Direct Vision Intracardiac Surgical Correction of the Tetralogy of Fallot, Pentalogy of Fallot, and Pulmonary Atresia Defects

Report of First Ten Cases*

C. WALTON LILLEHEI, M.D., MORLEY COHEN, M.D., HERBERT E. WARDEN, M.D., RAYMOND C. READ, M.D., JOSEPH B. AUST, M.D., RICHARD A. DEWALL, M.D., AND RICHARD L. VARCO, M.D.

Minneapolis, Minn.

From the Department of Surgery, University of Minnesota Medical School, Minneapolis

CLEARLY THE SURGEON'S ultimate goal in treating the tetralogy of Fallot type of congenital heart defects should be to restore the circulation to normal. This is accomplished by both closing the ventricular septal defect and removing the pulmonary stenosis, whether valvular or infundibular.

The advent of controlled cross circulation[1-4] as a successful method for direct vision intracardiac surgery has made such corrective surgery possible. Since our initial experience in 1954 with the curative procedure for the tetralogy defects, we have come to adopt that plan for all patients with this lesion currently in need of surgical treatment.

METHOD

Controlled cross circulation for achieving total by-pass of the heart and lungs of the patient has been used in all of the cases herein reported.† The basic technical features dealing with the application of this method to direct vision intracardiac surgery in man have been described elsewhere.[1-4] Several specific considerations, pertinent to open intracardiac surgery utilizing this perfusion method, merit another brief mention here because of their significance.

1. *Systemic Perfusion at Reduced Rates of Flow.* Through application of the low flow concept to this perfusion technic important simplifications in method became possible. These advantages have contributed significantly to achieving the successes. Based upon the azygos flow experimental studies[5, 6] we have perfused these patients during their total by-pass interval at a rate of blood flow approximately 1/6 to 1/4 of the resting cardiac outputs for normal individuals of comparable size and weight. These flow volumes have varied from 23 ml. to 29 ml. per kilogram of body weight per minute. For these ten patients so managed, the heart and lungs were totally by-passed from six to 21½ minutes. This group of patients, together with 47 additional individuals with other types of intracardiac lesions, have had total cardiac

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† Since the completion of this series of ten patients herein reported who have been consecutively operated upon by the cross circulation method of by-pass, one additional patient with the tetralogy of Fallot defect, J. W., age 22 months, U.H. #883-416, seriously ill as manifested by daily anoxic convulsive seizures and a preoperative femoral artery oxygen saturation of 44 per cent, has had his intracardiac defects successfully corrected during an open cardiotomy, lasting 17 minutes, utilizing a simple inexpensive disposable artificial oxygenator assembled from materials available in the average hospital supply room. The same cross circulation type pump was used with this oxygenator. The design and use of this oxygenator is described elsewhere.[9]

TABLE I. *Tetralogy of Fallot.*

Anatomy of the Right Ventricular Outflow Tract in Ten Patients Managed Consecutively by Intracardiac Correction		
Infundibular Stenosis		
Complete Atresia	1	8
High (A poorly developed or no infundibular chamber)	6	
Low (Well developed infundibular chamber)	1	
Valvular Stenosis		2*

*One patient had associated multiple (3) atrial defects.

and pulmonary by-pass for corrective intracardiac surgery at comparable rates of perfusion and at normal body temperature without a single instance of cerebral, hepatic, or renal dysfunction attributable to these low flow rates.

2. *Control of Body Temperature.* To avoid the increased myocardial susceptibility to the arrhythmias often associated with hypothermia, we have attempted to carry out all of these intraventricular procedures at normal body temperatures.

In our air conditioned operating rooms the small children and infants, particularly, when under anesthesia and after a bilateral thoracotomy, often undergo a rapid spontaneous fall in body temperature. We have been able to prevent this for the most part by placing an electric heating blanket beneath all of these patients on the operating table. Also, the patient's thermal status during the operative procedure is revealed by a temperature recording catheter inserted into the rectum. This instrument quickly provides an accurate reading on its dial for the monitoring anesthesiologist. Moreover, all the bank blood to be transfused into these patients is warmed to 38° C. in a water bath kept in the operating suite.

TABLE II. *Tetralogy of Fallot, Pentalogy of Fallot, and Pulmonary Atresia Defects.*
Intracardiac Corrective Operations*

Case No. and (Sex)	Age (yrs) and Wt. (Kg.)	Indications for Surgery	Diagnosis at Operation	Perfusion Rate ml/Kg./min.	Duration of Total By-Pass (min)	Femoral Artery % Oxygen Saturation		Results
						Preop.	Postop.	
1. M.S. (M)	11 (26.4)	Dyspnea, physical incapacity	I.V.S.D. with Infund. Stenosis	28	11½	83	96	Excellent
2. J.A.† (F)	14 (38.6)	Dyspnea, physical incapacity	"	25	9	83	—	Died, 1 hour
3. J.W. (M)	1½ (7.7)	Syncope, coma, dyspnea	"	26	6	22	—	Died, 12 hours
4. W.B. (M)	19 mo. (8.5)	Cardiac failure, terminal	"	23	9½	25	94	Excellent
5. G.F. (F)	2½ (9.3)	Dyspnea, syncope	I.V.S.D. with Valvular stenosis	25	15½	69	—	Died, 2 hours
6. L.K. (F)	22 mo. (10)	Dyspnea, physical incapacity, intense cyanosis	I.V.S.D. with pulmonary atresia	27	20	39	95	Excellent
7. J.S. (M)	3½ (13)	Dyspnea, incapacity, polycythemia	I.V.S.D. with Infund. Stenosis	27	12	41	—	Died, 11 hours
8. P.R. (F)	18 mo. (9.5)	Dyspnea, convulsions	" /	29	16	35	95	Excellent
9. S.C. (F)	7 (16.8)	Dyspnea, incapacity, polycythemia	"	25	16½	53	94	Excellent
10. D.S. (M)	10½ (26)	Dyspnea, incapacity, arrhythmia	I.V.S.D., I.A.S.D. with valvular stenosis	25	21½	75	91	Excellent

*Closure of the ventricular septal defects and relief of the pulmonary stenosis by infundibular resection or valvulotomy performed under direct vision through a right ventricular cardiotomy. One patient (Case 10) had in addition to the ventricular cardiotomy a simultaneous auriculotomy for repair of multiple (3) atrial septal defects.

†Had a subclavian-pulmonary artery anastomosis done two years previously, at which time the pericardium was opened for exploration.

I.V.S.D.—Interventricular septal defect.

I.A.S.D.—Interatrial septal defects.

TABLE III. *Intracardiac Correction of the Tetralogy of Fallot Defects.*

Cardiac Catheterization Data Before and After

Patient	Age at Surgery	Time Relationship of Catheterization to Surgery	Oxygen Saturations* Percent						Pressures (mm Hg)			
			Sup. Cava	Inf. Cava	R. At.	R. Vent.	Pul. Art.	Systemic Artery	R.A.	R.V.	P.A.	Systemic Artery
Case 1. M.S.	11 yrs.	2 yrs. before	45	—	53	44	46	83	—	93/0	8/4	90/70
		7 mo. after	66	71	66	Lo 65 Hi 71	74	96	8/5	85/0	30/8 (M-18)	110/70
Case 4. W.B.	19 mo.	1 mo. before	19	—	24	39	—	25	7/2	82/0	8/5	80/40
		6 mo. after	71	69	66	66	65	94	6/2	38/0	20/8 (M-14)	100/70

*All oxygen content determinations were made by the Van Slyke Method of Analysis.
M-mean pressure.

3. *Blood-Free Intracardiac Operating Field.* During the perfusion, despite cardiac inflow stasis by complete caval occlusion, there remain three sources which may contribute blood into the intracardiac operating field. These are: the coronary venous return; the bronchial collateral circulation between the aorta and the lungs, and thence back to the heart; and regurgitation through the aortic valve.

The utilization of a low flow rate during these perfusions has contributed to a substantial reduction in the quantity of blood returning to the heart from the first two above-mentioned sources. In all of these tetralogy cases operated upon to date, the usual location of the ventricular septal defects has been in immediate proximity to and extending up between the cusps of the aortic valve (Fig. 5). This high position has tended to render the latter valve incompetent during the manipulations incident to placing the closure stitches. To correct this annoying problem and to avoid the flooding of the operative field by refluxing blood, the origin of the ascending aorta is looped routinely in all cases by a cotton tape held in a Rumel tourniquet. This arrangement allows selective occlusion of the ascending aorta between the arterial perfusion inflow catheter and the aortic valve with the origins of the coronary arteries.

Intermittent occlusion of this tourniquet as needed then provides a virtually bloodless operating field in the case of the acyanotic types of septal defects. Though much improved over the situation without such a control, the tetralogy cases and other types of cyanotic heart disease still regularly exhibit varying quantities of pulmonary venous return *via* the bronchial collaterals. As previously emphasized,[4] this maneuver of tourniquet control of the root of the aorta, by the clearer vision afforded the surgeon, has contributed substantially to a reduction in the time needed for the necessary reparative surgery inside of the heart. Moreover, the total blood loss has been diminished through this limiting of the aortic regurgitation, as well as by lessening the coronary flow.

4. *Weighing Scale.* At the conclusion of a perfusion procedure for intracardiac surgery it is important for the patient's well-being and ultimate recovery that his blood volume be in a near normal range. Any accurate evaluation of this status has been complicated by the patient's losses from his operative wounds and elsewhere onto the drapes, thereby nullifying accurate measurements. Careful volumetric appraisals are made of the quantities of blood aspirated from the interior of his heart and chest

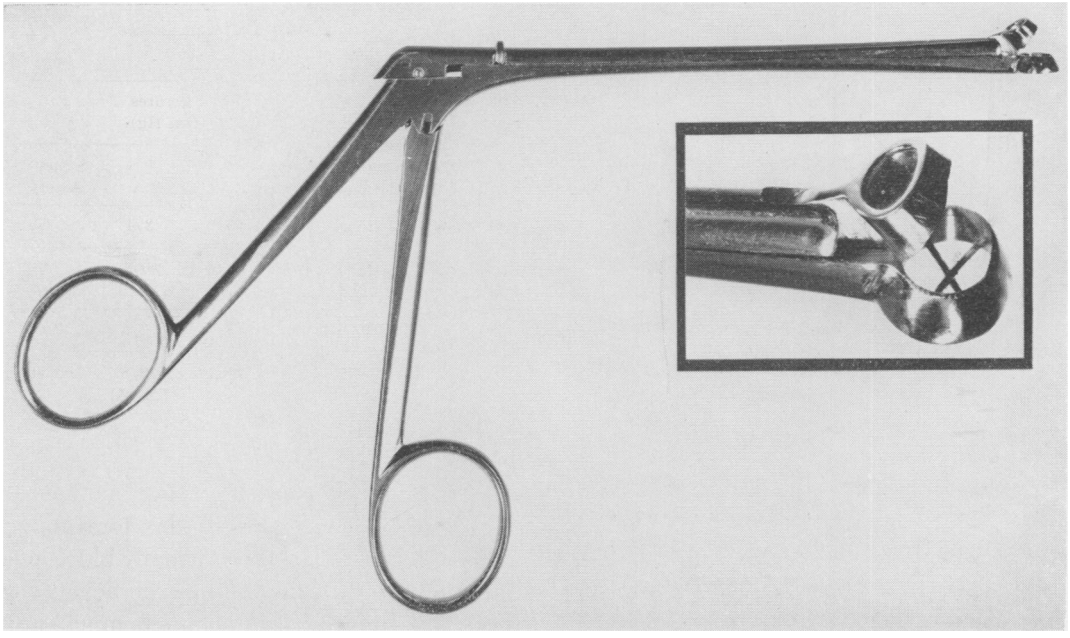

FIG. 1. Infundibulectomy scissors. For the resection of the tenacious and toughened muscle comprising the infundibular obstructions, a cutting instrument with blades that interdigitate has been found superior to a biting or rongeur type forceps. A Hartmann tonsil punch modified (inset) by the addition of a basket to retain the resected muscle pieces has met these criteria very satisfactorily.

cavities,* but at best these have proved to be only rough estimates. After an initial trial in these patients, we have discarded serial blood volume determinations because they were cumbersome, slow, and often imprecise. We believe that careful weighing of the patient in the operating room immediately preoperative and postoperative has provided a rapid, simple, and more accurate index of the blood volume changes over the short time span of the operative procedure.

