1948

Tetralogy of fallot

Robert W. Pollack
University of Nebraska Medical Center

THE
TETRALOGY OF FALLOT

Robert . . Pollock

Senior Thesis

Presented to the
College of Medicine
University of Nebraska

Omaha, Nebraska
1948

TABLE OF CONTENTS

INTRODUCTION

Until a few years ago, congenital heart disease had been neglected to a great extent. Previously nothing could be done for these patients. Very little in the form of definite diagnosis was attempted and most of the patients were just labeled as having congenital heart disease and were studied from purely an academic standpoint.

After Doctors Gross and Blalock published their work on the surgical treatment of some of these conditions the importance of the accurate diagnosis of the different types of congenital heart lesions was brought to light. From that time on, great strides have been made in the diagnosis and treatment of some of these conditions.

The congenital cardiac defect known as Tetralogy of Fallot, characterized by pulmonary stenosis, intraventricular septal defect, dextra position of the aorta, and right ventricular hypertrophy, has been known for many years. After Blalock published his first article describing the anasomosis of the subclavian artery to the pulmonary artery for the surgical treatment of this condition, the subject of Tetralogy of Fallot has been of increasing interest and importance in the

field of cardiovascular surgery.

Classification of Congenital Heart Disease.

Maud Abbott's (1) clinical classification of congenital heart disease is at the present time the generally accepted classification. She divided the patients into three groups. The first group includes those patients with no abnormal communications or shunts between the right and left sides of the heart. Cyanosis is not a part of this condition. This group includes patients with single dextracardia, anomalies of the pericardium, primary congenital hypertrophy of the heart, pure subaortic or aortic stenosis, pure mitral stenosis, and coarctation of the aorta. The second group includes patients arteriovenous shunt in whom arterial blood enters the pulmonary circulation. Cyanosis is usually not observed. There is possible terminal or transient reversal flow with cyanosis, due to entrance of venous blood into the systemic circulation. Patients in this second group have defects of the interauricular septum, defects of the intraventricular septum, localized defects of the aortic septum, and patent ductus arteriosus. The third group includes those patients in whom cyanosis is associated with the entrance of venous blood in large quantities into the systemic circulation. There are numerous causes for

this group, among these are; interventricular septal defect with dextra position of the aorta, tricuspid stenosis, tricuspid atresia with septal defects, transposition of the arterial trunks with defects in the ventricular septum, persistant ductus arteriosus and Tetralogy of Fallot.

Advances have been made in the surgical therapy in a representative of each of the three groups described by Abbott; coarctation of the aorta in the acyanotic group, patent ductus arteriosus in the usual acyanotic group but occasionally cyanostic group, and pulmonary stenosis, usually Tetralogy of Fallot, in the cyanotic group.

History.

The tetralogy of congenital cardiac defects now called Tetralogy of Fallot was first described by Sandiford (35) in 1777. William Hunter (17) described a case accurately about the same time. Hope (30) in his treatise on disease of the heart described a congenital heart disease in which there was constriction of the pulmonary valves or orafice or an opening out of the right into the left ventricle or both. He said this was associated with an unusually loud and superficial murmur with the loudest sound immediately over

the semilunar valves (this is about opposite to the inferior margin of the third rib.) When signs of a lesion in the mouth of the right ventricle coincide with cyannis, the evidence of a communication between the two sides of the heart is almost positive, and, as hypertrophy of the right ventricle is usually a concomitant, its presence is a corraboration circumstance.

Fallot's (20) description of the condition was thus not the first time this congenital anomaly was described. Fallot had post-mortums examinations on three hearts that showed the tetralogy. He referred to the thesis of Debeby published in 1878 which described the observations in 20 cases of the maladie bleue collected from the literature, which conformed to the conditions that Fallot found in his three cases, but the true significance of which Debeby missed. The reason for the continued use of the term "Tetralogy of Fallot" lies in the fact that Fallot realized that the lesion could be recognized clinically. He wrote: "Until now, clinicions have considered the precise diagnosis of anatomic lesions of morbus caeruleus (maladie bleue) of almost unsurmountable difficulty, as if it would be pronounced only as a vague and uncertain hypothesis. On the contrary, we see from our observations that cyanosis, especially in the adult is the result of a

small number of cardiac malformations well determined." He went on to enumerate the tetralogy which, under the circumstances, quite appropriately bears his name.

Embryological Defects.

The abnormality described by Fallot (20) consists of a true anatomopathologic type represented by the following tetralogy: "(1) stenosis of the pulmonary artery; (2) interventricular communication; (3) deviation of the aorta to the right; (4) hypertrophy, almost always concentric, of the right ventricle. Failure of the obliteration of the foramen ovale may occasionally be added in a wholly accessory manner".

McGinn and White (36) stated that the dextraposition of the aorta means that the aorta is further to the right than normal and that the position overlies the septal opening.

Other anatomical variations are often present. The aorta is usually wide and thick walled, and as stated before rides over the large defect in the interventricular septum. There is frequently present a narrow muscular channel inferior to the orifice of the pulmonary artery. This channel, though actually a part of the right ventricular chamber, has been called a "third ventricle."

Because of its narrow chamber, the so-called third ventricle, when present, contributes materially to the interference with the flow of blood to the lungs. Another frequently associated lesion is a biscuspid pulmonary valve. The leaflets are frequently fleshy in character and the conus is narrow and deformed or contracted at its lower bulbar orifice. The valve cusps, instead of attaching more or less directly to the wall of the pulmonary artery, as they do in the normal condition, attach to a pair of mounds of tissue which protrude from the intima of the pulmonary artery into the luman of the artery. This presents the full lateral excursion of the pulmonary cusps during systole. Thus the bicuspid valves tend to increase the degree of pulmonary stenosis.

Theories of Development.

Fallot (1) at the time of his original writing gave his interpretation as to the cause of the condition. He stated that "in persons with morbus caeruleus, the incompletely developed septum cannot by any means be considered as the analogy of the false septum of vertebrates to communicating ventricles. It seems to be more logical and more in conformity with the laws of physiology to regard the whole series of cardiac changes

enumerated as wholly the result of pulmonary stenosis. As to the cause of this, it is believed that it must not be attributed to a simple arresting of development but rather to a pathologic process developing during intrauterine life of the level of the pulmonary valves and of the region of the infundibulum which is contiguous to them."

The views of the earliest observers who believed that fetal endocorditis with subsequent stenosis of the pulmonary orifice and increased pressure in the right ventricle was the cause of the anomalies, has been abandoned. The pulmonary stenosis, interventricular septal defect and the dextra position of the aorta according to Abbott (1) is highly suggestive of the relationship that would result from the uncovering of the right reptilian aorta and obliteration of the left in the delayed torsion of Spitzer's theory (his type I and II of transposition). "Spitzer's theory ascribes such anomalies to the arrest or delay of the clockwise torsion that usually takes place in the growth of the primitive heart, between its fixed arterial and venous ends, during the process of septation, thus leading to an apparently counter-clockwise shunting of the parts with resultant reopening of the channel of the reptilian right aorta, and obliteration of the left ventricle

vessels. Anatomical proof, of the latter curious fact, is supplied by the confirmation of the right ventricle in which the trabecula septomarginalis or moderatos band is homologous (as described by Tendler) with the septum between the right aorta and pulmonary artery in the reptilian heart, that portion of the ventricle lying between the "anterior tricuspid ledge" of the latter and the crista supraventricularis being identified as the "outflow channel" of the right reptilian aorta, which is closed in the normal human heart by a clockwise tortion. An additional, very suggestive point, is seen in the persistence of the bicuspid pulmonary valve (normal in reptiles) and the development from pulmonary stenosis."

