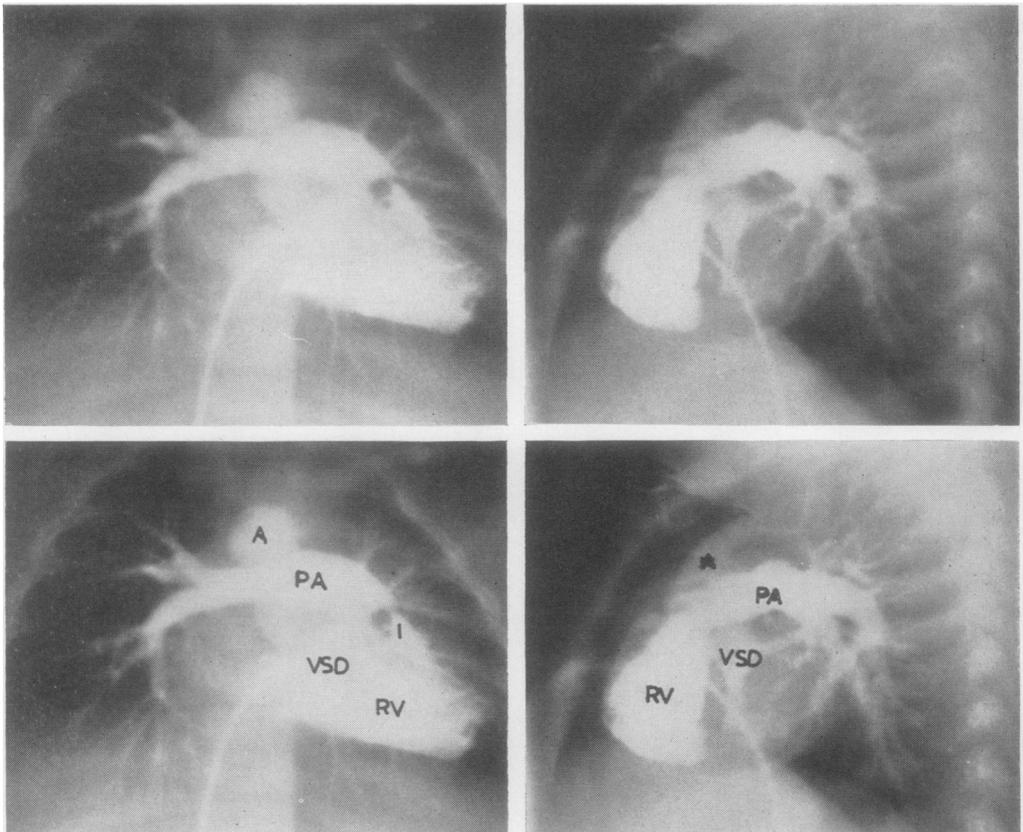


FIG. 2.—Selective angiogram from the right ventricle shows the stenosis mainly at valve level. The infundibulum is outlined. The ventricular septal defect is clearly shown with simultaneous filling of the aorta and pulmonary artery. The main pulmonary artery and its branches are well developed.

accepted for study. In seven acyanotic patients, the stenosis was always severe, permitting bidirectional ventricular shunt, or, at the most, a 30% left-to-right shunt.

There were 32 males and 12 females, their ages ranging from 11 months to 40 years, five being 3 years old or less and 28 between the ages of 3 and 15 years. The severity of the disability was assessed on symptoms and signs, as previously described (Vogelpoel and Schrire, 1960a, b, and c). Approximately 45% were mildly to moderately disabled (grades 1 and 2) and 45% were severely or grossly disabled (grades 3 and 4). The remainder had a few or no symptoms (Table I).