SELECTION OF PATIENTS

Since 1950 in our clinic, patients diagnosed as having pulmonic stenosis and a ventricular septal defect (Fallot's tetrad) have been considered as candidates for surgical management only if there has been definite evidence of recent and significant deterioration.[7] This state may be represented either by recurrent bouts of heart failure, decompensation of the pulmonary collateral channels (episodes of dyspnea on slight effort, syncope, coma, or convulsions), or a sustained rate of increase in the hemoglobin, erythrocyte, or hematocrit values. In the past, unless one or more of these disturbing developments have appeared to a significant degree, we have withheld palliative surgery in order to allow as many of these patients as possible to live into the era when curative surgery would be possible.

To date, all ten of the patients accepted for corrective surgery have been judged by our cardiologists to be in serious difficulty, manifesting one or more of these same indications of sustained and impressive deterioration (Table II).

These ten cases have been consecutively operated upon. Since embarking upon this program of curative surgery for this combi-

* This blood has been severely hemolysed by the forces applied by an effective aspirator. Hence, we have not considered it advisable to re-transfuse this blood in order to conserve on the total amount needed.

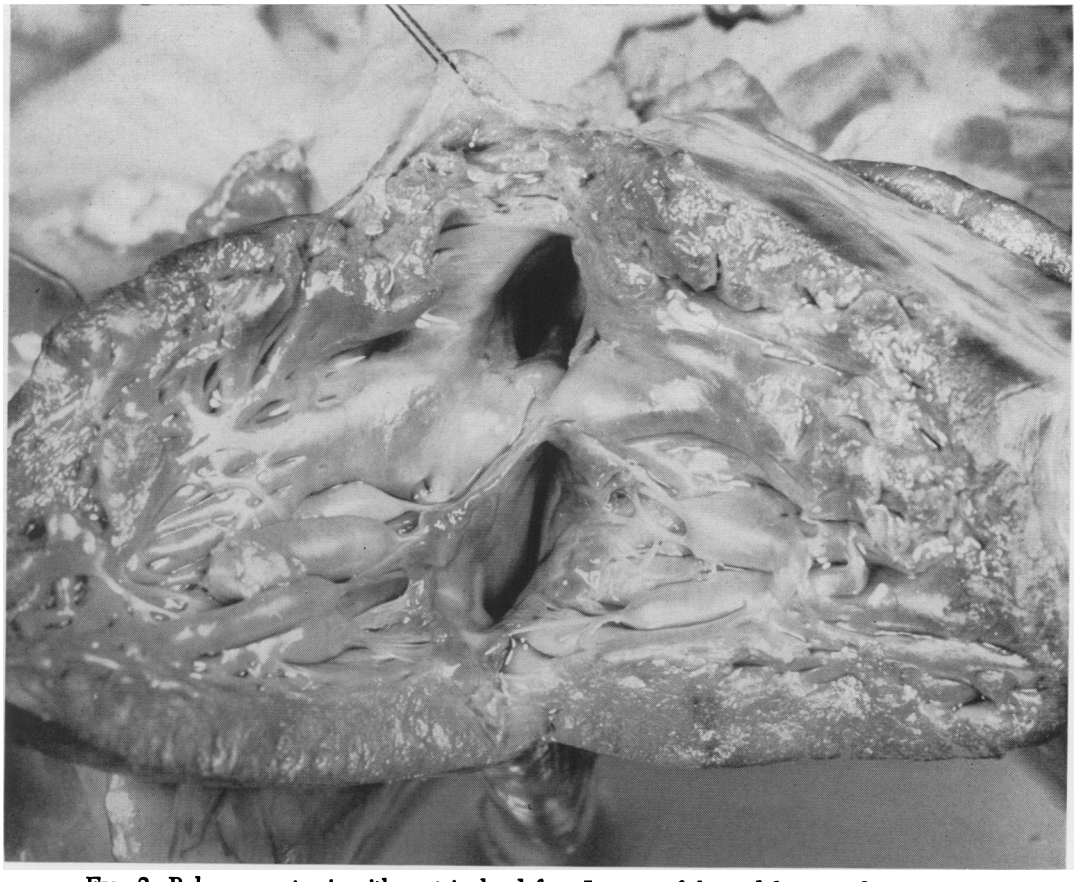

FIG. 2. Pulmonary atresia with ventricular defect. Interior of the widely opened right ventricle (J. W., age 6 months, who died of his defects) viewed from below looking upwards into the right atrium through the tricuspid valve orifice (located in the lower half of the picture); and into the aorta through the ventricular septal defect (located above the tricuspid valve in the photograph). The cusps of the aortic valve are clearly visible. The outflow tract is atretic, and is represented by the small dimple evident anterior to the ventricular defect.

nation of defects, there have been no anastomotic procedures deemed necessary even though in each case we have reserved the final decision as to the relative feasibility of either a corrective or an anastomotic procedure until after the pericardium has been opened and the external anatomy of the cardiac structures inspected.

THE INTRACARDIAC OPERATION

For several years prior to the availability of any satisfactory method for intracardiac surgery, it had been and continues to be our policy to be present at every autopsy upon a patient succumbing with a known or suspected congenital heart defect. With the heart still *in situ*, these defects have been exposed by a surgical cardiotomy, and a corrective procedure carried out. These mock operations have proved to be of incalculable value to us in undertaking these subsequent clinical procedures.

In each of these patients comprising this operative series, after completion of the preparations for total by-pass of the heart and lungs, adequate exposure to the cardiac interior for closure of the ventricular septal-defect and the relief of the pulmonic stenosis has been achieved by a vertical incision in the right ventricle, extending over most of its anterior surface and placed to avoid cut-

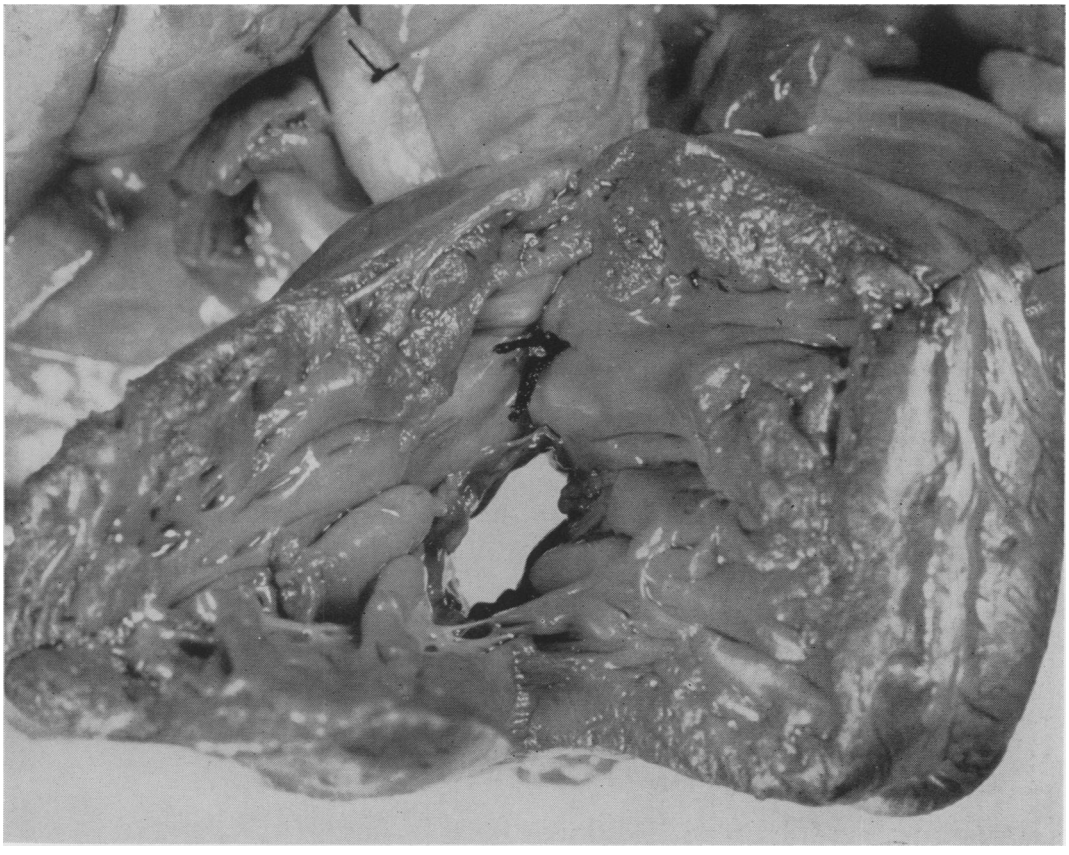

FIG. 3. Same heart as in Figure 2. The ventricular septal defect has been closed with two 000 silk U shaped stitches. Note also the typically very enlarged aorta at the top of the photograph.

ting major branches of the coronary arteries. If an infundibular obstruction was present, the ventriculotomy incision was extended directly through the stenotic area and up to the pulmonary valve. The intracardiac pathology was inspected and if necessary, papillary muscles that interfered with access to the ventricular defect were transected to be repaired later before closure of the cardiotomy.

Closure of the Ventricular Septal Defects. The septal defects have been closed by direct suture with interrupted stitches of 000 silk. The recognition of the importance of placing these closure stitches on the right ventricular aspect of the defect margins will allow complete closure of the defects without injury to the aortic valve cusps. This technic is accomplished by the use of U shaped or half purse string stitches at either end of the defect with the placement of as many Cushing type mattress stitches in between as necessary. Properly placed, this type of stitch accomplishes approximation of the top of the muscular septum (the inferior margin of the ventricular defect) firmly to the base of the aortic septum with its attached aortic cusps (which is the superior margin of the defect). Also, when tied, this type of stitch everts the defect margins toward the left ventricle, thus preserving intact the function of the aortic valve. In several patients, these closure stitches were placed in the same manner, but tied over a pledget of polyvinyl sponge (Ivalon) as portrayed in Figure 15. Study of Figures 2 to 5 will facilitate an appreciation of these technical suggestions.

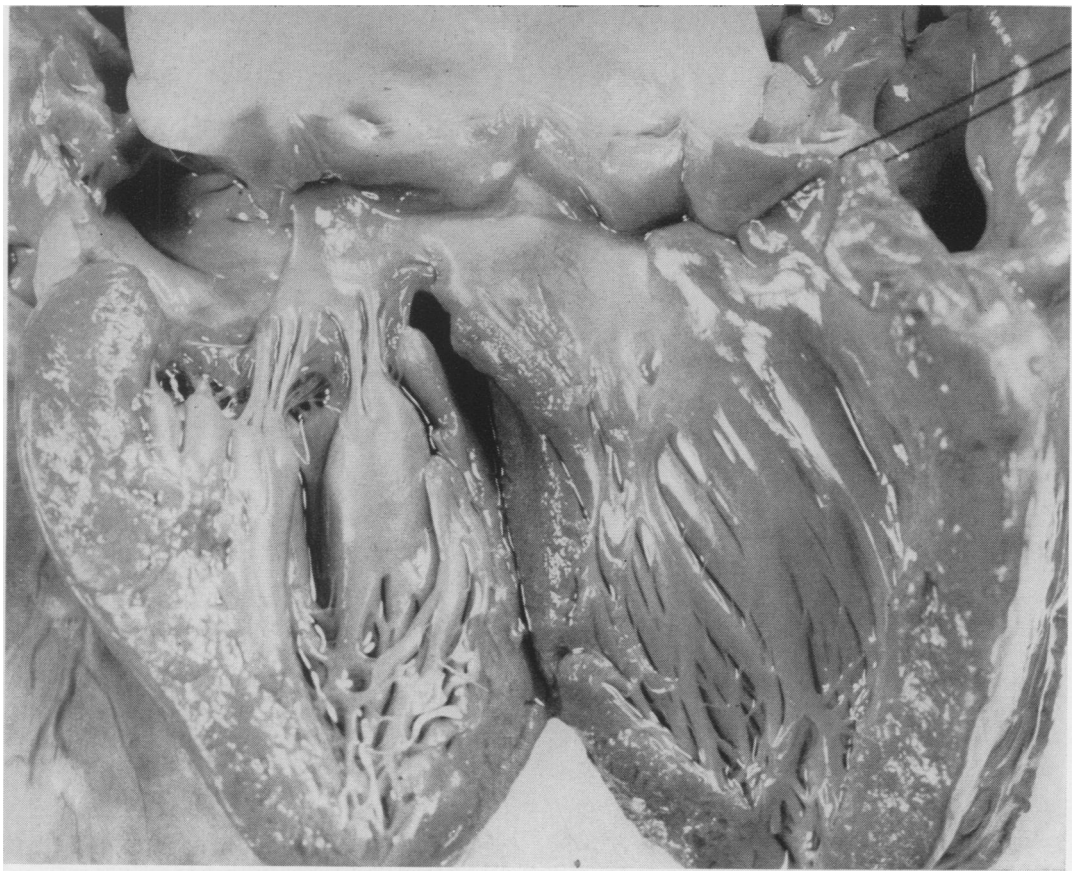

FIG. 4. Same heart viewed from the left ventricle after closure of the ventricular defect. Note restoration of the normal anatomy. The previous site of the septal defect is inconspicuous. The aortic valve function remains unimpaired by the closure.