Herndon, Voss and Donovan (29) stated that they doubted the theory of Spitzer, and they were inclined to believe Lev and Saphir (33), who thought that the essential process in the development of the Tetralogy of Fallot is an abnormal development of the bulbo auricular spur which presents usual absorption of the arterial bulb into the left ventricle and thus a sufficient degree of normal clockwise rotation around this point. As a result counter rotation of the unfixed lower end of the arterial bulb takes place, causing a more or less parallel instead of a spirally

twisted position of the aorta and pulmonary artery, and imperfect union or nonunion between the lower end of the aortic-pulmonary septum and ventricular septum. They explain the pulmonary stenosis by the presence of a hypertrophic and rotated septal muscle bundle.

Edwards, Bulbulian and Milton (18) thought that the information derived from a review of the transformation in the embryo, in the truncus arterisus and in the conus arteriosus communis would help to explain the mechanism of the formation of the defects to form the Tetralogy of Fallot. The truncus arteriosus is the relatively early arterial vessel leaving the heart, while the conus, is the conically shaped part of the heart which joins the ventricle of the young embryo and the truncus arteriosus. The conus and the truncus together are a continuous channel and for practical purposes may be considered as two superimposed portions of a single tube. The line of division lies at the level of the pulmonary and the aortic valves, the truncus lying cephalad and the conus caudad to this level. In the lining of the truncus-conus channel, two opposed and spiralled rows of swellings, the truncus conus ridges, develop. As the result of growth, these two ridges meet in the central axis of the channel and fuse to form a septum. This septum divides the truncus

arteriosus into two vessels, the pulmonary artery and the ascending aorta, which are about equal size. The basic abnormality in the Tetralogy of Fallot seems to lie in the truncus conus ridges. Instead of being lined up in a celertrodorsal to sinistroventro plane, in the upper part of the truncus, which is normal, the ridges line up dorsoventrally and at the same time, are eccentrically placed. Also Edwards, et al. (18) says there seems to be only a minimal degree of spiralling of the ridges and the septum formed by their fusion. This would account for the narrow pulmonary artery and the wide aorta. The dextra position of the aorta would be the result of the minimal degree of spiralling of the truncus-conus ridges.

They say the explanation for the bicuspid pulmonary valve lies in the truncus-conus ridges being excentrically placed. Thus the resulting septum formed by the fusion of the opposing ridges are smaller than normal. The resulting small septum contributes but one pulmonary cusp instead of the normal two, the lateral wall contributes its normal one cusp. Thus results the bicuspid valve.

Also because of the basic misplacement of the truncus-conus ridges, the septum resulting from their fusion is not in line with the upper portion of the

muscular interventricular septum and is thus not in position to make its important contribution to the fomating of the membranous portion of the intraventricular septum. The result is a defect in this portion of the septum.

Basis of the Cyanosis.

Lundsgard and Van Slyke (35) have shown that there are four important factors in producing cyanosis; (1) the total hemoglobin content; (2) the proportion of blood passing from right heart to left through unareated channels; (3) the rate of utilization of oxygen by the peripheral tissue; (4) the extent of areation of the blood in the lungs. The importance of the absolute amount of reduced hemoglobin present in the capillary blood, not the percentage of desaturation of the hemoglobin in the causation of cyanosis was first stressed by these men. It has become common teaching to state that cyanosis only appears when the mean value of the reduced hemoglobin in the capillaries was 5 gm per 100 cc of blood. Buchell (13) states that such a general and still useful rule was soon found to have some exceptions. It has become apparent that the degree of cyanosis is related also to the increased amount and distribution of the hemoglobin pigments in the dermal capillaries

and venules. Lundsgard and Van Sylke have also shown the great importance of the pulmonary factor in Tetralogy of Fallot. The oxygenation of the blood in the lungs depends on the vital capacity of the individual, the rate of flow blood through the lungs, the partial pressure of oxygen in the inspired air and also special pulmonary factors. These investigations showed that in most, if not all cases, in which there was pronounced polycythemia, secondary changes occurred in the lungs in such a way that all the blood passing through the lungs was no longer in effective contact with the oxygen in the alveoli. They thought this could be easily demonstrated because in almost all cases in which there is polycythemia, cyanosis can be greatly lessened by the prolonged inhalation of oxygen. The fact that all the blood which circulated through the lungs is not fully oxygenated made it seem improbable that if more blood circulated through the lungs a larger proportion of the blood would be oxygenated.

Blalock (5) through his operation of anastomosing the subclavian artery to the pulmonary artery has shown, that increasing the blood supply to the lungs does help the blood to be more fully oxygenated and the return to more normal conditions and decreasing or completely stopping the cyanosis.

Dautrebande, Marshall and Meakins (16) in 1929 found it impossible to obtain reliable determinations of the venous or right to left shunt of the blood. Bing and Vondam (3) in 1847 demonstrated in 36 out of 48 patients studied; the flow of blood through the pulmonary artery per square meter of body surface was below 3 liters per minute, which is normal cardiac index. The systemic flow on the other hand calculated the same way showed marked variations above and below the values of the normal cardiac index. They showed by comparison of the data obtained for systemic flow, with that for the pulmonary artery, that the systemic exceeded the pulmonary artery flow by from 0 to 9.7 liters per minute. This indicated that in Tetralogy of Fallot some of the returning mixed venous blood, unable to pass through the stenosed pulmonary artery into the lungs, coursed through the intraventricular septal defect and the overriding aorta, directly into the systemic circulation. Consequently the overall direction of the intracardiac shunt was from right to left. Gibson (23) estimated that when 30% or more of the blood sent into the systemic circulation is venous blood there will be cyanosis. Segall (50) in his patient considered the venous-arterial shunt to be 40% of the total cardiac output, into the aorta. There is, however, some concomitant flow from

left to right through the septal defect. This is indicated by the observation that the oxygen content of the right ventricular blood exceeded that of the right auricle, in the majority of cases.

The anatomical defects in Tetralogy of Fallot are structurally fixed. Compensatory processes designed to maintain oxygen supply to the tissue are present. These are the increased capacity of the blood to carry oxygen, made possible by the occurrence of polycythemia and an increase in the volume and the rate of flow in the systemic circulation. By this latter mechanism, Burchell (13), there will be a certain similarity in the oxygen tention of the arterial blood in the patients, with Tetralogy of Fallot at rest, and the value for the oxygenation of arterial blood will not necessarily correlate with the degree of the anatomical defect. With exercise in these patients, there is an abrupt drop in the degree of saturation of the hemoglobin in arterial blood due to increased peripheral utilization. Certain limitations of Burchell's instruments precluded the exact quantitative analysis of this. This explains the long known clinical observation, that exercise may increase the cyanosis in congenital heart disease. In the normal person there is no decrease under conditions of exercise.