TABLE I
SERIES OF CASES OF TETRALOGY OF FALLOT

Age (yr.)	Sex	Cyanosis	Disability	Previous Surgery				
0-1 .. 2	} Male	Present	Severe 21	6				
2-5 .. 9					32	Moderate 20		
6-10 .. 14							Female	Absent 7
11-15 .. 8								
16-40 .. 11								

Total series 44

Cardiac catheterization is useful in establishing the diagnosis, but the precise localization of the obstruction is notoriously difficult, especially when high infundibular stenosis is present (Brock and Campbell, 1950; Neufeld, DuShane, and Edwards, 1961). Moreover, when there is diffuse narrowing and hypoplasia, or multiple stenoses, this is often not shown by catheterization. Dye dilution studies are of help in estimating the magnitude, direction, and site of the shunts. A good, selective angiocardioqram provides the most valuable information, since it outlines the detailed anatomy in each case (Fig. 2). Even with the heart displayed by open heart surgery, cardiac standstill and the ventriculotomy so disturb the functional pathology that it is not always easy to be sure of the exact anatomical deformity.

The pre-operative and operative findings were in disagreement in seven subjects. The dominant stenosis was at valvular level in 14 cases (32%), at infundibular level in 15 cases (34%), and at both levels in 15 cases (34%). Brock's figures were 35% with valve stenosis, 43% with infundibular stenosis, and 22% with combined infundibular and valve stenosis (Brock, 1957).

SURGICAL TECHNIQUE

The success of open intracardiac surgery depends, to a great extent, on whether the technique used allows the accurate, unhurried correction of the lesion, and also on whether the

procedure can be completed with the minimum of myocardial damage and without any significant change in the internal environment of the patient. This applies particularly to the complete correction of the tetralogy of Fallot. In our hands, this operation has been time-consuming, requiring on an average 100 minutes of total body perfusion and between 30 and 40 minutes of cardiac asystole (see Table II).

In the first 10 patients, normothermic high flow perfusions were used (2.4 litres per square metre per minute for patients with a body surface of up to 1 square metre, and for those with a greater surface area 2.1 litres per square metre per minute). In nine patients the heart was stopped by means of potassium arrest and in one with anoxic arrest (Table II).

TABLE II
OPERATIONS FOR TETRALOGY OF FALLOT

<i>Perfusion (average duration, 100 min.)</i>		
Normothermic	10
Hypothermic	34
<i>Cardiac Arrest</i>		
Anoxic	1
Potassium	9
Hypothermic	34
<i>Repair of Ventricular Septal Defect</i>		
Direct suture	17
Plastic patch	25
<i>Complete Correction (42)</i>		
Relief of stenosis	10
Infundibular resection	3
Valvotomy	4
Resection + valvotomy	3
Relief of stenosis + R.V. outflow reconstruction	32
To valve ring	13
Across valve ring	10
To bifurcation	9
<i>First-stage Correction (2)</i>		
Valvotomy	1
Stenosis relieved and reconstruction across valve ring	1

Although the above results were satisfactory (two deaths), the large bronchial return in the very cyanosed patients made accurate surgery difficult, especially the reconstruction of the main pulmonary artery. In addition, reports on the dangers of anoxic and potassium arrest (Gott, Bartlett, Johnson, Long, and Lillehei, 1960) and on the safety of hypothermic arrest persuaded us that generalized hypothermia, hypothermic arrest of the myocardium, and a low flow perfusion, using the pump oxygenator, would give better conditions. In the last 34 cases this technique was used (Barnard, Terblanche, and Ozinsky, 1961).

Except for the first patient, a median sternotomy was always used. The thymus was dissected free from the root of the pericardium, which was opened vertically in the midline from the dome of the diaphragm to the root of the great vessels, exposing the ascending aorta, main pulmonary

artery and its two branches. The exterior of the heart and its main vessels were inspected, special attention being directed to the main pulmonary artery and its branches.

At this stage, the surgeon should decide whether a two-stage operation or the complete correction is to be performed. Where the reconstruction necessitates the insertion of a prosthesis up to the bifurcation of the main pulmonary artery, the pulmonary artery and the commencement of its branches are dissected free from the root of the aorta. If the underdevelopment of the right ventricular outflow extends beyond the bifurcation of the pulmonary artery, complete correction is followed by right ventricular hypertension and usually the patient dies.