Correction of the Pulmonic Stenosis. Next, the right ventricular outflow tract obstruction was dealt with either by resecting the infundibulum, performing a valvulotomy, or, as in one patient, with pulmonary atresia creating a new outflow tract for the right ventricle to which the pulmonary artery was anastomosed (Figs. 12 and 13). A specially modified Hartmann tonsil punch (Fig. 1) has been found especially useful for the resection of the infundibular obstructions, although the ordinary surgical scissors has also served satisfactorily.

The site and the amount of the infundibular muscle requiring resection in order to establish an adequate outflow tract may be expected to vary considerably, but in general for this resection these bites of tissue are preferably taken from the posterior-lateral aspect of the right ventricular outflow tract, with lesser amounts, or none at all, being resected from its posterior-medial aspect because of the nearness in this area of the aortic valve and the defect closure stitches.

In these tetralogy hearts it was early noted that a significant relief of the infundibular stenosis could be obtained by merely cutting directly through the stenotic area, and then closing this cardiotomy incision by sewing only the outer layer of the cardiotomy wound (Figs. 7 and 13C). In all of these patients with infundibular stenosis, this method of cardiotomy closure with continuous 000 silk doubly sewn has been utilized along with the infundibulectomy.

For the single patient in this series with

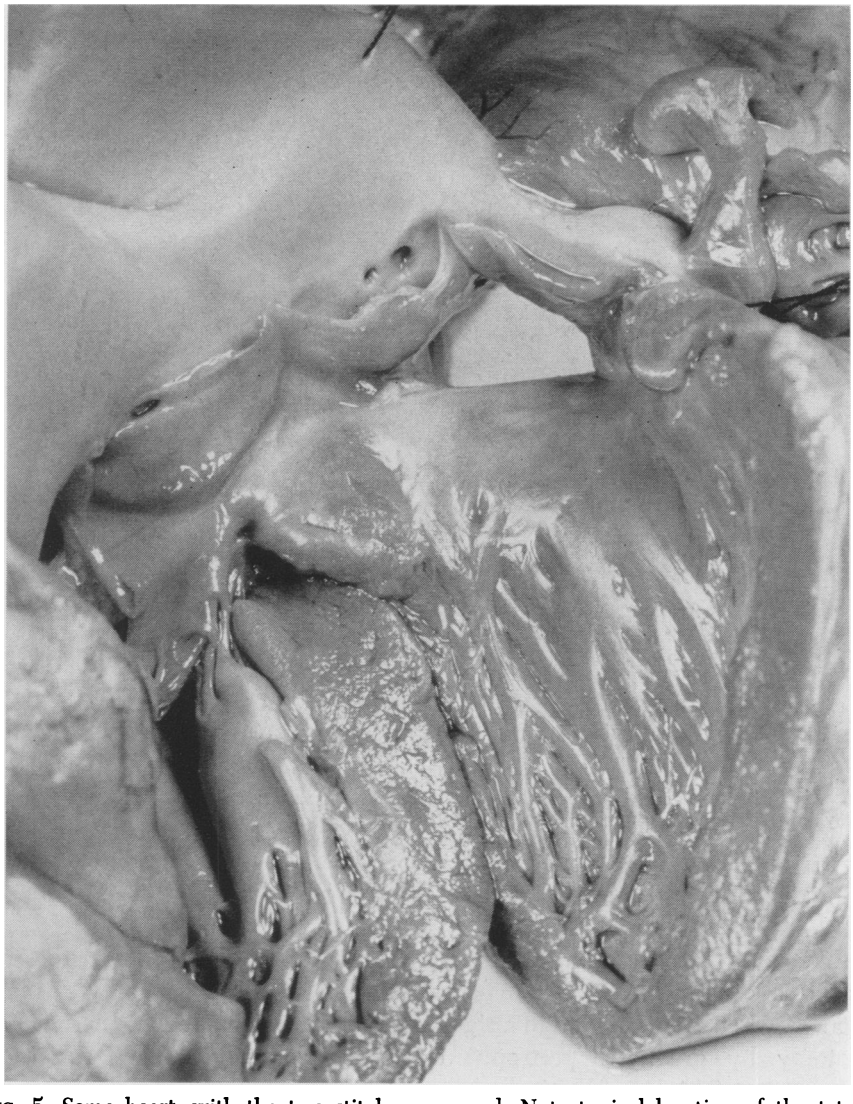

FIG. 5. Same heart, with the two stitches removed. Note typical location of the tetralogy ventricular defect extending up between the cusps of the aortic valve, and the aorta is seen arising entirely from the left ventricle.

the pentalogy of Fallot defect, the associated atrial defects were also closed under direct vision through an auricular cardiotomy.

A further description of the surgical technics formulated and found to be of value in the curative management of these defects will be presented in the sections that follow.

SURGICAL ANATOMY

All ten patients had high lying ventricular septal defects immediately adjacent or actually extending up between the cusps of the aortic valve (Fig. 5). Visualized from within the right ventricle, the tetrad septal defects less often have lain in the concealment of the tricuspid leaflets. Characteristically, they have occupied a more cephalad position within the right ventricle (Fig. 2). This position is in contrast to the usual location of ventricular defects occurring as an isolated lesion. Moreover, the septal defects of the tetralogy patients have tended to have

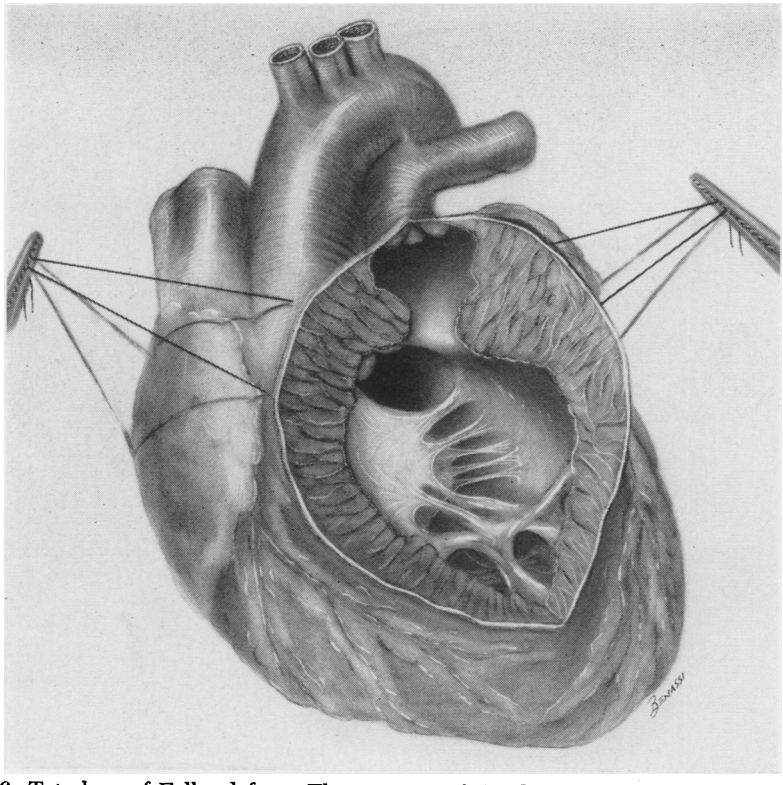

FIG. 6. Tetralogy of Fallot defects. The exposure of the defects as viewed in Case 1 (M. S., age 11 years) through the right ventricular cardiotomy incision. This patient had a well developed sub-valvular chamber. The ventricular defect was typically located cephalad to the tricuspid leaflets, and between the latter and the crista supraventricularis. The cusps of the aortic valve are characteristically easily seen from the right ventricular approach.

margins that were more muscular and less fibrous, and in certain hearts the defects were obscured when viewed through a right ventriculotomy by greatly hypertrophied papillary muscles. This muscle tissue has had to be transected to expose adequately the extent of the defect, and then resutured after closure of the septal defect had been completed to prevent any tricuspid insufficiency ensuing.

Among this tetralogy group the ventricular septal defects have been quite constant in their position and with regard to these characteristics. In contrast, the anatomy of the right ventricular outflow tract, particularly in its crista supraventricularis portion, has been rather variable. As indicated in Table I, seven out of these ten patients had an infundibular obstruction to within a few millimeters of, or involving the pulmonary valve. There was virtually no subvalvular infundibular chamber in these patients. This finding might have been anticipated from the criteria set for the selection of the patients submitted to the curative operation. This type of outflow obstruction that we have observed in the majority of these tetralogy patients operated upon is well recognized as being associated with the development of serious difficulties during infancy and early childhood.

Earlier descriptions of the tetralogy malformation have placed considerable emphasis upon the matter of a dextroposed aorta. It was even suggested that this would effectively hamper any surgical correction. Although our operative experience is yet small, it has wholly confirmed our previously ex-

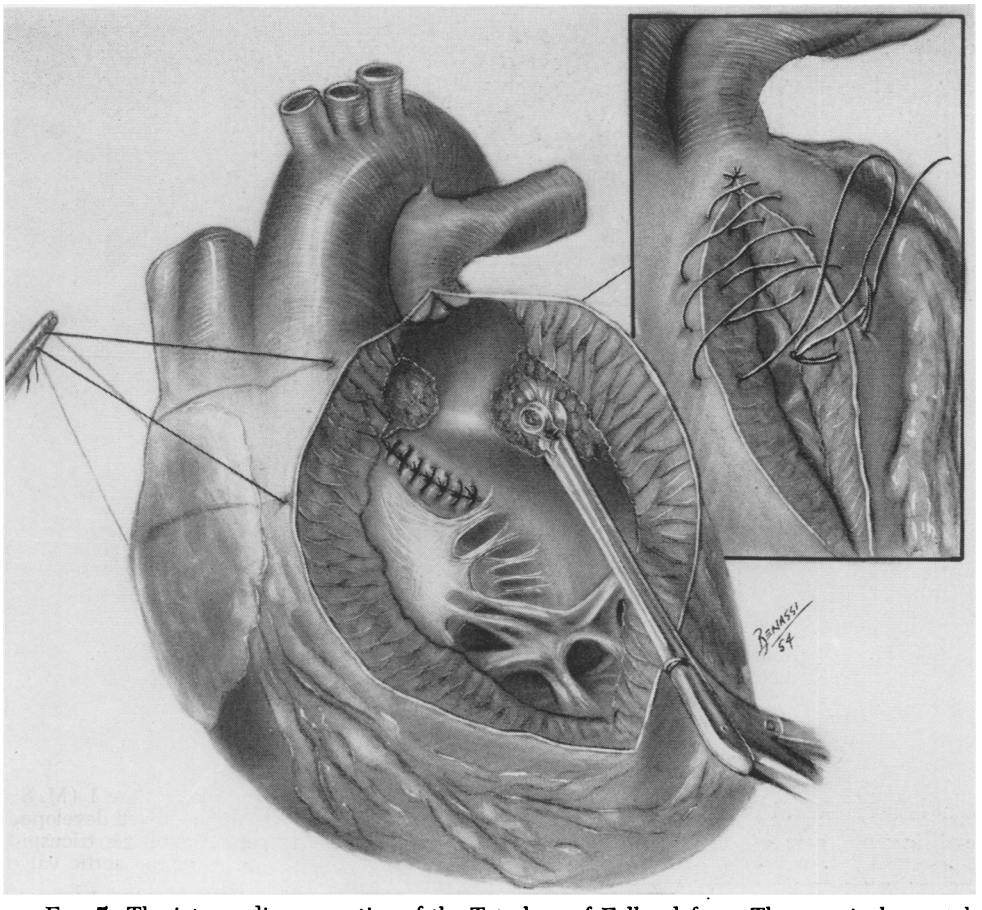

FIG. 7. The intracardiac correction of the Tetralogy of Fallot defects. The ventricular septal defect has been closed by interrupted silk sutures, and the infundibulectomy is in progress. The inset illustrates the method of closing the right ventricular cardiotomy incision by sewing together with continuous 000 silk only the superficial portions of the cut muscle edges over the stenotic area of the outflow tract. Away from this narrowed area the stitches may again be taken full thickness. This maneuver, which has been utilized in those patients with an infundibular type stenosis, leaves an internal myotomy which further augments the diameter of the pulmonary outflow tract.

pressed opinion[7] based upon observations made in the postmortem room in fresh autopsy specimens, in regard to the feasibility of completely curative cardioplastic procedures for these lesions. The apparent dextroposition of the aorta in this malformation has presented no serious obstacle to corrective surgery. It has not precluded the successful closure of these defects.