A summary of the causes of cyanosis in Tetralogy of

Fallot shows that many interrelated physiologic variables must be considered. The variables are; First, the central factors, (a) the proportion of arterial to venous blood being mixed in the aorta, (b) the degree of oxygen unsaturation of the venous component. Second, the peripheral factors, (a) the size of the capillary and venous vascular beds in and under the skin, (b) the rate of flow of the blood through these vascular beds. Third, the degree of polycythemia. Fourth, the nature of the dermal area viewed. Fifth, the source of light that is used.

Incidence.

Fallot (20) himself pointed out that the Tetralogy of Fallot was by far the commonest type of pulmonary stenosis, and Maud Abbott (1) found it present in 77 percent of 110 cases of pulmonary stenosis, and in 66 per cent of 40 cases of pulmonary atresia or 115 cases of her 1,000 cases of congenital anomalies studied, were Tetralogy of Fallot. Leech (32) reported on 13,115 post mortums at John Hopkins Hospital and found 120 cases or 1.29% of true congenital anomalies. Volini (55) reported 99 cases or 1 percent of the 9,500 autopsies showed congenital heart defects, at Cook County Hospital. He found only one case of Tetralogy of Fallot, in this entire series. Clawson (14) reported on 15,597 autopsies

performed in the University of Minnesota 141 cases of congenital heart deformities, were found. Of these 18 were still births, 83 died the first 5 months of life, 111 or (78.7%) were dead or died before the first year of life. There were 9 cases of Tetralogy of Fallot, or 6.3%, one still birth, 7 died in the first decade and one in the third.

Diagnosis.

The raised pressure, in the right ventricle behind the pulmonary artery, sends a large venous-arterial shunt into the dextraposed aorta, through the interventricular septal defect. The effect of this in the circulation is intensified by increased deoxygenation at the pulmonary orifice. In addition, long standing capillary changes and the effect of the polycythemia and great hypertrophy of the right ventricle, that develope as compensatory features, produce the symptomatology of morbus coeruleus even seen in adult life.

The history of patients with Tetralogy of Fallot is usually quite typical but varies with amount of stenosis and interventricular shunt of the blood. The symptoms are frequently absent in the first weeks or months of life and when it first appears it may only occur in transitional attacks. The patient complains of attacks

of dyspnea and there is usually marked limitation of activity.

Fashena (21) said, that frequently it can be obtained from the history that when the patients are deeply cyanotic they lie on their chests with their knees drawn up under them to make breathing easier. Also, after walking a few steps many of the patients will assume a squatting position with their head down between their knees until the dyspnea passes. The physiology of this maneuver is obscure, but many of the older patients, when questioned, report that it makes them feel better and tends to help the "all gone" feeling that they often experience.

On physical examination, there are many characteristic signs. On inspection there is usually signs of cyanosis of lips, cheeks, ears, fingers and toes. There is also clubbing of the fingers and toes. The oral manifestations are quite prominent. The bucal nuscosa shows a pronounced wrinkling, this is possibly due to a transient edema according to Kaver, Lord and Green (27). There is severe marginal gingivitis and bleeding is produced on very slight pressure. The teeth of the children with Tetralogy of Fallot are normal in size and shape but there is usually a delay in the eruption time of both the deciduous and permanent

teeth. Also the incidence of cavities is markedly elevated.

A loud systolic murmur is heard best at the pulmonary valve area and in the third left interspace, (at times it is accompanied by a systolic thrill). Fashena (20) states, that in approximately 5 per cent of the patients a systolic murmur is absent. Garb (22) also, says, that with high blood viscosity, the absence of a murmur does not rule out gross structural cardiac abnormalities. Furthermore, the presence of a murmur in polycythemia probably indicates a much more extensive abnormality than it would if the blood picture were normal.

The blood picture of the patient with Tetralogy of Fallot is quite a variation from the normal. The red cell count is greatly increased, it varies from 7 to 12 million. The hemoglobin also is greatly increased and ranges from 18 to 26 gm. The hemocrib readings may be in the eighties.

Talbotts (52) showed that in his metabolic studies of a patient with Tetralogy of Fallot there are profound variations from normal, in acid-base equilibrium of the body and changes in renal function. His oxygen saturation of the blood varied between 62 and 58 percent. The oxygen capacity was about 35 vol. percent, the carbon dioxide capacity was about 33 vol. percent.

The arterial pH was less than 7.20.

Bing, Verdon and Gray (3) stated that the oxygen consumption was reduced and the B.M.R. was low. Their lowest was -48. They also showed that in normal individuals the standard exercise (stepping up and down a step 20 cm high 30 times in one minute) resulted in a significant increase in the oxygen consumed and a slight rise in the carbon dioxide produced, whereas in the Tetralogy of Fallot the oxygen consumed and the carbon dioxide produced both fell below their resting values.

On X-ray examination of the chest, the findings are: (1) absence of fullness of the normal pulmonary conus; (2) the shadow at the back of the heart to the left of the stermum, is concave and not convex; concave shadow in this region, in cases of persistent cyanosis always means that the pulmonary aorta is misplaced, absent or deminished in size; (3) in the left anterior oblique position the pulmonary window is abnormally clear; (4) the heart frequently is normal in size but sometimes is moderately enlarged, due to right ventricular hypertrophy. As a result of this, the heart is enlarged to the left and the apex is lifted from the diaphragm and has a blunt appearance. This is the "coeuren sabot" silhouette. (5) the dextra

position of the aorta can be seen in conventional x-rays but is not a very reliable observation. In many cases dextra position of the aorta is not suggested by the roentgenogram, (6) masking of the aorta by the trachea in the right oblique position. If stenosis is solely of the valve, there will be usually noted an enlarged pulmonary arc.

E K G shows evidence of marked right axis deviation, usually accompanied by accentuation of the P waves, and occasionally an inverted T wave.

Robb and Steinberg's (49) method of rapid injection of 25 to 45 cc of a 70 percent solution of diodrast into the arm vein and making x-rays of the heart has made it possible to visualize the chambers of the heart and the great vessels. The interval, between the injection and the exposure, depends on the region to be visualized. Through this procedure, the stenosed portion of the pulmonary artery can be seen and also the enlarged and right ventricle. Grishman, Steinberg and Sussman (26) say, that fluorographic multiple exposure technique is essential for successful visualization of all the features to be considered. Through this visualization they can help make the diagnosis of Tetralogy of Fallot.

In the case of the patient with Tetralogy of Fallot, McGuide, and Goldman (37) have shown that the circulation

time from the arm to the tongue is greatly shortened. In these cases the stimulating time does not have to go through the pulmonary circulation before it reaches the tongue so it gets to the tongue much quicker by way of the interventricular shunt. They have reported circulation times of 4 seconds, in cases of congenital heart disease with venous arterial shunt. They found that the average circulation time for normal children was 10.6 sec.