The right ventriculotomy was placed in the right ventricular outflow, without dividing the fibres of the sinus of this chamber and the major coronary vessels. By doing this, the efficiency of right ventricular systole is not as likely to be disturbed by the cardiomy. On opening the right ventricle, the anatomy of the various defects was studied.

The first part of the correction consisted of relieving the obstruction. Infundibular stenosis was relieved by resecting the two limbs of the crista, care being taken not to damage the aortic valve or the septal arteries. In the 42 patients in whom complete correction was attempted, pure infundibular stenosis, requiring resection only, was encountered in three patients, two of whom were acyanotic and one of whom had had a Brock procedure six years before (Brock and Campbell, 1950). Pure valvular stenosis was best corrected by exposing the fused leaflets through a small pulmonary arteriotomy. The commissures could be clearly seen and accurate incision of the fused commissures prevented gross incompetence. This procedure relieved the obstruction adequately in three patients. Pulmonary valvotomy plus infundibular resection only was performed in a further four patients (Table II).

In the remaining 32 patients there was either underdevelopment of the infundibulum, pulmonary valve ring, main pulmonary artery or its branches, necessitating reconstruction of the underdeveloped area in order to complete the relief of the obstruction. This was achieved by the insertion of a plastic roof. In four patients we used a curved Ivalon prosthesis and in the remaining 29 a woven Teflon prosthesis. When reconstruction of the pulmonary valve ring and/or the main pulmonary artery was necessary, the cardiomy was extended across the valve ring

into the pulmonary artery and, where required, across the bifurcation.

In 13 patients the prosthesis extended up to the valve ring, in 10 across the valve ring into the pulmonary artery, and in nine the patch reached the bifurcation of the main pulmonary artery (Table II).

The prosthesis was partially inserted up to the valve ring before closing the ventricular septal defect. Aspiration of blood in the left heart, through the open ventricular septal defect, prevented the return of bronchial circulation backward through the pulmonary artery, and therefore a dry field could be better maintained.

CLOSURE OF THE VENTRICULAR SEPTAL DEFECT.

—Since the ventricular septal defect in the tetralogy of Fallot is always large, complete closure without producing complete heart block poses a problem. In the 42 patients in whom the defect was closed, this was achieved either by direct suture (17 patients) or by using a prosthesis sutured in position (25 patients). Before beginning the closure, the anatomy must be studied, particularly the relation of the defect to the aortic valve cusps and the tricuspid annulus. In the first 10 patients, sutures were placed through the full thickness of the margins of the defect, and complete heart block resulted in two. In the last 32 patients, the technique described by Kirklin, McGoon, and DuShane (1960), to avoid damaging the conducting bundle, was used with only one failure. When a prosthesis was required, a compressed Ivalon patch was used.

The ventricular septal defect was in the anterior part of the septum in three patients. In this type of defect, the slightest disturbance in the aortic annulus resulted in aortic incompetence. These defects were best closed with a prosthesis, as closure by direct suture was more likely to cause leaking of the valve.

Associated lesions, such as atrial septal defect or patent ductus arteriosus, could be identified and repaired at the same operation.

If the surgeon decided that he could not relieve the right ventricular outflow obstruction, a limited valvotomy was performed to relieve the stenosis and permit a mild left-to-right shunt. This was done in two patients, one of whom required plastic reconstruction of the narrow pulmonary valve ring, which was the main obstruction.

After discontinuing bypass, the venous pressure was used as a guide to the replacement of blood. The patients were usually perfused until the venous pressure was raised and a good systemic

pressure obtained. All patients were routinely digitalized after the bypass was discontinued.

The pericardial incision was tightly closed with a continuous suture and a drain was left in the pericardial cavity to drain any blood and prevent tamponade. This was removed 48 hours after surgery. By closing the pericardial cavity, the outflow patch is protected against infection. The pericardium becomes adherent over this area, strengthening the graft and minimizing the danger of aneurysmal dilatation at a later date.