Actually we are inclined for several reasons to the belief that dextroposition of the aorta is more often illusory than real. Reference to Figure 2 discloses a photograph of the interior of the right ventricle of a patient with a severe form of the tetralogy defect (complete atresia of the pulmonary outflow tract*). There was apparent dextroposition of this aorta upon external inspection of the great vessels. Figure 5 is a photograph of this same ventricular defect from the left ventricular chamber. It is quite apparent that the aortic origin from the left ventricle

* J. W., age 6 months, U.H. 877898, who died suddenly during one of his episodes of anoxic unconsciousness just 12 hours prior to scheduled surgical correction of his defects. The pathology seen in this heart was virtually identical to that observed subsequently in Case 6 of the operative series.

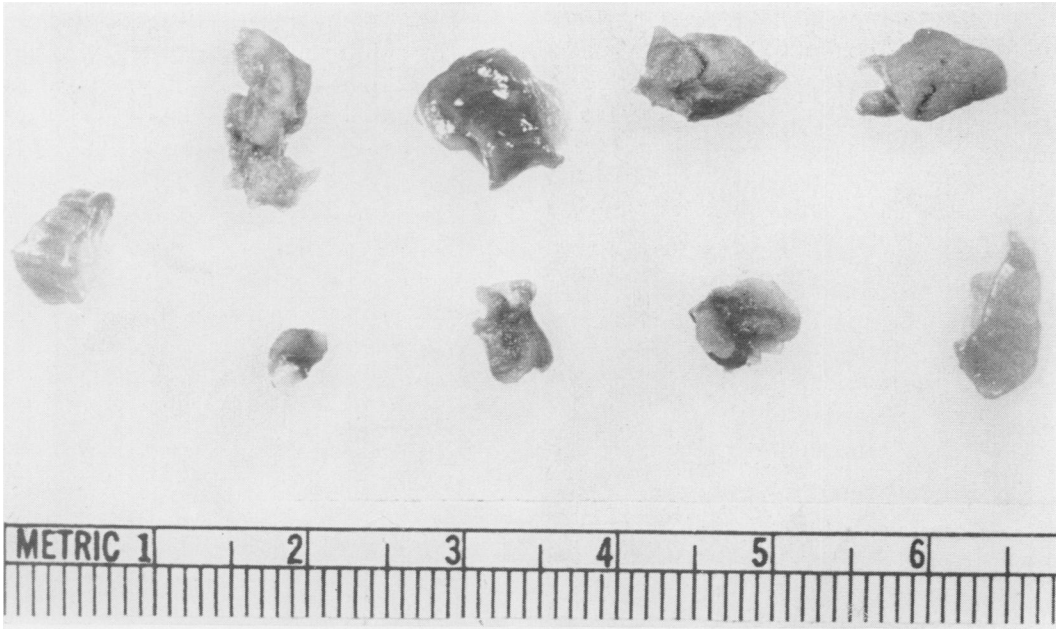

FIG. 8. Resected infundibular muscle obtained at operation in Case 1 (M. S.).

is entirely normal. In fact, if one excises this same portion of the ventricular septum in a normal heart, it creates the impression of an over-riding aorta if viewed from the right ventricle. This is so because normally the left ventricular outflow tract does encroach into the posterior wall of the right ventricle.

This concept is further supported by a careful inspection of venous angiograms in the left anterior oblique position. In such films there can be seen no essential difference between the opacified left ventricular outflow tract in a tetralogy patient, and in one with a normal heart. On the contrary, the child with transposition of the great vessels, and hence a true right ventricular origin of the aorta, demonstrates at angiography a marked difference. Here the aorta is dextropositioned. As corroborative evidence that the septal defect in these tetralogy patients can be readily sutured without encroachment or complications referable to the dextroposed aorta, we suggest reference to Figures 3 and 4. These photographs show the appearance of this defect after its closure in the autopsy room through a right ventricular cardiotomy incision of surgical extent. Only two U shaped stitches of 000 silk were necessary for a complete anatomic restoration. These photographs effectively deny the allegations of those who propose that the dextroposition of the aorta constitutes a significant barrier to corrective surgery for this lesion. To reiterate, our observations made during the last several years on this fresh* autopsy material studied carefully *in situ* have convinced us that this slight malposition constitutes no substantial obstacle to complete correction of the septal lesion.

Assuredly, a major factor contributing to this deception about the true amount of dextroposition of the aorta in tetralogy patients is the fact that the pulmonary artery is considerably diminished in size, while the aorta is proportionately enlarged. Also as the right ventricle hypertrophies, it ro-

* It should be emphasized that accurate anatomical observations are impossible in specimens that have been rendered rigid and distorted by formalin fixation.

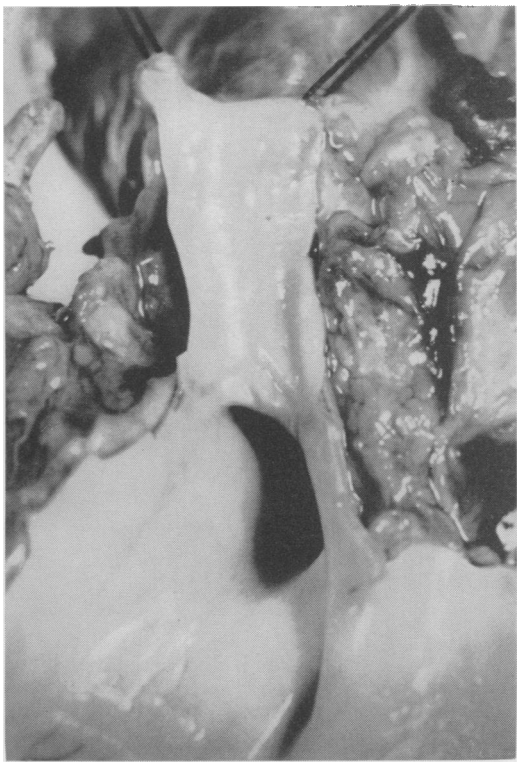

FIG. 9. The subclavian-pulmonary artery anastomosis from a tetralogy patient. This patient (J. A., Case 2) was critically ill from insufficient pulmonary blood flow despite this widely patent end-to-side anastomosis made two years previously. At autopsy the opened left subclavian artery (held up by the threads) is seen smoothly healed to the pulmonary artery below. This late morbidity and mortality is one of the obvious disadvantages of a palliative surgical procedure.

tates the entire heart to the patient's left, and magnifies any appearance toward dextroposition.

RESULTS

Ten patients, all seriously ill with various forms of pulmonic stenosis associated with a ventricular septal defect, have been consecutively managed by corrective surgery. The sum of their intracardiac defects have been corrected during an interval of open cardiotomy ranging from six to 21½ minutes. Pertinent data about these procedures are summarized in Table II.

Among the anatomic varieties of this defect successfully managed by corrective surgery was a patient (Case 6) with atresia of the main pulmonary artery as well as a ventricular defect, in which it was technically possible after closing the ventricular defect to create a new opening at the top of the outflow tract in the right ventricle, and to anastomose the pulmonary artery to this newly created orifice. Another patient (Case 10) with the pentalogy of Fallot defects was successfully treated by simultaneous atrial and ventricular cardiotomies for closure of the corresponding septal defects present, and then valvulotomy for relief of the pulmonary stenosis.

All of the patients were safely maintained during their interval of total by-pass by the cross circulation method. There have been no problems referable either directly or seemingly to the method *per se*. Three of the four deaths were due to technical misadventures associated with the learning phases of unfamiliar intracardiac technics. The fourth death was apparently due to a persistent complete heart block. Ventricular fibrillation has not occurred during the by-pass interval.

Abnormal bleeding has not occurred in the 20 patients and donors even though both were heparinized, and several of these cyanotic patients had abnormally prolonged bleeding and clotting times preoperatively. Postoperatively the bloody drainage from the thorax has been comparable to that measured in cardiac cases operated upon without use of by-pass technics.

The six patients who recovered from the operative procedure are all clinically well. All have been discharged from the hospital, and there has been no late morbidity or mortality amongst them. These six patients have had a systemic artery oxygen saturation* performed postoperatively, and in

* The samples were drawn from the unanesthetized patients breathing room air and were analyzed by the Van Slyke manometric method.

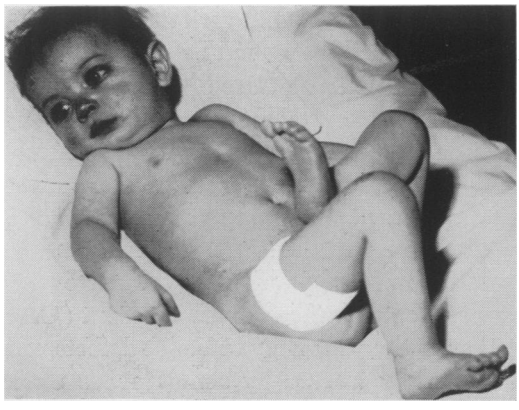

FIG. 10. Tetralogy of Fallot patient (W. B., age 19 mo.) prior to surgery. The systemic arterial desaturation (25 per cent) is evident. The patient was unable to sit up alone.

each these measurements were within the normal range. This is in clear contrast to their preoperative de-saturation (Table II).

Two of these six asymptomatic patients have undergone right heart recatheterization performed six and seven months respectively after their corrective surgery. The first patient (Case 1, Table III) exhibits physiologic results indicative that the anatomic repair is not perfect. Corresponding data obtained in the second patient (Case 4, Table III) are completely normal, indicating complete anatomic correction.

As a group, these tetralogy patients subjected to the curative operation, because of their normal lungs, have had a benign postoperative convalescence contrasting with the significant morbidity and mortality experienced during this same period in those patients with isolated ventricular septal defects, in many of whom the lungs have been ravaged preoperatively by the effects of pulmonary hypertension.

The donor mortality in these operative procedures remains zero, as it has for all other cases in which we have employed the cross circulation method to date. There has been no donor complications of significance in this group, either immediate or delayed in nature. Although the use of a donor in these procedures remains a significant responsibility, it is quite apparent that for the patient it is unlikely that a safer or more physiologic method of total by-pass will be developed.

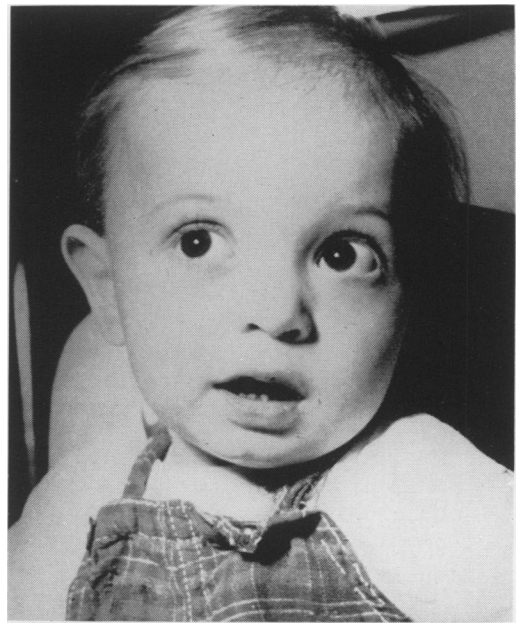

FIG. 11. Same patient, seen four weeks postoperatively, when he already had acquired the ability to sit unassisted. The femoral arterial oxygen saturation is 95 per cent, and recatheterization has indicated normal right heart measurements.