McGinn and White (36) say that there are five findings which when they occur in association permit the diagnosis of the Tetralogy of Fallot; (1) cyanosis of lips, toes, fingers, etc.; (2) clubbing of fingers and toes; (3) loud systolic murmur heard over third left interspace; (4) marked right axis deviation on E K G; (5) x-ray evidence of the sabor-shaped heart, due to enlargement of the right ventricle without enlargement of the pulmonary artery.

Differential Diagnosis.

The differential diagnosis of Tetralogy of Fallot should include all types of congenital heart disease associated with cyanosis. It is possible to eliminate from consideration all defects with which cyanosis is not associated, except as a terminal event or in the presence of heart failure; this group includes un-

complicated patent ductus arteriosus, simple auricular septal defect and uncomplicated ventricular septal defect.

The differential diagnosis is made easier the older the patient is, because the factor of age alone may serve to exclude the highly complicated primitive defects which are most difficult to diagnose but are usually incompatible with life beyond a few weeks or months.

The majority of the children who have congenital heart disease with persistant cyanosis and polycythemia, are examples of the Tetralogy of Fallot. These are the conditions which must be taken into consideration when a differential diagnosis is made, particularly if the question of surgery is in consideration.

The Eisenmenger complex, consists of a ventricular septal defect, with overriding of the aorta, so that a portion of the right ventricular or venous blood enters directly into the aorta. The degree of cyanosis, therefore, tends to be less intense than it is in Tetralogy of Fallot and it is likely to have its onset later in life. Likewise, the degree of oxygen saturation of the patient, in arterial blood although always subnormal, does not drop so precipitously on exercise as it does in the presence of a lesion associated with pulmonary

stenosis. The auscultatory findings are not notably different from those found in Tetralogy of Fallot. A nondiagnostic systolic murmur is usually present but one is likely to hear a clear second pulmonic tone which is not a feature of Tetralogy of Fallot. In contrast to the roentgenologic findings in Tetralogy of Fallot, a large pulmonary conus shadow, with exaggerated pulmonary shadows and pulsations in the hili of the lungs, is usually seen.

It is significant, that in several individuals with Tetralogy of Fallot, extensive collateral circulation has been responsible for the erroneous clinical diagnosis of Eisenmengers complex, because of pulsations in the hili of the lungs on fluoroscopic examination due to the collateral circulation.

One must also consider the possibility of a persistent truncus arteriosus. This defect is always associated with a defect in the ventricular septum. Cyanosis may be minimal or even absent. A loud systolic murmur with a thrill, usually is encountered in the presence of this lesion and the murmur may be transmitted widely over the base of the heart and into the cardiac vessel. There is both right and left ventricular enlargement, as well as increased vascular hilar pulsations.

Complete transposition of the great vessels, in

which both the aorta and the pulmonary artery are of adequate proportions, may be consistent with life beyond the period of infancy. In a majority of cases death occurs in early infancy. The heart, in complete transpositions, increases in size rapidly and has a peculiar globular configuration. There is a reversal of position of the aorta and the pulmonary artery, the aorta lying anterior to the pulmonary artery.

Cor triloculore biatriatum is the condition of complete absence of the ventricular septum. There is complete mixture of venous and arterial blood in the single ventricle and cyanosis is a prominent feature. This condition is most frequently encountered in infants and only rarely in children or young adults. On E K G examination the pattern of large biphasic Q.R.S. complexes in all leads is noted in the presence of this type of lesion.

Other congenital anomalies associated with severe cyanosis are even more difficult to recognize clinically. Atresia of the aortic orifice, atresia of the mitral or tricuspid orifice, cor bilocular and persistence of the atrioventricular ostium commune are rare types of lesions which, owning to the severe derangement of the circulation, rarely present a diagnostic problem, except in the infant. The clinical picture of a child

3 years of age or older is usually characteristic and should be recognized readily. The roentgenologic examination of the heart serves as the best adjunct in determining whether or not the defect in question is one associated with pulmonary stenosis.

Prognosis.

The duration of life in patients with Tetralogy of Fallot depends upon the amount of right-to-left shunt in one hand and erythrocytosis on the other. The amount of right-to-left shunt is determined by the mechanical factors in the heart, the size of the pulmonary vein orifice, the size of the intraventricular septal defect, and the relations between the pressure developed in the right and left ventricles. Under severe intracardiac conditions the efficiency of the myocardium plays the center role in determining the duration of life, because upon it depends the maintainence of the circulatory rate and the restriction of the right-to-left shunt to within physiological limits.

The average age of death in 85 cases reported by Maud Abbott (1) was 12½ years. However, the outlook may not always be so discouraging. The oldest patient in Fallot's original series was 36 years of age. White and Sprague (57) reported a man who died in his

sixtieth year after leading a useful and active life. McGinn and White (36) mentioned a patient who was still leading an active life at the age of 62.

It has been generally supposed that death is caused by the development of bacterial endocarditis. Perlman and Meyers (42) believe that this is not true and state that only one case in Abbott's series had bacterial endocarditis and one of Fallot's patients died of terminal endocarditis and, Pescatose, Wolffe and Dizilio's patient died of septic endocarditis. The remainder of cases of Tetralogy of Fallot which have been reported, died of causes not related to the cardiac defect.

Since Blalock (11) started his surgical treatment of cases of Tetralogy of Fallot in 1945, the prognosis for these people is much better. Adequate follow up of these patients has not been possible because of the short time since the first operation was performed but the outlook for these patients in life expectancy and functional capacity is greatly increased. Without operation, Taussig (54) states the chance of survival to puberty is only 50 per cent. Cerebral thrombosis is frequently the complicating condition.

Treatment.

Congenital defects are not rare in infancy and it

is only because of the high mortality rate that the incidence after the age of 12 years is low. The complex nature of the defect which adds to the difficulty of diagnosis particularly in infants and the lack of successful therapeutic methods resulted for years of a general neglect of this field. One should be able to diagnose the particular defects which are present. The possibility of surgical therapy for certain types of congenital heart defects makes it all important that these types be recognized. The advances in this field are recent as shown by White's (56) statement in 1937, he said "there is no curative treatment surgical or medical, for congenital cardiac defects."

The first attempt at curative treatment was done by Doyen (5) in 1913. The patient was 20 years of age and had a congenital pulmonic stenosis. He introduced a small tenotone into the right ventricle and an attempt was made to divide what was thought to be a stenotic valve. The patient died several hours later and examination showed narrowing of the conus rather than stenosis of the valve. There were no other curative attempts in the treatment of Tetralogy of Fallot until Blalock (11) reported his operation in 1945.

The treatment of the Tetralogy of Fallot surgically was the second congenital heart disease to be successfully

attacked; the first being the patent ductus arteriosus by Gross (24) 1939.