The clinical picture of tubular necrosis was met in two patients, giving rise to extra anxiety in the immediate post-operative period. Treatment with restriction of fluids, resins and attention to diet, along conventional lines, promoted recovery. Sludging of the viscous polycythaemic blood at low temperatures, with low perfusion rates, was

probably responsible. (Since this observation, in two cases, as soon as bypass was begun a portion of the patient's blood was removed and replaced with donor blood with a normal haematocrit, thus reducing the viscosity. This proved to be satisfactory.)

RESULTS

Clinical examination during the post-operative period showed that all patients developed a raised jugular venous pressure and hepatomegaly a few days after surgery, and this usually persisted for several weeks. Digitalis was routinely given in full doses post-operatively, starting with an intravenous dose while the patient was still on the operating table.

Full examination was repeated three or more weeks after surgery, at the time of discharge from

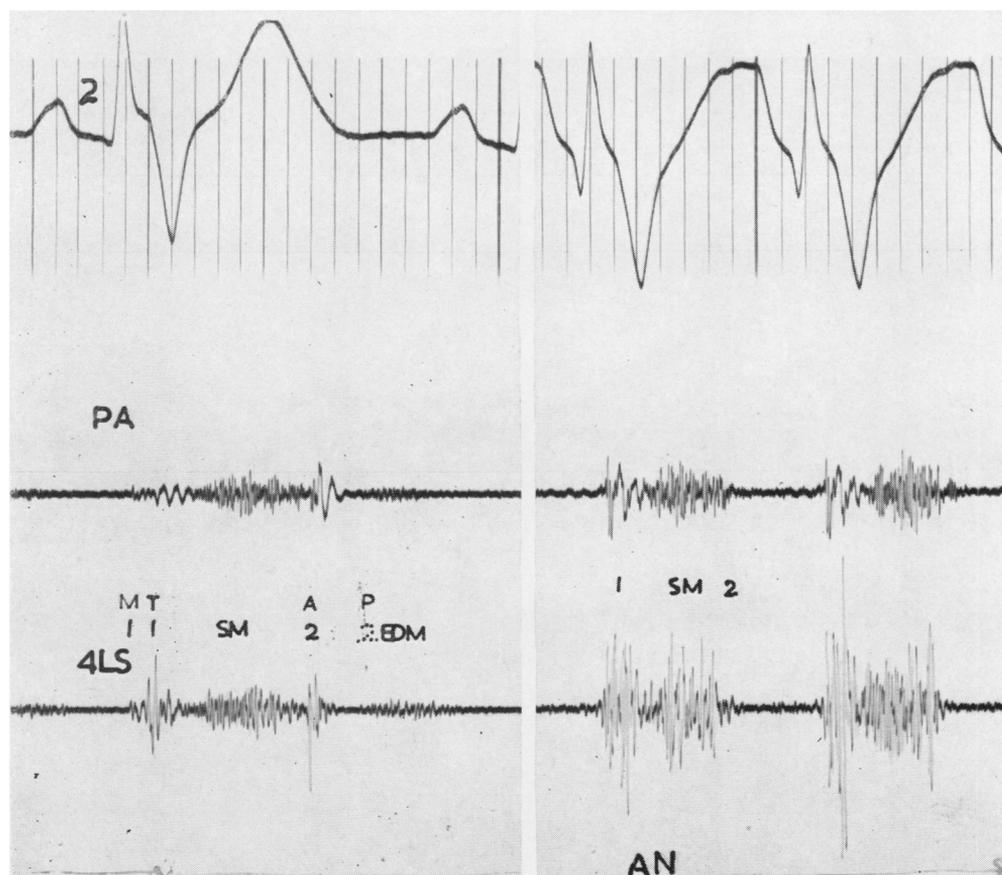


FIG. 3.—Post-operative phonocardiogram (one year after complete repair) shows the right bundle branch block associated with wide splitting of both heart sounds. The systolic murmur is ejection in type, situated between the tricuspid sound (T1) and the pulmonary sound (P2). Because of the ventricular asynchrony, the murmur continues right up to the aortic sound (A2). The delayed pulmonary diastolic murmur identifies the pulmonary valve closure (P2). After amyl nitrite inhalation the systolic murmur promptly intensifies, indicating that the ventricular septal defect is closed; the first sound intensifies, the second softens and the diastolic murmur disappears.