Additional details concerning the pathology of the lesions observed and the surgical technics formulated for their correction are presented in the following case reports.

REPORTS OF CASES

Case 1. M. S. was 11 years old at the time of his admission to the University Heart Hospitals March 28, 1954, for a subclavian-pulmonary artery anastomosis for his tetralogy of Fallot defect. A presumptive diagnosis of congenital heart disease had been made by the patient's family physician at 6 months of age. This was confirmed by more precise studies carried out in our cardiac clinic at the age of 6 years. The patient was followed at regular intervals in the outpatient clinic. At the age of 8 years his exercise tolerance had decreased to an ability to walk only 2 blocks, but he was still getting along in school tolerably well. At that time the parents were told of the availability of an anastomotic operation at such time as they wished it, or if his exercise tolerance diminished still further. A year later his exercise tolerance had decreased to less than 1 block with frequent stops for squatting.

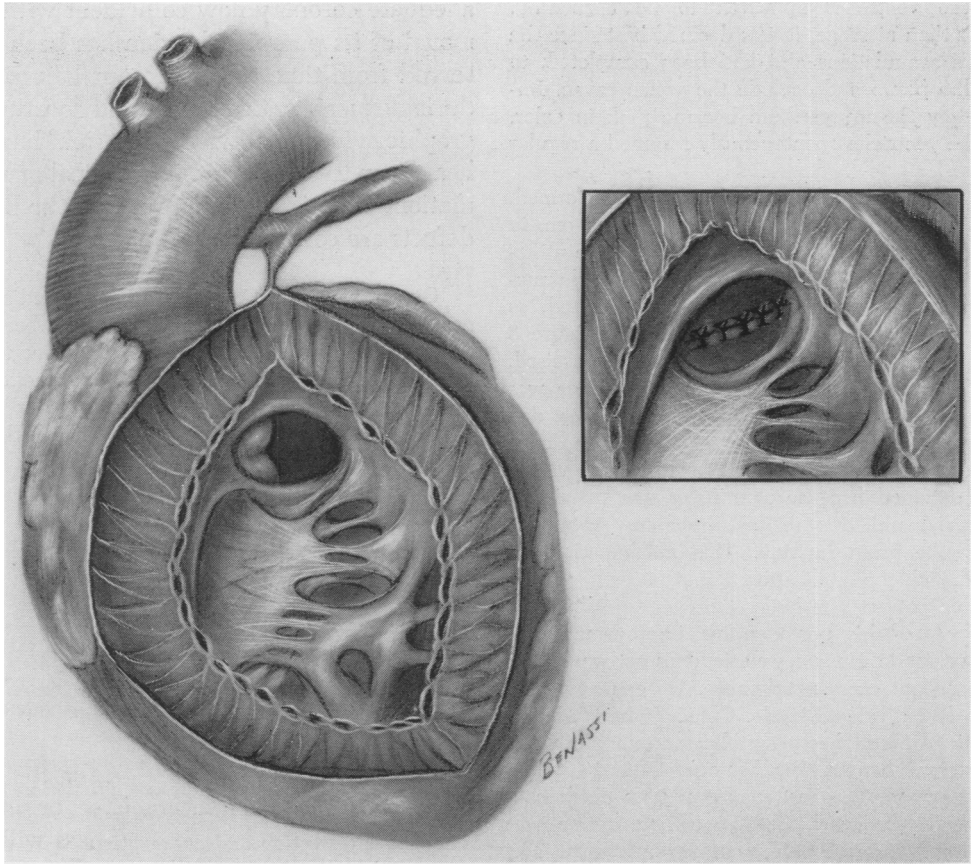

FIG. 12. The intracardiac correction of pulmonary atresia. The anatomy observed in Case 6 (L. K., age 22 mo.) through a right ventricular cardiotomy. The main pulmonary artery was tiny, and there was a solid muscular barrier at the site of the outflow tract. Inset shows the ventricular septal defect closed by direct suture (000 silk).

He was no longer able to attend school, and palliative surgery was scheduled. Between that time and his admission in March 1954, direct vision intracardiac surgical correction of ventricular septal defects had become a reality, and his parents elected to have the intracardiac operation performed. Neither parent was a compatible donor, so the patient was discharged for re-admission at a later date.

Physical examination revealed a thin, cyanotic boy, small for his age. His digits were clubbed. Auscultation disclosed a Grade III systolic murmur, loudest along the left sternal border in the 4th and 5th interspaces.

The hemoglobin was 16.4 Gm. The electrocardiogram showed marked right ventricular hypertrophy and strain. Cardiac fluoroscopy revealed a normal sized heart with a right-sided aortic arch, right ventricular enlargement, and pulmonary vascular markings within the normal limits.

Cardiac catheterization had been carried out previously (Table III), and disclosed right ventricular hypertension, pulmonary stenosis of an infundibular type, and systemic arterial de-saturation.

Surgical operation was performed August 31, 1954, utilizing a volunteer donor (H. H. age 29 years) from the boy's home town, for the cross circulation procedure. Upon opening the pericardium the heart had the typical appearance of a tetralogy. The aorta was large, 4 to 5 cm. in diameter, while the main pulmonary artery, although far smaller (1½ cm. in diameter), appeared to be of ample size. About 4 cm. proximal to the site of the pulmonary valve was a definite infundibular type stenosis with a well demarcated chamber distal, in which a thrill was palpable (Fig. 6).

The dissection and cannulations proceeded easily and rapidly. Despite this, and perhaps indicative of this boy's low cardiac reserve, as the last catheter was slipped into place the heart went into arrest

and the ventricles appeared to be fibrillating. Rather than massage, it was deemed best since the donor's cannulations also had been completed, to start the pump and occlude the venae cavae. Immediately the myocardium became pink in color, and the ventricles spontaneously resumed a regular beat.

A right ventricular cardiotomy was performed, exposing a high ventricular septal defect 3 cm. in diameter, which was closed by direct suture utilizing 6 stitches of 000 silk. The infundibular obstruction was resected and the cardiotomy closed, as indicated diagrammatically in Figure 7. Figure 8 is a photograph of the infundibular muscle excised.

The patient's postoperative convalescence was uncomplicated; he was acyanotic and was discharged on the 14th postoperative day. The donor was discharged 72 hours postoperatively, having been detained because of a fever due to a mild superficial phlebitis in his saphenous vein just below the groin incision. This inflammation resolved with the application of hot packs.

This patient has been seen periodically since surgery, and his postoperative improvement has been dramatic. He is acyanotic, does not squat, and has excellent exercise tolerance. He can now play baseball and ride a bicycle. Clinically he is asymptomatic. He still has a systolic murmur along the left sternal border. His hemoglobin is 13.3 Gm. His postoperative chest roentgenogram discloses a slightly larger heart. Cardiac catheterization was performed 7 months after corrective surgery. The findings indicated a significant increase in the volume of pulmonary flow. Also, he still has some residual stenosis of the right ventricular outflow tract which is probably fortunate since there is a small left to right shunt (Table III). Thus, although this patient clinically is well, and judging from the experience of Brock,[8] he may be expected to remain so, his cardiac malformations were not completely cured anatomically.

Comment. At the time that the patient's heart arrested, the fact that the donor's cannulations were completed and the donor and patient could be rapidly linked together in order to relieve the work load upon the patient's heart undoubtedly contributed to the successful result. This policy of timing the donor's and patient's cannulations has been followed regularly, especially in poor risk patients, and has also proved lifesaving in other patients.

The spontaneous defibrillation of the normothermic canine heart supplied with an adequate coronary flow coincident with removal of its pumping burden has been observed from time to time in our laboratory during experimental studies, and constitutes graphic evidence of why these "sick" human hearts can tolerate extensive surgical manipulations so satisfactorily, provided the basic defects are corrected.

Case 2. J. A., age 14 years, was admitted as an emergency transfer from her local hospital because of extreme dyspnea associated with pronounced cyanosis and progressive physical incapacitation that necessitated confinement to bed. Her past history disclosed that in August 1952 she had had a subclavian-pulmonary artery anastomosis performed with good relief of symptoms for about 1½ years. At that operation the pericardium had been opened for exploratory purposes. Auscultation of the chest on this admission disclosed a loud systolic murmur along the left sternal border. A diastolic murmur, previously present, could not now be heard. Chest roentgenograms at this time revealed a heart of normal size, right ventricular enlargement, sparse pulmonary vasculature, and moderately severe emphysema.

On September 23, 1954, corrective surgery was undertaken by means of the cross circulation technic, utilizing her father (age 42 years) as the donor. The initial dissection and cannulations were much more difficult in this patient due to the adhesions secondary to the previous intrapericardial exploration.

An attempt was made to dissect out the location of the main coronary artery branches before making the cardiotomy incision but the heart tolerated these manipulations so poorly they were terminated, and the cardiotomy was made without this advantage. It was noted that the cardiotomy incision cut across several unusually large coronary arteries. The infundibular stenosis, located less than 2 cm. below the pulmonary valve, was very tight. The ventricular defect was visualized and closed with 6 interrupted 000 silk stitches, and then the infundibular obstruction was resected. The fixation of the heart within the pericardial sac due to the previous adhesions also interfered with the surgical exposure during these procedures.

Upon release of the cavae the heart beat was strong and vigorous in the left ventricle, but the right ventricular contractions were weak. About 60 minutes later the heart arrested and resuscitation was unsuccessful.

At autopsy, it was found that the right main coronary artery, embedded in a thick layer of fibrous tissue covering the surfaces of the heart, had

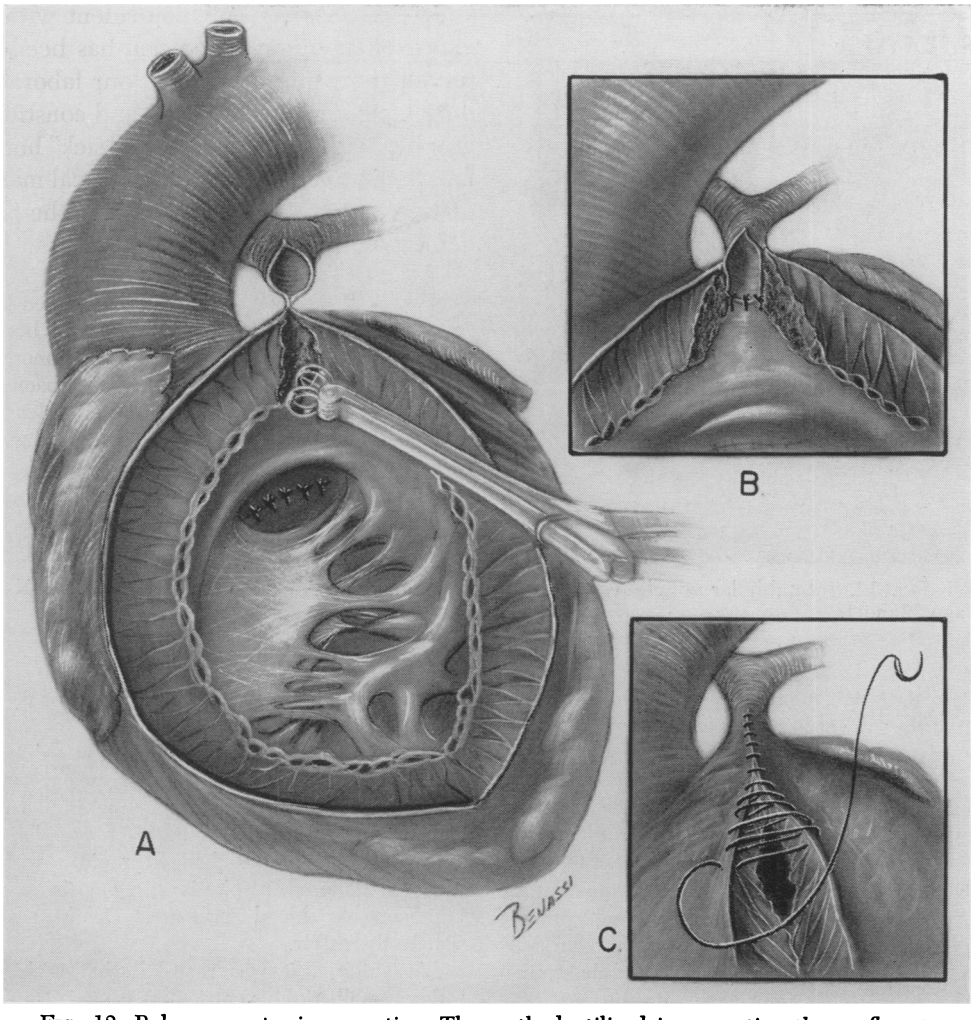

FIG. 13. Pulmonary atresia correction. The method utilized in correcting the outflow tract obstruction in patient L. K. (A) A new outflow channel is being cut through the solid infundibular muscle. The main pulmonary artery has been opened distally for orientation. (B) The opened pulmonary artery has been pulled down and anastomosed to the newly created outflow tract. There was no pulmonary valve present in this patient. (C) The cardiotomy incision closed by a continuous 000 silk stitch sewn superficially to further enlarge this outflow channel.

been transected by the upper end of the cardiotomy incision. The ventricular septal defect was well closed, and the surgical widening of the right ventricular outflow tract appeared adequate. The previous left subclavian-pulmonary artery end-to-side anastomosis was widely patent (Fig. 9).