Blalock and Taussig (11) thought that the importance of the diminution of flow of blood to the lungs had not been fully appreciated mainly, because studies on cyanosis had been made on older children and young adults. They thought that it was only when these factors were not of vital importance that the individual survived to that age. They went on to say that all infants, with pulmonary atresia in whom the closure of the ductus arteriosus cuts off the circulation to the lungs, die at an early age. In all cases of complete pulmonary stenosis, death occurs before complete cessation of circulation of blood through the lungs, thus in such patients there is always slight patency of the ductus arteriosus. In cases of Tetralogy of Fallot with an extreme pulmonary stenosis, the ductus arteriosus may become entirely obliterated before death.

From their observation, it was indicated that many gross malformations of the heart were compatible with life provided there is adequate circulation to the lungs, and further that lack of circulation to the lungs, is the primary cause of death in many of these children. Also Taussig (11) has seen many infants with Tetralogy of Fallot, in whom the cyanosis was not apparent until the ductus arteriosus closed. It was the appreciation

of these facts and the previous experience with the experimental use of large arteries for the purpose of conducting blood to sites not usually supplied by such vessels, that led them to the development of the Blalock operation.

The feasibility of anastomosing a systemic artery to one of the pulmonary arteries in experimental animals had been demonstrated by Levy and Blalock (34); Eppinger; Burwell; and Gross (19) and Leeds (32).

Before the operation was performed on patients, experimental studies were carried out on dogs to determine if the creation of an artificial ductus arteriosus would be helpful in treatment of patients with Tetralogy of Fallot. Attempts to produce the desired degree of pulmonary stenosis in the dogs were unsuccessful. They tried constricting ligatures of silk, rubber tubes, bands of ox fascia and large silver Goldblatt clamps. Unilateral pneumonectomy and anastomosing the pulmonary artery and vein, produced marked cyanosis but was unsatisfactory for testing in artificial ductus arteriosus. The removal of lobes of one or both lungs and the anastomosis of the severed proximal ends of the pulmonary artery and vein usually results in a high degree of arterial oxygen unsaturation. The creation of an artificial ductus arteriosus under these conditions

usually resulted in an increase in the oxygen saturation of the arterial blood. This increase was due to the passage of a greater quantity of blood through the pulmonary circulation. Thus, a greater number of red corpuscles was exposed to the pulmonary circulation. Although this is much different from the condition in Tetralogy of Fallot, the results of these studies strengthened the impression that the patient would be improved if the pulmonary blood supply was increased.

Before performing the operation on a patient, Blalock and Taussig (11) had many doubts as to how the patient would stand the operation. They feared that an intensely cyanotic child would not stand the operation which necessitated opening the plural cavity and the temporary occlusion of one of the pulmonary arteries. They also were worried about what the effect of the ligation of the subclavian or innominate would be on the arm or the brain. With the polycythemia, the increased blood viscosity and chances of thrombosis gave them much concern. The possibility of post operative endocarditis was also a factor to be reckoned with. For these reasons, their first clinical attempt to increase the circulation to the lungs was postponed almost a year after they decided the procedure was sound.

Up to June, 1947, Blalock (7) has performed 450 of these operations and the previous pre-operative doubts are now proved to have been of no reason for concern. From their experience, it appears that this type of patient can tolerate the use of inhalation agents for general anesthesia. Open drop ether and cyclopropane with high concentration of oxygen have been used and this apparently increased the oxygen content of the arterial blood because cyanosis was definitely decreased. Whether the patient would tolerate the occlusion of one of the main pulmonary arteries for the period during which the anastomosis was being made, a time of about 30 to 90 minutes being necessary was questioned. They stated that the cyanosis did not seem to be greatly affected by the clamping of this artery. This is possibly explained by the fact that the other lung is able to utilize the excess blood almost as effectively as could the two lungs.

The effect of the ligation of the subclavian or the innominant on the arm or the brain was first thought to be insignificant. The pulse is absent for some time post operatively and the part was slightly colder than that of the opposite, but immediately after operation it was evident that its circulation was adequate to maintain life of that part. The innominant is, however,

not used unless absolutely necessary because of the possibility of cerebral complications.

With the polycythemia and the increased blood viscosity, the chances for thrombosis was greatly increased, so they decided at first to use dicoumarol in small quantities, starting 24 hours after the operation. This was regulated to keep the clotting time about two weeks. This procedure was later discontinued when a patient died from interpulmonary bleeding. Heparin is now used and only if the patient presents evidence of weakness of the side opposite the operation.

Blalock (7) in 1946 reported that up to that time none of his patients had developed bacterial endocarditis.

Taussig (53) does the diagonistic work for Blalock and she states that the operation on the patient with Tetralogy of Fallot is indicated only in cases in which the primary difficulty is lack of circulation to the lungs. It is possible, only provided that there is a pulmonary artery to which to anastomose the systemic artery, that the pressure in the systemic artery is higher than the pulmonary artery and finally, that the size and the structures of the heart are such that it can tolerate the altered circulation. The heart should not be enlarged. There should be no fullness in the pulmonary conus and no pulsations in the pulmonary fields.

Usually there is an absence of hilar shadows. However, in patients with extensive collateral circulation these shadows may become pronounced, but they are composed of an aggregate of fine shadows and never show expansile pulsations.

The factors influencing selection of patients are, (1) the age and size of the patient, (2) the severity of the anoxemia and (3) the height of the red blood count, the hemoglobin content and the hematocrit reading.

The optimum age is between 3 and 7 years. Blalock's (9) youngest patient to be operated successfully was 5 months and the oldest 26 years. Patients under 18 months of age should not be operated if they will live to a greater age. There are several reasons for this limitation: (1) one is less certain of the correctness of the diagnosis, (2) the mortality rate accompanying operation is higher in this age group, and (3) it is frequently necessary in infants to utilize the innominant or the carotid artery rather than the subclavian and this adds to the danger of the operation.

Taussig (53) also says, that if the patient is in danger of dying of anoxemia, early operation is clearly indicated. Attacks of dyspnea are common and are often severe when the patient is between 4 and 18 months of age. Prolonged syncope is an indication for operation.

The degree of incapacity varies greatly. With an oxygen saturation of the arterial blood of 30 per cent most children can walk only a few feet. With an oxygen saturation of 20-25 percent few can walk at all. When the saturation is below 20 percent there is cause for great concern.

The height of the red blood cell count, the hemoglobin content and the hematocrit readings are important factors in selection of cases. If a patient has a red blood cell count of 8,000,000 or 10,000,000 or a hematocrit reading of 70-80 mm. he is in great danger of cerebral thrombosis. For such children operation is indicated.

On the average, the pulmonary arterial pressure taken at the time of surgery is between 150 and 200 mm. of water; Blalock (8) says when the pressure is over 350 mm. of water, the anastomosis is not advisable and should not be performed unless other evidence points strongly toward the presence of significant reduction in the pulmonary blood flow.

There are several conditions associated with persistant cyanosis which would not be benefited by the operation. These are the Eisenmenger complex, complete transposition of the great vessels, and aortic atresia, and should be watched for carefully.

Due to the tendency of these patients to thrombosis and because dehydration increases this tendency, prior to operation, Blalock (5), (6) and (11) says they should receive one or two quarts of liquid a day.

Penicillin is given one or two days preoperatively and is continued several days following the operation.