TABLE III
RESULTS IN TETRALOGY OF FALLOT

Correction		Survived	Mortality (%)
Complete	.. 42	35	17
First stage	.. 2	1	50
Total	.. 44	36	19

hospital. On auscultation and phonocardiography (Vogelpeol and Schrire, 1960), a systolic murmur was always present. A delayed pulmonary early diastolic murmur (Fig. 3) was heard in 29 of the 36 survivors. Right bundle branch block was nearly always present when the ventricular septal defect was in the usual position, so that wide splitting of the second heart sound was found (Fig. 3). The murmur continued up to, or beyond, A2, so it was not always easy to decide whether the residual murmur was due to a persistent defect in the ventricular septum or to an outflow tract ejection murmur. Amyl nitrite was generally of great help in differentiation (Vogelpeol and Schrire, 1960b). The electrocardiogram showed sinus rhythm in all but two patients, one of whom had a transient complete heart block. Right bundle branch block of a

peculiar type, associated with surgery, was generally present (Fig. 4). Radiologically, the heart was usually larger than pre-operatively, especially if pulmonary incompetence was present, and the pulmonary vasculature was increased, but plethora was absent except in the three patients with a persisting septal defect. Two of these were closed at a second operation, with subsequent disappearance of the plethora.

Most patients were examined every three to six months after surgery, except those who live hundreds of miles from the clinic. The latter were re-assessed before recatheterization, approximately one year after surgery. The improvement was often dramatic, particularly in patients severely disabled before operation, and in every patient the results of surgery were rewarding. The post-pericardiotomy syndrome was not infrequently present, especially in the older patients, but was never troublesome. Cyanosis sometimes persisted for the first few days after surgery but, except for one patient with an atrial septal defect, always disappeared, and clubbing improved.

One year or more has elapsed since surgery in the first 22 patients. Relief of the outflow