The donor's convalescence was uncomplicated, and he was discharged 72 hours postperfusion.

Comment. Two considerations in this girl's clinical course are particularly worthy of emphasis. First, she illustrates some of the hazards for the patient of having the pericardial sac opened prematurely and without having available a surgical procedure for correcting all of the defects that one may reasonably expect to encounter. Secondly, she is an example of the late morbidity and mortality accompanying palliative procedures. This girl was critically ill from insufficient pulmonary blood flow, even though her previously made subclavian-pulmonary artery anastomosis was found to be anatomically intact at autopsy.

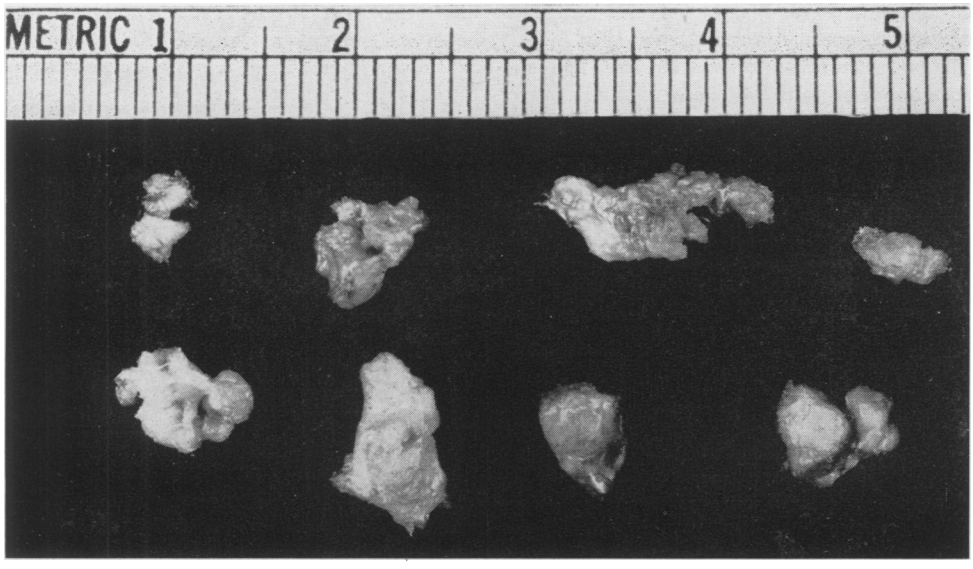

FIG. 14. Infundibular muscle resected in correction of the pulmonary atresia (Case 6, L. K., age 22 mo.).

Case 3. J. W., age 1½ years, was a very ill infant at the time of his admission for surgical treatment. He was experiencing episodes of unconsciousness associated with extreme cyanosis occurring several times each day. He was unable to sit up unsupported. Angiocardiography and cardiac catheterization had established the diagnosis of tetralogy of Fallot. Preoperatively, his hemoglobin was 18 Gm., with an erythrocyte count of over 9 million. Both the bleeding and clotting times were markedly prolonged (over 18 minutes on repeated determinations). The platelets prior to surgery were 57,000; his blood fibrinogen level was also decreased. Corrective surgery was undertaken October 19, 1954, utilizing a cousin (age 34 years) of the child's father as the donor.

Upon entering the chest the aorta was large (3 cm. in diameter) as compared to the main pulmonary artery (3 to 4 mm. in diameter). The latter was so soft to palpation that it could not be certain that any blood was flowing through it. There was extensive collateralization about the pulmonary hila and chest wall. The right ventricular cardiotomy extended through a very tight infundibular stenosis (2 mm. diameter) located 5 mm. below the pulmonary valve. The ventricular defect was 2 cm. in diameter, and seemed unusually easy to close with 4 000 silk stitches. One bite of infundibular muscle, 4 x 5 mm., was resected and appeared to relieve the infundibular stenosis quite adequately. Upon release of the venae cavae the heart took over with a strong beat. Despite the initial small size of the pulmonary artery, the pulmonary flow appeared adequate.

The patient's color improvement was dramatic. The bleeding and clotting times postoperatively were normal. He appeared to be making an excellent recovery when 12 hours after completion of surgery his condition changed rapidly with the onset of labored respirations, cyanosis, and an imperceptible blood pressure. Death occurred shortly thereafter.

At autopsy, it was revealed that within this rather small right ventricular cavity, the thick hypertrophied anterior papillary muscle coursing transversely across the ventricle had been mistaken for the inferior margin of the ventricular defect, and stitches had been placed between it and the margins of the upper portion of the defect, leaving the lower portion of the ventricular defect open.

The donor's convalescence was uncomplicated, and he was discharged 36 hours postperfusion.

Comment. Our failure to close his ventricular septal defect combined with an adequate resection of his pulmonary outflow obstruction probably produced an intolerable left to right shunt, and death ensued from acute cardiac decompensation.

Case 4.* W. B., age 19 months, an intensely cyanotic infant, was admitted to the hospital in

* A more detailed report upon this patient has been presented elsewhere.[10]

cardiac failure and was regarded by our pediatric cardiologists as terminal in appearance (Fig. 10C). After digitalization he improved somewhat. He had been cyanotic from birth, with progressively severe manifestations of dyspnea, cyanosis, and failure to develop normally. The severity of this child's difficulty, together with the finding of cardiomegaly on chest roentgenogram, favored the diagnosis of complete transposition of the great vessels. However, angiocardiography and cardiac catheterization subsequently provided a correct interpretation by ruling out the presence of complete transposition, and suggesting the diagnosis of Fallot's tetralogy. Corrective surgery with direct suture closure of his septal defect and resection of his infundibular obstruction was done on December 3, 1954, utilizing his father (age 28 yrs.) as the donor. The ventricular septal defect was 2.5 cm. in diameter and the infundibular stenosis was located just below the pulmonic valve, with an orifice only 3 mm. in diameter.

The child's improvement was dramatic and immediate, and has persisted. Postoperatively, he quickly acquired an ability to sit alone before leaving the hospital (Fig. 11). Recently this child returned to the hospital 6 months after corrective surgery for postoperative studies. His mother stated that he was now as active as his normal siblings. Recatheterization at this time disclosed a normal circulation with normal intracardiac pressures (Table III). The chest roentgenograms taken postoperatively have shown a progressive diminution in the size of his cardiac silhouette, as compared with its preoperative appearance.

The donor's convalescence was uncomplicated, and he was discharged 36 hours postperfusion.

Comment. This boy, who was desperately ill prior to operation, represents the first patient, as far as we are aware, to have the tetralogy of Fallot defects completely corrected as evidenced by his postoperative clinical course and confirmed by subsequent objective measurements.

Case 5. G. F., a 2½-year-old girl, was first examined in our outpatient clinic at age 7 months, when a diagnosis of tetralogy of Fallot was made on the basis of intense cyanosis, feeding difficulties, listlessness, clubbing of her digits, and a loud, systolic murmur along the left sternal border. During the next 2 years the patient was seen frequently in the outpatient clinic, and was hospitalized twice due to a progressive increase in her disability.

Corrective surgery was undertaken December 10, 1954, by the cross circulation technic, utilizing her father (age 26 years) as the donor. Closure of the ventricular septal defect (1.5 cm. in diameter) was made more difficult by the fact that her defect lay underneath a very prominent supraventricular crest. The latter, in turn, over-hung as a curtain the septal defect. The defect closure was obtained by means of 7 silk 000 stitches. The pulmonary stenosis was valvular, and was relieved by valvulotomy.

Following the closure of the cardiotomy and release of the cavae the heart took over readily. However, it was significant that despite this good heart action the anesthesiologist was unable to detect a peripheral blood pressure. Likewise, immediately postoperatively her color was characterized by an ashen pallor, and the heart beats were difficult to hear because the second sounds were absent. This child expired 2 hours after surgery.

At autopsy the explanation of the unusual manifestations noted clinically in the postoperative course was clearly evident. The ventricular defect was tightly closed and the pulmonary stenosis adequately relieved, but 2 of the cusps of the aortic valve had been sutured back against the aortic wall by misplaced sutures. This created a free aortic regurgitation, and death was due to the acute heart failure so induced.

The donor was discharged in good condition 24 hours postperfusion.

Comment. This patient's tragic outcome re-emphasized to us the lesson, previously learned from autopsy specimen studies, concerning the advisability of placement of the closure stitches on the right ventricular aspect of the defect margins.

Case 6. L. K., an intensely cyanotic, underdeveloped, 22-month-old female infant, who had considerable dyspnea at the time of her admission for the first time to the University Hospitals in December 1954. Cyanosis had been present since birth. Poor appetite had been constant. During the last year the parents had noted a definite decrease in the child's ability to do things requiring physical exertion. Chest roentgenograms and angiocardiography December 23, 1954, were characteristic of severe pulmonic stenosis with a ventricular septal defect. Cardiac catheterization was not performed, but preoperatively the femoral artery oxygen saturation was 39 per cent. The preoperative diagnosis was severe tetralogy of Fallot or pulmonary atresia.

Surgical exploration was undertaken December 31, 1954, with the father (age 43 years) available as the donor. Upon opening the pericardium the heart was intensely cyanotic and the main pulmonary artery was small (4 mm. in diameter at its widest portion) and very soft. There was no thrill in