Blood and plasma should be present at the time of operation so that they can be given if the loss of blood is excessive. A slow continuous drip of glucose or saline, or both, is advisable during the operation so that if necessary a large quantity of fluid can be given on a moments notice. A leg vein is used for this proceedure.

Since the operation was devised to compensate for an inadequate flow of blood to the lungs, it seemed advisable to Blalock (11) that the anastomosis be made in such a manner that the blood from the systemic artery would be able to reach both lungs. It was obvious that a suture anastomosis could not be made to the main pulmonary artery since occlusion of this vessel for more than a few minutes causes death. Therefore, the anastomosis is made just distal to the division of the main pulmonary artery and furthermore, the side of the chosen vessel should be used in order that the blood might flow to both lungs.

In the Blalock (9) operation, the incision is made from the lateral margin of the sternum to the midaxillary line. Usually the incision is beneath the breast, (3rd interspace). The present plan is, that the incision is made on the side opposite to which the aorta descends. The reason for this is, that it is desireable to gain access to the innominate and its branches and this vessel lies on the left when the aorta descends on the right and on the right when the aorta descends on the left. Twenty per cent of the patients have a right aortic arch.

The innominate, subclavian or the carotid may be used for the anastomosis, depending on the size of the vessel desired. Since the subclavian vessel is usually chosen, the fact that the subclavian branch of the innominate makes a much better angle with its parent vessel following the anastomosis, than the subclavian, which arises from the aorta, makes it important that the operation be done on the opposite side to the aorta.

Blalock (11) by June 1947 had 38 right arches, and Taussig diagnosed correctly the position of the aorta prior to the operation in all of these cases.

The exposure is usually more difficult on the right than on the left. It is necessary to free an adequate length of the pulmonary vessel and there is no

hesitation in opening the pericardium to do this. In the majority of the cases, however, the pericardial cavity has not been entered. After exposure of an adequate length of the pulmonary artery, the pressure within the vessel is usually measured by the use of a needle and a water manometer. If the vessel is small and the pressure within it is obviously low, this test is omitted for fear of damaging the artery. The average reading was between 150 and 200 mm. If the pressure is greater than 350 mm., it is believed that the anastomosis should not be performed.

In an occasional patient, the subclavian artery is so short that an anastomosis is difficult or impossible. This difficulty may sometimes be overcome by occluding both the innominate and carotid arteries temporarily with arterial clamps and removing the clamp from the subclavian. This leaves the entire length of this latter vessel free for anastomosis. However, even this maneuver may not give an adequate length of vessel. It may be necessary to ligate and divide the subclavian and ligate and divide the common carotid artery as high as possible and to use the proximal end of the carotid for the anastomosis. It shoud be emphasized, however, that the carotid should be used only when the employment of the subclavian is impossible, for it ex-

poses the patient to the danger of a cerebral complication. As a matter of fact, the most easily performed anastomosis is usually that in which the aorta descends on the right and the incision is made on the left. This is due to the fact that a greater length of the pulmonary artery is more easily accessible on the left than on the right. Furthermore early branching of the left pulmonary artery is less likely to be encountered.

In every patient that Blalock (5) has exposed there has been a branch of the arch of the aorta which could be used for the anastomosis. Such branches have not always been of the ideal size or length, but at least one or another could be used.

In most children over two years of age, the subclavian artery is large enough to transmit large quantities of blood and is the vessel of choice. Preoperative studies will have given a good idea as to the size of the artery that is required. Sometimes Blalock (5) says the artery may be smaller or larger than was anticipated or the length of the vessel may make it necessary to change the choice of the vessel during the operation.

There are a number of reasons why an end to side anastomosis is preferred to an end to end one, even though the former is more difficult. Among these are:

(1) an end-to-side anastomosis allows the shunted blood to pass to both lungs; (2) the size of the opening into the side of the pulmonary artery may be made to correspond very closely in size to that of the end of the systemic artery, whereas there may be much discrepancy in an end-to-end anastomosis; (3) if following an end-to-end anastomosis in a patient who has atresia or marked stenosis, a collapse of the lung developes on the side of the operation, the limited flow of blood through the opposite lung may not be sufficient to maintain life; and (4) if an end-to-side anastomosis is performed and if later in life the pulmonary flow appears to be inadequate, the operative proceedure may be repeated on the opposite side, whereas it may not be possible to do this if an end-to-end anastomosis has been performed. If one of the pulmonary arteries is divided and an end-to-end anastomosis is made, it is doubtful whether the patient would tolerate occlusion of the opposite artery for the length of time that is necessary for the accomplishment of an anastomosis.

Despite these objections to an end-to-end anastomosis, it must be said that very dramatic results have followed the use of this method in most cases in which it has been used. Points in its favor are: (1) it may be performed more easily and quickly;

(2) it may be used when there is a very short pulmonary artery and an end-to-side anastomosis is difficult or impossible; and (3) traction on the mediastinal contents is lessened. All things considered, however, the end-to-side anastomosis is preferred and has been used in most operations.

After both arteries to be used in the anastomosis have been disected free, the right or left pulmonary artery is occluded proximally with a mechanical device, the tips of which are covered with rubber. This instrument has a long handle which makes it useful in stablizing the mediastinum and in approximating the pulmonary artery to the systemic vessel. During the first operations a Bulldog clamp was used. Distal occlusion of the pulmonary artery is produced by traction on pieces of braided silk which have been placed around the branches of the pulmonary artery. Bulldog clamps were first used but the present method leaves a greater length of artery free for the anastomosis. If the artery is unusually long, Bulldog arterial clamps are used. After occlusion has been produced, a transverse opening is usually made in the pulmonary artery between the points of constriction. This opening is made slightly bigger than the circumference of the end of the systemic vessel. Some of the adventitia surrounding the

opening in the pulmonary artery is excised, but the pulmonary artery should not be weakened too much by removal of the adventitia around the entire circumference. If the pulmonary artery is very much smaller than usual and if an end-to-side anastomosis is to be attempted, the opening in the artery should be in a longitudinal rather than a transverse direction.

After the incision in the pulmonary artery is made the anastomosis is begun. 00000 silk on small round needles (Deknatel) is used. The suture is essentially a continuous one in which intima is approximated to intima. The suture includes the entire thickness of the wall of the artery. An attempt is made to place the sutures 1-2 mm. apart and approximately 2/3 of the posterior row is placed before the suture is drawn tight. Each end of this running suture is then tied to interrupted sutures which are placed at appropriate points. This continuous suture is begun again and the posterior row is completed, at which point another interrupted suture is placed. The anterior row of suture is easier to place. It is usually interrupted at one additional point. The suture around the entire circumference of the anastomosis is in reality a single row of a continuous out-in and-over suture which everts the intima and which is interrupted in several places in order not

to constrict the luman of the opening.

After completion of the anastomosis, the traction on the braided silk around the branches of the pulmonary artery is released and the instrument which has been occluding the pulmonary artery proximally is removed. If a spurting stream of blood comes from some point along the suture line, an additional suture is placed. If there is simply a slow ooze of blood, slight pressure is made and it will usually stop without the use of further sutures. Several minutes after all bleeding ceases, the arterial clamp is removed from the systemic vessel. Usually no further bleeding occurs. If there is bleeding, the arterial clamp is replaced and the suture line is inspected again.