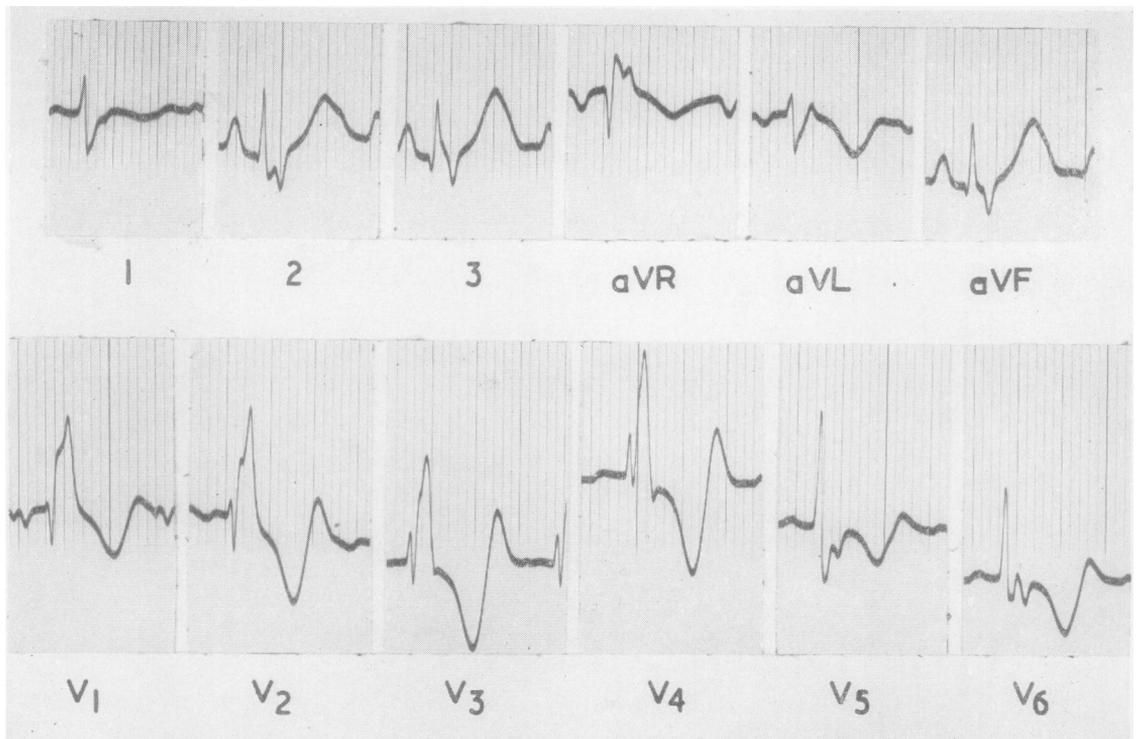


FIG. 4.—The electrocardiogram shows the typical right bundle branch block which follows repair of the ventricular septal defect.

obstruction alone was attempted in two (first stage procedure). In one it was life-saving; the other developed an aneurysm of the pulmonary artery at the site of the patch and died five months after surgery.

There were five deaths in the 20 patients in whom complete correction was attempted. Three died of unrelieved stenosis, one of whom developed complete heart block; one infant had chronic cerebral anoxia and blindness before operation, with extensive brain damage at necropsy; the remaining patient was probably inadequately perfused.

Fourteen patients have been recatheterized using dye dilution techniques as well as saturation data to detect residual shunts. Eight had virtually normal haemodynamics; three had residual shunts at ventricular level (suspected clinically), in two of whom it was trivial, and the third was a difficult operation on a patient who had already had a Blalock and a Brock procedure previously; three had moderate residual pulmonary stenosis. In one patient a bidirectional shunt at atrial level was present, through an atrial septal defect which was deliberately left open at the time of surgery.

In the remaining 22 patients, the follow-up period has been inadequate. However, on clinical grounds, the response to surgery has been excellent. A residual shunt has not been diagnosed clinically in any patient. There have been two deaths. One, a severely ill child, died of digitalis toxicity shortly after a successful operation. The other died four months after surgery in intractable congestive cardiac failure. The failure was partly due to aortic incompetence due to bicuspid valves, made incompetent during repair of the ventricular septal defect.

DISCUSSION

The complete surgical correction of Fallot's tetralogy requires removal of the obstruction to right ventricular outflow and closure of the ventricular septal defect. In pulmonary stenosis with intact ventricular septum, the right ventricle must generate sufficient pressure to overcome the stenotic resistance and maintain adequate pulmonary flow. In Fallot's tetralogy, however, the ventricular septal defect acts as an escape route preventing the right ventricular pressure from rising above systemic, and protecting the right ventricle from the severe resistance to outflow. If the defect is closed (producing pulmonary stenosis with intact ventricular septum), the right ventricular outflow obstruction must be adequately relieved if the patient is to survive. This aspect

must always receive consideration before closure of the defect is attempted.

RELIEF OF RIGHT VENTRICULAR OUTFLOW STENOSIS.—Right ventricular outflow obstruction is dependent on the primary developmental malformation and the acquired changes secondary to the reduced flow, as already described. The surgeon may be called upon to deal with either one or both situations (see Table IV).