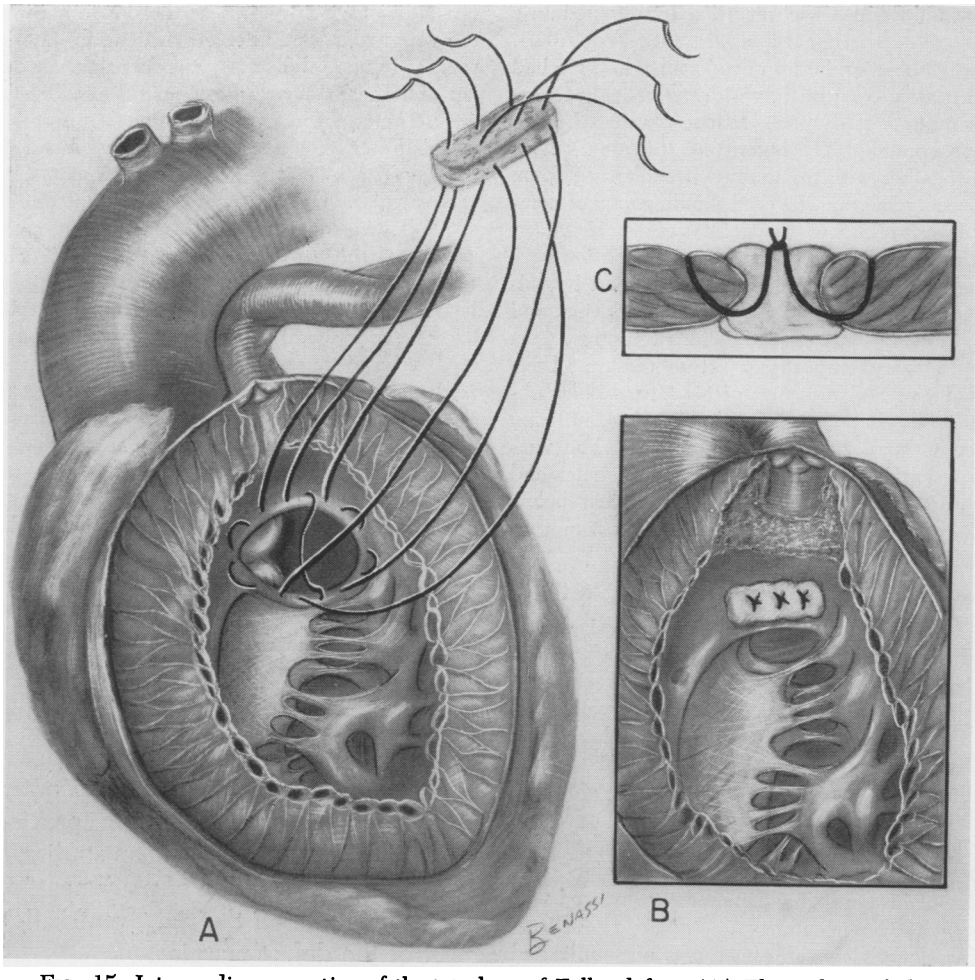

FIG. 15. Intracardiac correction of the tetralogy of Fallot defect. (A) The technic of closure of the ventricular septal defect utilizing stitches, placed in the same manner, but tied down on a pledget of compressed polyvinyl sponge (Ivalon). This method was used in patients 8 and 9 in this series. The stitches were placed first, as shown. A half purse-string suture was placed at either end of the defect, with one or more Cushing type mattress stitches between. The placement of these types of stitches on the right ventricular aspect of the defect margins guards against injury of the aortic valve cusps. The needles were then threaded through the Ivalon sponge. (B) The closure stitches tied over the plastic sponge and the infundibular outflow barrier resected. (C) A schematic cross sectional view through the area of the septal defect closure with the Ivalon sponge in place.

this artery, and no apparent flow of blood within it. The aorta was large, some 8 times the diameter of the pulmonary artery.

Upon opening the right ventricle there was no outlet of the right ventricle detectable under direct vision with the use of a fine probe. Instead, there was a high infundibular stenosis of solid muscle about 1½ cm. in length, constituting a complete atresia of the outflow tract. A ventricular septal defect (1½ cm. in diameter) was also present, and this was closed by direct suture (Fig. 12) with 000 silk.

Then, to achieve orientation the pulmonary artery distal to this outflow barrier was opened and the muscular obstruction was cut out with the muscle biting forceps. The widest available portion of the pulmonary artery was then pulled down and anastomosed by interrupted silk stitches to this newly created outflow tract, as indicated diagrammatically in Figure 13. The infundibular muscle resected is shown in Figure 14. There was no pulmonary valve present.

She awoke immediately after surgery, was responsive, but for the next 48 hours her color, al-

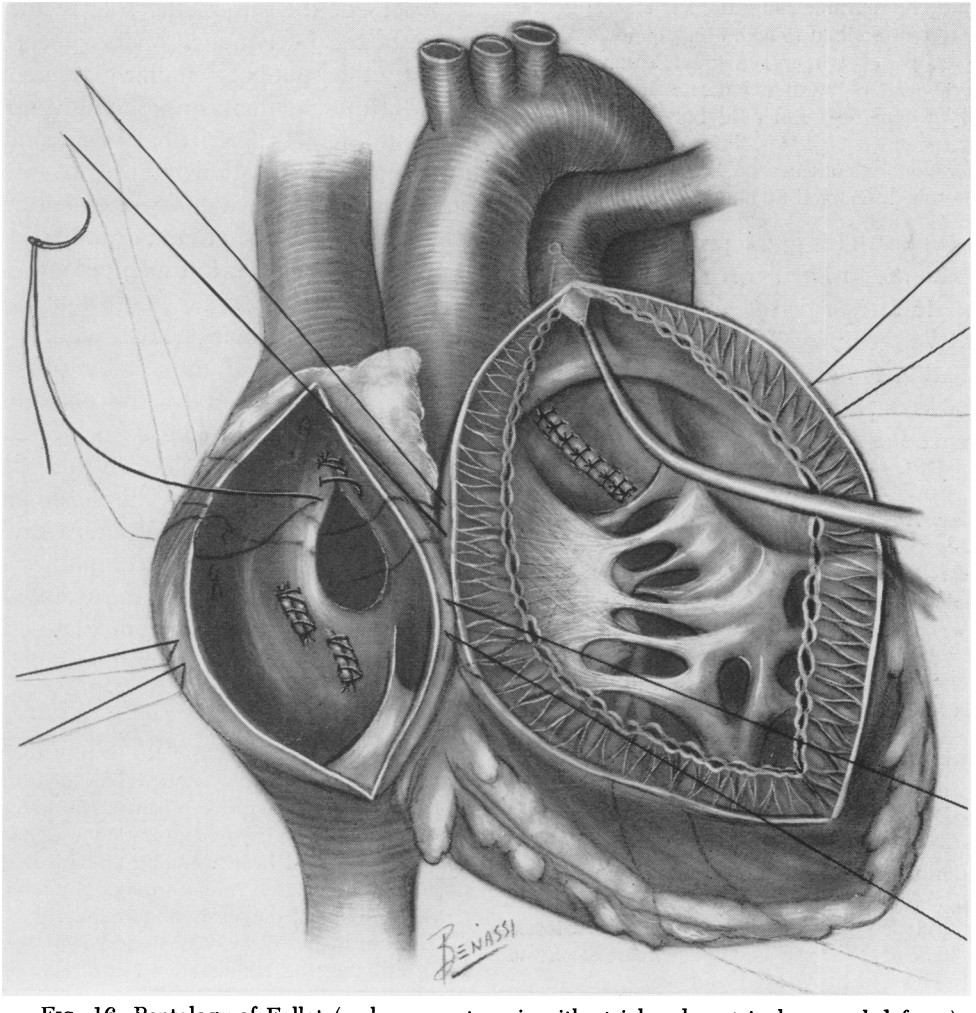

FIG. 16. Pentalogy of Fallot (pulmonary stenosis with atrial and ventricular septal defects). The anatomy observed and the operative corrections carried out in Case 10 (D. S., age 10½ years). Synchronous right sided bichamber cardiotomies gave excellent exposure for suture of the multiple atrial and ventricular septal defects, as illustrated. The pulmonary stenosis in this patient was entirely valvular, and was corrected by valvulotomy.

though much improved over its preoperative state, had an ominous ashen hue, associated with a very rapid pulse (160 to 170/min.); respirations, 54 to 68/min.; and a low blood pressure (68-74 mm. Hg systolic). We interpreted this clinical picture as being consistent with a pulmonic stenosis in a patient with intact septa. During this period she was digitalized and empirically maintained upon intravenously administered heparin to minimize any further narrowing of her new outflow tract due to the temporary deposition of fibrin. It was presumed that she was experiencing some difficulty adjusting to the small pulmonary artery.

Following this initial period, she began a progressive and continued improvement. Her convalescence was somewhat prolonged by the appearance of a right chylous thorax* which was treated non-operatively by replacing her daily losses from the thoracic drainage catheter by intravenous plasma until the fistula dried up. She was discharged 39 days postoperatively in excellent condition. A femoral artery oxygen saturation at that time was 95 per cent.

Follow-up visits in the clinic have disclosed that she has maintained this result with normal exercise tolerance and color. She has both a systolic

* A retroesophageal right subclavian artery joining the descending aorta was utilized for the arterial cannulation.

and diastolic murmur along the left sternal border. The latter is ascribed to lack of a pulmonary valve. Postoperative chest roentgenograms compared with the preoperative studies indicate increased pulmonary vascularity, and mild but definite cardiac enlargement.

Her donor's convalescence was uncomplicated, and he was discharged 36 hours after the operation.

Comment. This girl represents the first of our patients to have corrective surgery for complete atresia of the pulmonary outflow tract. We recognize that physiologically her circulation is not completely normal, since she lacks a pulmonary valve, but available evidence suggests that this is a well tolerated defect.

Case 7. J. S., age 3½ years, had demonstrated a steady progression of difficulties due to dyspnea and diminished exercise ability which permitted him to walk less than 1 block. He also had developed a sustained polycythemia, with an erythrocyte count above 10 million per cubic millimeter.

Corrective surgery was undertaken February 4, 1955, utilizing a cousin (age 35 years) as the donor. The external cardiac configuration was typical of a tetralogy defect, with the aorta being about 3.5 cm. in diameter and the pulmonary artery 1 cm. in diameter. The ventricular septal defect (2 cm. in diameter) was closed by direct suture, and a high infundibular obstruction with a small ostium located within a few millimeters of the pulmonary valve was resected. Following closure of the cardiotomy and release of the cavae a complete heart block was noted. This block persisted and worsened. In the postoperative period the ventricular beats became progressively slower and less responsive to stimulants. Death from cardiac arrest occurred 11 hours after completion of the surgery.

At autopsy it was found that the large ventricular defect had been correctly closed, and there was an adequate sized pulmonary outflow tract, but in rongeuring out the high infundibular obstruction a portion of the pulmonary valve had also been resected.

The donor was discharged 2½ days after the perfusion. This discharge was delayed 24 hours by the presence of a temporary fever of undetermined origin.

Comment. The most likely explanation of the occurrence of the complete heart block in this patient would be to ascribe it to the stitches placed in the ventricular septum for closure of his defect. However, we are inclined to doubt this as the sole factor because of the infrequency of its appearance in the other patients similarly treated. In many of these patients undergoing ventricular septal defect repair it has been common to observe a dissociation of the atrioventricular beats during the actual placement of the stitches, but this arrhythmia usually corrects itself promptly once the manipulations are completed. It may be suggested that the injury to the pulmonary valve, while not a serious lesion in itself and usually quite well tolerated as indicated by the outcome in Case 6 above, could have been a factor in perpetuating the heart block in this patient under these conditions by diminishing the effective right ventricular output. At any rate we are unable to offer any completely adequate explanation for the unsuccessful result in this case, since the immediate operative procedure had been without incident otherwise.

Case 8. P. R., age 18 months, was at the time of surgery a critically ill cyanotic infant having daily tonic and clonic convulsive seizures. Her preoperative femoral artery oxygen saturation was only 35 per cent. She had been cyanotic and in difficulty since her birthdate. Angiocardiography substantiated the clinical diagnosis of tetralogy.

Intracardiac surgery was undertaken April 5, 1955, utilizing the child's aunt (age 34 years) as the donor. Although there had been no complications and the operative procedure had progressed rapidly, by the time the patient's cannulations had been completed the heart action was very feeble. At this point the circulations of the patient and donor were rapidly linked together, the pump started, and the patient's caval inflow occluded. Subsequent to these maneuvers, the heart immediately turned pink and resumed a healthy beat. Upon entering the right ventricle it was necessary to transect a large papillary muscle to expose the defect (2 cm. in diameter) which still lay partially concealed by a well developed crista supraventricularis. Three stitches of 000 silk were placed for closure as diagrammed in Figure 15. Then, the needles on the free ends of these stitches were threaded through a small pledget of Ivalon and tied down, giving a satisfactory closure (Fig. 15). Approximately 5 bites removed the infundibular stenosis located about 4 mm. below the pulmonary valve. The transected papillary muscle was re-

sutured. The cardiotomy was closed in the usual manner, sewing the upper portion near the outflow tract superficially to maintain a maximum outflow tract. The infant's convalescence was uncomplicated. She has been completely well since the recovery from her operation, and has remained entirely free of all seizures. Her color is now normal.

The donor's recovery was uneventful, and she left the hospital 36 hours after the procedure.

Comment. This infant's initial reaction to the thoracotomy illustrates again the limited cardiac reserve present in some of these seriously ill patients. This cardiac status improves immensely the moment its work load is removed, while it continues to receive an adequately oxygenated coronary flow. Successful accomplishment of the intracardiac repairs almost guarantees maintenance of this improved state because of the lessened burden upon the heart and the improved oxygen content of its coronary flow.