After the anastomosis is complete and the constricting devices have been removed, a continuous thrill is usually felt in the pulmonary artery. In many cases this can also be felt in the lung tissue at a considerable distance from the hilus. In infants and even in older children you may not feel a satisfactory thrill if the systemic arterial pressure is low.

After every effort has been made to be certain that the anastomosis is patent, including gentle massage of the opening, the mediastinal pleura is closed with interrupted sutures. The retractors are removed and the

lung is partly inflated. Usually at this time the color of the patient is definitely improved. The 3rd and 4th ribs are approximated. The tip of a catheter is placed temporarily in the pleural cavity. The muscle, fascia, subcutaneous tissues, and skin are approximated with interrupted sutures. Aspiration is carried out and the catheter is removed.

After the operation Blalock (5) places the patient in an oxygen tent. The length of time that the administration of the oxygen is continued varies from a few hours to a number of days.

If the patient has a pronounced polycythemia and the loss of blood during the operation is minimal, whole blood equal to one percent of the body weight is removed at the conclusion of the operation.

The intervenous needle that was placed in the leg is left in place until the patient begins to take adequate fluid by mouth. The patient should be given adequate but not excessive fluids. If the blood pressure should go down, plasma or whole blood should be immediately given because cerebral thrombosis or closure of the anastomosis may occur.

Heparin is given if the patient shows evidence of weakness of the side of the body opposite that of the operation.

The patient is observed carefully for evidence of accumulation of air or blood in the pleural cavity and aspiration is performed if it is deemed necessary.

If evidence of heart failure appears, digitalis is given.

The penicillin that was begun preoperatively is continued two weeks.

The anastomosis makes it possible for a greater volume of oxygenated blood to return by way of the pulmonary veins to the left ventricle. Consequently, a greater volume of oxygenated blood is pushed into the pulmonary circulation as a result of the aortic blood being diverted to the pulmonary circulation. The volume of blood going to the systemic circulation is thus decreased and that returning to the right heart is decreased, and that to the left heart is increased. The importance of the amount of blood which reaches the lungs for areation is demonstrated in Blalock and Taussig's (9) patients by the extent of the rise in the oxygen saturation of the arterial blood, as a result of the operation. In one instance Blalock and Taussig (9) obtained a rise of 36.3 to 82.8% and in another from 35.5 to 83.8%.

The improvement in the general condition of the patients has paralleled the alterations in the satura-

tion of the blood and the decrease in the polycythemia. A change in the color of the mucous membrane is usually apparent shortly after completion of the operation. Cyanosis of the nail beds disappears more slowly and the recession or disappearance of the clubbing of the fingers and the toes requires longer time. A number of children who could walk only a few feet prior to operation can postoperatively walk a mile or more. Some of the children can even engage in fairly strenuous exercise. Improvement is reported in the oxygen saturation, red blood cell count, hemoglobin and the hematocrit readings. Blalock (5) reports a case in which the oxygen saturation rose in nine days from 35.5 to 80 percent and subsequently to 83.7 percent. In three months, the red blood cell count declined from 10.1 to 5.5 million and hemoglobin fell from 26 to 13.8 grams and the hematocrit reading changed from 81 to 38.

The shunt of the blood should not be excessive because this will place an undue strain on the heart. Only one of Blalock's (5) patients has developed enough enlargement to cause concern. He says a number of patients have shown an increase in size of the heart in the early postoperative period but this enlargement has not been progressive.

In the statistics of 110 patients operated Blalock

(6) did not perform the anastomosis in six patients. In four of these the pulmonary arterial pressure was too high and it was thought that the anastomosis was not indicated. These patients survived the exploratory operation. The other two patients died. In one of these the pulmonary artery was not found at operation and was located only with difficulty at time of autopsy and was small and in an abnormal position. In the other patient the chest was opened and it was immediately apparent that the patient would not stand the operation and he was closed immediately.

In two of Blalock's patients the subclavian was anastomized by mistake to the small artery to the upper lobe of the lung. Both of these patients died.

Blalock (9), by February 1946, had done four patients in which an end-to-end anastomosis was done and all four survived the operation and were improved.

After the ligation of the subclavian artery, Blalock (5) has had no trouble with the involved arm. Sympathetic nerve block was not done on any of the patients. He reports that the arm is a slight bit cooler than the opposite one for varying period of time. Motion and sensation were little, if at all, affected. The radial pulse has reappeared in some patients. He also reported the innominant artery was used in 23 cases.

Seven of these patients died. In two of these patients the preoperative diagnosis was in error. In the other five cases the death was attributed to cerebral ischemia or thrombosis.

In five cases the common carotid was used because of an abnormality in the vessels. He noted no substantial evidence of disturbance of cerebral circulation in these patients.

Of Blalock's (5) first 55 patients, 40 have shown definite improvement, three have shown little change and 12 have died. Two of those that died, on autopsy, were shown not to be Tetralogy of Fallot and should not have been operated.

Up to June 1947 Blalock (7) has operated 375 patients from 4 months to 26 years. His overall mortality is approximately 17 percent.

After surveying Blalock and Taussig's (11) work, Potts, Smith and Gibson (44) thought it would be of great help if the subclavian or the innominate artery wouldn't have to be sacrificed. They thought of the possibility of directly anastomosing the aorta and the adjoining pulmonary artery. Gross and Hufnagle (25) and Blalock and Park (10) proved from their experiments that clamping of the aorta completely for any length of time was impossible because of the damage caused to the spinal cord.

Potts, et al. (44), concluded that the anastomosis could only be performed safely if substantial amount of blood flowed through the aorta during the operation. Potts and Smith (44) made this possible by devising a new clamp. The clamp has two flanges which encircle the aorta and, when closed, pinch off a small but sufficiently large portion of the aorta to allow room for an anastomosis. The major portion of the aorta lies in the hollow of the clamp and rests against it. When the clamp is closed the lumen of the aorta is reduced to approximately half its regular size. This is sufficient for adequate blood supply to the spinal cord and they have had no complications from this.

Potts (45) states that for operation the patient must have cyanosis, the patient must not have a diastolic murmur, the heart must not be enlarged and the pulmonary artery pressure must be below 300 mm. of water. The best time for operation is between three and seven years of age.