TABLE IV
PATHOLOGY OF TETRALOGY OF FALLOT

Site of Ventricular Septal Defect	Right Ventricular Outflow Stenosis	
	Normal Distal Development	Abnormal Distal Development
Posterior .. 41	Infundibular .. 3	Infundibular .. 13
Anterior .. 3	Valvular .. 4	Infundibular plus valve area .. 11
	Infundibular plus valvular .. 4	Infundibular plus valve area plus P.A. .. 9
	11	33

Stenosis with Adequate Distal Development.—Once the stenosis has been removed, the right ventricle can cope adequately without a significant rise in pressure. This situation is met where the stenosis is not extreme, so that a bidirectional shunt—or a small left-to-right shunt only—is present. Of the 44 patients, 11 were in this group, three requiring only infundibular resection, four requiring pulmonary valvotomy, and four requiring a combination of these two procedures.

Sufficient blood flows from the right ventricle into the pulmonary circuit to develop the outflow tract. Where isolated infundibular or valve stenosis occurs, post-stenotic dilatation permits a capacious chamber to develop distal to the obstruction. In this group, the surgical correction of the stenosis offers little difficulty.

Stenosis with Inadequate Distal Development.—Free flow of blood from the right ventricle is dependent on adequately developed channels from the ostium of the infundibulum to the pulmonary capillaries. In this type of patient, a segment of the outflow tract is underdeveloped or, rarely, the whole circuit from the ostium proximally to the small pulmonary arteries distally is hypoplastic. The pulmonary valves are frequently malformed, fused or even absent. There were 33 patients in this group.

For practical purposes, five groups can be differentiated: (1) Infundibular stenosis and underdevelopment of the infundibular chamber, up to the pulmonary valve ring (13 patients); (2) infundibular stenosis, underdevelopment of the

infundibular chamber and the pulmonary valve area (11 patients); in the pulmonary valve area, the valve alone may be stenosed, the valve and valve ring may be narrowed, or the valve ring alone may be affected; (3) infundibular stenosis, underdevelopment of the infundibular chamber, pulmonary valve area and main pulmonary artery up to the bifurcation (seven patients); the right and left pulmonary arteries are often surprisingly well proportioned; (4) infundibular stenosis, underdevelopment of the infundibular chamber, pulmonary valve area, main pulmonary artery, and distal pulmonary arterial branches (two patients); (5) absence of a pulmonary artery (one patient) or stenosis in association with (1) to (4).

The surgical management of pulmonary outflow tract stenosis with inadequate distal development is controversial. Brock (1959) suggested that a two-stage operation was the treatment of choice. The first operation was designed to relieve the stenosis partially, so that a bidirectional or a small left-to-right shunt results. The ventricular septal defect was not closed at this stage, but remained as a safety valve. It is hoped that, after this operation, the increased blood flow to the lungs will develop the outflow tract to such an extent that septal closure can be achieved at a later date, without resorting to plastic reconstruction. We have attempted this in two desperately ill infants with grade 4 disability. In the first, surgery was life-saving; the cyanotic attacks were relieved and further progress has been satisfactory. In the second, marked distal pulmonary arterial hypoplasia was present, the patient developed a pulmonary arterial aneurysm at the site of the insertion of the prosthesis and died suddenly five months after surgery.

Lillehei, Cohen, Warden, and Varco (1955), Kirklin, Ellis, McGoon, DuShane, and Swan (1959), and many others preferred a single operation. The stenosis was relieved and the underdeveloped outflow tract was reconstructed by the insertion of a plastic or pericardial roof. In some cases this necessitated the extension of the roof from the infundibulum across the valve ring into the main pulmonary artery (one-third of our patients). This may have to be extended right into the bifurcation (one-quarter of our patients). Once the obstruction had been relieved, the ventricular septal defect could be closed.