The use of a pledget of Ivalon sponge in this case was to test its effectiveness in lessening any tendency for the closure stitches, which for the most part must be placed in muscle tissue, to cut out under the continued impact from each heart beat. Delayed opening of a septal defect snugly closed at operation has been recognized as a possibility in these cases, but we have relatively little evidence that it has ever occurred. In the first six patients with isolated interventricular septal defects who have undergone recatheterization following corrective surgery there has been a complete closure of the defect in all.[3] We are undecided as to whether the one patient in this series of tetralogy patients who has been found to have a shunt remaining (Case 1, Table III) represents a delayed opening or an incomplete initial closure.

Case 9. S. C., age 7 years at the time of surgical correction, was first studied at our hospital at 1 year of age because of cyanosis of 9 months' duration. She was one of fraternal twins; the other twin sister being normal. The mother soon noticed the onset of dyspnea, cyanosis, and episodes of syncope associated with crying in this patient.

For the next 6 years the patient got along tolerably well. The patient was not brought back to the clinic during that time. During this interim of 6 years her cyanosis had become intense, and at the time of her next clinic visit in 1955 her hemoglobin was 27 gm., the hematocrit was 82 per cent, and her weight was 36 pounds as compared to 56 pounds for her twin sister. Interestingly, the mother stated that the patient's mental development had always remained superior to her twin, although she had never been able to attend school. The parents were now concerned because of the onset of generalized convulsive seizures occurring every few weeks during the 6 months preceding this admission. Upon physical examination she was intensely cyanotic, underdeveloped, and appeared undernourished, with marked clubbing of fingers and toes. Angiocardiography confirmed the diagnosis of tetralogy. The preoperative femoral artery oxygen saturation was 53 per cent.

Surgical correction of the patient's cardiac anomalies was undertaken April 12, 1955, using her father (age 39 years) as the donor. At operation the heart was definitely enlarged for a tetralogy. There was a definite infundibular stenosis visible externally about 2 cm. below the pulmonary valve. There was no thrill palpable distal to this obstruction, and this, together with the softness of the pulmonary artery, suggested that little if any blood was flowing from the right ventricle directly into the pulmonary artery. The aorta was large (5 cm. in diameter), the pulmonary artery was much smaller but was considered good sized (1.5 cm. in diameter). The right ventricle was opened and the cardiotomy extended directly through the infundibular stenosis. There was ostium of only 2 mm. diameter through this stenotic area, which was about 1 cm. in length. The ventricular defect, about 2 cm. in diameter, was visualized and closed in a manner similar to that described for the preceding case (Fig. 14). Infundibulectomy was then performed for this severe stenosis, taking out 8 to 10 bites, which resulted in a very adequate appearing right ventricular outflow tract. The interior of the heart was noticeably more bloody in this patient during the by-pass interval due to her well developed bronchial collateral circulation.

Postoperatively the patient's immediate color change from intensely cyanotic to normal was dramatic. Her convalescence was uneventful save for a chylous fistula which responded readily to the type of non-operative management already mentioned. At discharge from the hospital the patient's color and clinical appearance were normal. Her first femoral artery oxygen saturation performed 6 weeks postoperatively was 94 per cent. A chest roentgenogram 3 months postoperatively shows the heart to be unchanged from its preoperative size, and with a more normal configuration and a significant increase in the vascular markings.

The donor's convalescence was delayed one day by fever, and he left the hospital 2½ days after the perfusion.

Case 10. D. S., age 10½ years, was an underdeveloped, cyanotic boy with a very limited exercise capacity due to exertional dyspnea. Cyanosis and a systolic heart murmur had been present since 6 weeks of age. For the year just prior to surgical correction he had suffered also from transient episodes of complete heart block. These attacks were unrelated to digitalis therapy and recurred after the withholding of this remedy. Roentgenograms disclosed diffuse cardiomegaly with normal or slightly decreased pulmonary vascular markings. Preoperative cardiac catheterization and angiocardiography suggested the likelihood of an atrial septal defect in addition to those defects usual to a tetralogy.* His preoperative electrocardiogram showed right axis deviation and right ventricular preponderance, with marked irregularity, sometimes bigeminy involving many multifocal premature ventricular contractions as well as nodal beats.

Surgical correction of all of these defects was undertaken May 6, 1955, utilizing a friend of the family (age 31 years) as the donor. Upon entering the chest through the usual bilateral anterior thoracotomy in the fourth interspace and opening the pericardium, the heart was seen to be much larger than the usual tetralogy heart but otherwise had the typical appearance. The aorta was large (5.0 cm. in diameter), and the main pulmonary artery was about 1½ cm. in diameter, with a palpable thrill in it. The right atrium was only moderately enlarged, but it had an unusually thick wall. The previously mentioned cardiac irregularity was conspicuous, and in addition the heart was especially irritable, racing into a sustained tachycardia upon being touched. Another abnormality of importance to recognize before opening the heart was the presence of a sizable left superior vena cava draining into the coronary sinus.

Despite the fact that care was taken to perform the necessary cannulations rapidly and with the gentlest possible cardiac manipulations the patient's heart was virtually in arrest at their completion. Thus, the patient was linked to his ready donor and the pump was turned on. Cardiac inflow stasis was secured by occluding the tapes previously placed about the right and left superior and the inferior venae cavae. The heart immediately turned pink in color, the contractions increased in vigor, and although its rhythm had been grossly irregular for more than a year previously, its beat immediately became perfectly regular. The right ventricle was opened first and a large ventricular septal defect (4 cm. in diameter) was visualized and closed by direct suture of the opposing margins, utilizing a number of interrupted 000 silk stitches (Fig. 16). The pulmonary stenosis was entirely valvular and was relieved by valvulotomy. With the right ventricle open, a large atrial septal defect could easily be visualized through the tricuspid valve. To expose this lesion an atrial cardiotomy was performed, which disclosed this larger defect (2.0 cm. in diameter) located low in the atrial septum, being separated from the ventricular defect by only a narrow (3 to 4 mm.) band of tissue, and also two other atrial defects (½ cm. each in diameter) in the *septum secundum* area. All of these openings were sutured with continuous 000 silk reinforced with occasional interrupted stitches (Fig. 16). Both cardiotomies were then closed and the cavae released. The heart took over readily and maintained a completely regular rhythm in contrast to its grossly irregular preoperative status.

The patient's color improvement was evident immediately postoperatively. About 72 hours postoperatively he again had the onset of a pulse irregularity, which was due to ventricular extrasystoles. These ectopic beats were thought to be originating from foci about some of the numerous intracardiac stitches, and he was treated by digitalization and pronestyl therapy. The arrhythmia responded to this therapy. A chylous fistula also occurred postoperatively, and required daily intravenous plasma and serum albumen replacement until it dried up. A femoral artery oxygen saturation at the time of discharge from the hospital was 91 per cent.

The donor was discharged 3 days postperfusion. His only complication was a fever of unexplained origin, lasting 48 hours.

Comment. This boy represents the first patient in our series with a pentalogy of Fallot defect to have complete correction of his defects.

The successful outcome in this boy again illustrates the advantages, for those patients in whom a low cardiac reserve may be predicted from their preoperative status, of having the donor's cannulations completed and the pump calibrated so that the hook-up can be completed rapidly if necessary. The reversion of the cardiac rhythm to normal as soon as the pumping burden was removed is graphic confirmation of the experimentally

* For brevity, it seems convenient to refer to this combination of defects as the pentalogy of Fallot.

deduced observations that the heart is immediately improved if it is relieved of its work load while being supplied with coronary flow. It is this fact that has made these operations possible in critically ill patients at a reasonable risk. Chylous thorax has been a rather annoying complication in these tetralogy patients but has responded well to nonoperative management. In a much larger series of acyanotic patients subjected to this same form of operative management this complication has not occurred at all, indicating that there is a much greater lymphatic flow in these cyanotic individuals.

SUMMARY AND CONCLUSIONS

Ten patients, all of whom were seriously ill with Fallot's tetralogy, have been consecutively operated upon by direct vision intracardiac curative procedures, utilizing controlled cross circulation for the necessary interval of total by-pass of the heart and lungs. All of the six survivors have a normal clinical appearance, and are asymptomatic postoperatively. The systemic artery oxygen saturations have been measured postoperatively in these six patients, and have been found to be within the normal range in all in contrast to their significant preoperative desaturation. The first two patients operated upon have now had right heart recatheterization performed six to seven months postoperatively. The first patient remains asymptomatic, but has a small left to right shunt high in the right ventricle. The other boy has normal oxygen contents and pressure measurements demonstrating unequivocally that complete anatomical and physiologic restoration to normal can be achieved in a patient born with these tetralogy defects.

Also numbered among the cases successfully managed by these corrective technics are two more complicated varieties of the tetralogy defect. The first of these was the 22-month-old girl with complete atresia of the main pulmonary artery at its origin, in whom it was possible to close the ventricular septal defect and to create a new opening at the top of the outflow tract in the right ventricle to which the pulmonary artery was anastomosed. The other was the 10½-year-old boy with a pentalogy of defects in whom simultaneous atrial and ventricular cardiotomies were well tolerated. It is significant for the future that the four seriously ill infants in this series, a therapeutic problem by any palliative method of management, were successfully treated by intracardiac correction in three instances.

As a result of these gratifying successes, together with the four instructive failures, the curative operation has been adopted in our clinic as the method of choice for all patients with the tetralogy of Fallot defects who are in need of surgical treatment at this time. For the lesser degrees of disability, we recommend temporary postponement of any surgical operation at this time, and periodic re-evaluation of the patient's cardiac status.

For the patient currently in need of surgical treatment and for whom an intracardiac procedure is not available, we recommend an anastomotic operation without the opening of the pericardium. This operation provides significant palliation, and does not interfere with the intracardiac operation should it be deemed necessary subsequently.

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DISCUSSION.—DR. ALFRED BLALOCK, Baltimore, Maryland: I suspect it is a mistake for an old conservative surgeon to discuss this paper. I must say that I never thought I would live to see the day when this type of operative procedure could be performed. I want to commend Dr. Lillehei and Dr. Varco and their associates for their imagination, their courage and their industry.

I don't think they have supplied the ultimate answer in the type of circulation tha tthey used in the film. It is my guess that the ultimate answer will be the artificial heart lung as developed by our President, Dr. Gibbon, and as it is being used now by more and more people.

Dr. Gibbon has used it successfully for intracardiac surgery. I have heard that Dr. Kirklin, of the Mayo Clinic, has now used it successfully in closing several ventricular septal defects.

It may very well be that Dr. Gibbon and others will want to use the azygos flow principle that has been used so successfully by Dr. Lillehei, Dr. Varco and others. One of the things that this work has done has been to accelerate progress in intracardiac surgery. While the artificial heart lung is being further developed, certainly great progress has been made.

I would agree that the direct attack for intracardiac lesion is the ideal one. However, I would like to remind some that the mortality rate at the present time with the anastomotic procedures in the treatment of the tetralogy of Fallot is considerably less than that with the open operative procedure.

For example, we have had two groups of 45 consecutive survivals following anastomotic operations during the last eight or ten years. I must say that when we see some of the people upon whom an anastomosis was performed eight or ten years ago, the results are very gratifying.

The thing that has surprised me most about this contribution by Dr. Lillehei is that he seemingly can close an intraventricular defect where the aorta is overriding this defect. From the many specimens that I saw postmortem, I would not have dreamed that that could have been done, because this aorta straddles the defect, and there is nothing between the defect and the aorta. So, to me, that, perhaps, is the most important part of their contribution.

I would like to ask Dr. Lillehei several questions. I cannot expect him to answer them with great certainty. Dr. Bradford Cannon has commented on one of them, and that is this: Will the defect remain closed? Certainly the intraventricular defect, I think, is less apt to remain closed because of the great pressure there, than is the intra-auricular defect, where the pressure is lower.

In a recent open procedure in our hospital a defect was closed. Cardiac standstill was present. The heart was massaged and the sutures cut out, but this was vigorous massage and that may not take place with the normally beating heart.

I would like to ask another question that I doubt can be answered. Is there some concern about the myocardial scar that results from a big incision such as this? In other words, in subsequent years will the patient have difficulty with the coronary circulation as a result of it?

Another question for Dr. Lillehei is this: In the patient with the very small pulmonary artery, will this small pulmonary artery be able to carry the blood if the ventricular defect is closed?

In closing, I predict that the mortality accompanying the anastomotic procedure in the treatment of the tetralogy of Fallot will remain lower for quite some time than that accompanying the direct attack under vision. It seems likely that the more perfect results will be obtained in those