We feel that there is a place for both procedures in the treatment of severe tetralogy. In the majority of patients, a single procedure can be performed with a reasonable mortality. As far as is known to date, plastic reconstruction and pul-

monary incompetence do not lead to serious complications later in life. It must be accepted that chronic overwork inevitably throws a strain on the heart. The right ventricle is ideally suited to eject large volumes of blood at a low outflow pressure (Rushmer and Thal, 1951). Moreover, complete destruction of the free wall of the right ventricle produces no immediate effect on the circulation (Starr, Jeffers, and Meade, 1943; Bakos, 1950; Kagan, 1952). Experimental excision of the pulmonary valves in dogs (Barger, Roe, and Richardson, 1952; Ratcliffe, Hurt, Belmonte, and Gerbode, 1957; Fowler and Duchesne, 1958) produced remarkably little alteration in function but cannot be regarded as benign. In man, foetal congestive cardiac failure due to pulmonary incompetence has been described (Smith, DuShane, and Edwards, 1959). However, in most patients pulmonary incompetence has been compatible with a long, relatively symptom-free survival (Olesen and Fabricius, 1956; Price, 1961; Schire, 1961). The effect of surgically produced pulmonary incompetence still remains to be determined in the future.

It has been our policy to do a one-stage correction in groups 1 to 3. In group 4 the right ventricular pressure cannot be sufficiently reduced, even after reconstruction of the outflow tract right up to the bifurcation of the main pulmonary artery, to permit safe closure of the ventricular septal defect.

CLOSURE OF THE VENTRICULAR SEPTAL DEFECT.—Although the closure of the ventricular septal defect by direct suture, without using foreign material, appears to be preferable, the muscular postero-inferior border of the large defects encountered in the tetralogy of Fallot does not hold stitches well, especially when there is tension on the sutures. In the 17 patients in whom the defect was closed by direct suture, two developed serious ventricular septal leaks after correction and required re-operation. We prefer to close the ventricular septal defect by the insertion of a compressed Ivalon patch, circumferentially sutured in place with 3-0 silk mattress sutures. The same precautions can be taken to avoid heart block as described by Kirklin *et al.* (1960) for repair by direct suture. This was done in 25 patients, only one of whom developed a significant leak.

CONCLUSIONS

Forty-four patients with Fallot's tetralogy of varying severity were subjected to surgery with cardiac bypass.

The pre-operative diagnosis and site of stenosis was established by catheterization and/or angiocardiology and confirmed at surgery.

Complete repair of the defects was attempted in all but two patients. In 10, infundibular resection and/or pulmonary valvotomy adequately relieved the stenosis. Five of these subjects were acyanotic and two had had previous palliative procedures. The remaining 32 required plastic repair of the pulmonary outflow tract, varying from repair of the right ventricle alone to repair of the pulmonary outflow tract from the infundibular ostium to the bifurcation of the main pulmonary artery.

The ventricular septal defect was closed in 42 patients either by direct suture (17) or by means of a plastic patch (25). The importance of adequately relieving the obstruction to the pulmonary outflow tract before closing the defect is emphasized. Failure to relieve the stenosis resulted in half the deaths.

Of the two patients in whom no attempt was made to close the ventricular septal defect, one responded excellently and the other died with an aneurysm of the pulmonary artery five months after surgery.

There were seven deaths in the 42 patients in whom complete repair had been attempted. Three died of unrelieved stenosis. This includes one of the two patients in the series with established complete heart block following surgery.

The response to surgery has been gratifying in the surviving patients. Symptoms and cyanosis have disappeared and effort tolerance has been restored to normal. Three patients had persistent large ventricular septal defects, two of which were repaired at a second operation. Recatheterization of the first 14 survivors has shown almost complete restoration of normal haemodynamics, or small insignificant shunts or gradients, in the majority.

From the surgical point of view, patients with Fallot's tetralogy can be divided into two main groups: (a) Those patients with adequate outflow tract development distal to the stenosis: these can be relieved by infundibular resection and/or pulmonary valvotomy. (b) Those patients with inadequate outflow tract development: these require infundibular resection and plastic repair, the extent of which varies with the length of the hypoplasia. All require a plastic roof in the right ventricle; in some it extends to the pulmonary valve, in others across the pulmonary valve and, where the hypoplasia is extreme, the patch must extend right across the valve up to the bifurcation of the main pulmonary artery. In a few exceptional cases, hypoplasia

extends into the small pulmonary arteries and the obstruction cannot be relieved by plastic repair. A "two-stage" procedure is recommended for this type of defect.

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