Potts, et al. (44), use cyclopropane and oxygen anesthesia and also cannulate the saphenous vein at the ankle for continuous administration of plasma and emergency administration of blood. The chest is entered through the fourth interspace. The left pulmonary artery is identified and freed from the

surrounding structures. The pulmonary artery is occluded to determine if the right pulmonary artery alone can carry sufficient blood to maintain life during the anastomosis. The aorta is then dissected from its bed. The vagus nerve is identified and displaced posteriorly. The intercostal arteries arising from that portion of the aorta to which the clamp is to be applied are ligated. Silk sutures are placed around the pulmonary artery encircling the artery twice, one as far distally as possible and the other as far proximally as possible. The clamp is then applied to the aorta and closed slowly. The adventitia is then dissected from that portion of the aorta protruding from the clamp. With a sharp, small blade knife a longitudinal incision 4/16 inches is made in the clamped off portion of the aorta. The two ligatures doubly encircling the pulmonary artery are tied to the corners of the clamp, thus occluding the vessel and alinging it with the aorta. Opposite to the aorta an incision is made in the pulmonary artery. This incision is made slightly diagonal so that at completion of the anastomosis the vessels, in assuming their normal relationship, would not produce a kink in the pulmonary artery. The anastomosis is made with No. 5 Deknatel silk on a No. 9 curved atraumatic needle. The suture is begun at the upper angle, it is carried

through the pulmonary artery from the outside in, then across through the aorta from the inside out and tied. This brings the knot outside the vessels. In combination with this suture, the posterior lip of the pulmonary artery is sewed to the posterior lip of the aorta by a running over and over stitch. These stitches should be about 1 mm. apart, and include all layers of both vessels. At the lower angle of the wound, the suture is locked and then continued as a running suture bringing anterior lip of the pulmonary artery to the anterior lip of the aorta, intima to intima. The long end of the suture is tied to the short to complete the anastomosis. The ligatures holding the pulmonary artery are cut, first the distal then the proximal. The aortic clamp is then slowly removed. A thrill of the blood flowing through the new opening can be felt. The lung is then re-expanded and the incision closed. Any time during the operation you can back out, re-expand the lung and wait for the patient's condition to get better and then go back in for ten minutes or so.

Postoperatively Potts (45) reports there is no restriction of activity. The patients are not allowed to enter competative athletics. The major problem postoperatively is that of readjusting the child in the home. This psychiatric problem should be watched for

because they have been the center of attraction before the operation.

The main advantages to this operation are that the anastomosis can be made whatever size desired. Also there is no sacrifice of either the subclavian or the innominate artery.

This operation has been performed 45 times by Potts (44) with four deaths, a mortality of 8.8 percent.

	operated	deaths	mortality
4 months - 2 years	10	1	10.0%
2 years - 5 years	19	2	10.5%
5 years - 13 years	16	1	6.3%
	45	4	8.8%

Three of these patients died of cerebral thrombosis and one of a tension pneumothorax, the patient had tuberculosis. The other 44 patients have been helped greatly. None of the patients have died since they left the hospital and there is a higher mortality in the patients awaiting operation than in those operated.

At the present time this operation is only in an embryonic state and time will be the only factor that will tell if the formation of the artificial ductus arteriosus is of any lasting effect. The reports indicate that the immediate post-operative results are of definite value to the child but time is necessary to determine if endocarditis, or heart failure will

develop, if the anastomosis will grow with the individual or not, and if the operation will give the patient a normal life expectancy. Potts (46) at the present time is working on experiments to determine if the anastomosis will grow with the individual or not, and he expects to publish his results early in 1948.

CONCLUSIONS

The person with a congenital heart is in a much better position today than he has ever been in the past, because he may be one of the types that may be benefited by operative procedure. The prognosis of these patients is much improved.

It was through the studies of Gross, Blalock, and Potts that a treatment of the congenital heart evolved. As yet these operations are not simple and it takes the greatest of skilled operators to perform them. The Potts operation appears to be more logical, more versatile and have less complications than the Blalock operation, but an insufficient number of patients have been reported to make any all-inclusive conclusions. Now that supposedly successful procedures have been discovered, surgical teams will develop in leading medical centers. There is little doubt that in the near future, numerous new operations will be discovered and that the Blalock and Potts operations will be modified and improved, and the old barrier of doing nothing for the unfortunate child with the congenital heart condition will be torn down.

The definite results of the present operations will not be forthcoming for many years, because only

in years to come will the cardiac status of the patients as adults be known. There appears to be no reason at the present time why children with Tetralogy of Fallot should undergo the extreme incompacity and the danger of death from anoxemia or cerebral thrombosis.

BIBLIOGRAPHY

1. Abbott, M. E., Atlas of congenital cardiac disease, New York, American heart association, 1936.

2. Bedford, D. E., and Parkinson, J., Right-sided aortic arch, Brit. J. Radiol., 9:776-798, (Dec.) '36.

3. Bing, R. J., Vondom, L. D., and Gray, F. D., Jr., Physiologic studies of congenital heart disease, Bull. John Hopkins Hospital, 80:107-121, (Febr.) '47.

4. Blalock, A., Effects of an artificial ductus arterisus on experimental cyanosis and anoxemia, Arch. Surg., 52:247-252, (March) '46.

5. Blalock, A., Physiological and surgical treatment of congenital cardiovascular defects, Bull. New York Acad. of Med., 22:57-80 (Febr.) '46.

6. Blalock, A., The surgical treatment of congenital pulmonary stenosis, Ann. Surg., 124:879-887, (Nov.) '46.

7. Blalock, A., The surgical treatment of congenital pulmonary stenosis, Science, 105:633, (June) '47.

8. Blalock, A., The surgical treatment of tetralogy of Fallot, Surgery, 21:146, (Jan.) '47.

9. Blalock, A., The techniques of creation of an artificial ductus arteriosus in the treatment of pulmonic stenosis, J. Thoracic Surg., 16:244-255, (June) '47.

10. Blalock, A., and Park, E. A., The surgical treatment of experimental coarctation (stresia) of the aorta, Ann. Surg., 119:445-456, (March) '44.

11. Blalock, A., and Taussig, H. B., The surgical treatment of malformations of the heart, J.A.M.A., 128:189-202, (May) '45.

12. Bromer, R. S., The role of the Roentgenologist in the diagnosis of congenital cardiac lesions, J.A.M.A., 120:251-254 (Sept.) '42.

13. Burchell, H. B., Symposium on tetralogy of Fallot, Basis of cyanoses in tetralogy of Fallot, Proc. Mayo Clin., 22:162-165, (April) '47.

14. Clawson, B. J., Types of congenital heart diseases in 15,597 autopsies, Journal-Lancet, 64:134-136, (May) '44.

15. Cournand, A., and Richards, D. W., Jr., Pulmonary insufficiency, Am. Rev. Tuberc., 44:26-41, (July) '41.

16. Dautrebande, L., Marshall, W. R., and Meakins, J. C., Studies of the circulation in three cases of morbus caeruleus, J. Clin.Investigation, 8:123-135, (Dec.) '29.

17. Dry, T. J., Symposium on Tetralogy of Fallot, Introduction, Proc. Mayo Clin., 22:161-163, (April) '47.

18. Edwards, J. R., Bulbulian, A. H., and Milton, H. R., Symposium on Tetralogy of Fallot, Pathologic and embrologic considerations in Tetralogy of Fallot, Proc. Mayo Clin., 22:166-172, (April) '47.

19. Eppinger, E. C., Burwell, C. S., and Gross, R. E., The effects of patent ductus arteriosus in the circulation, J. Clin. Investigation, 20:127-143, (March) '41.

20. Fallot, L. A., Contribution a l'anatomie pathologique de la maladie bleue (Cyanose cardiaque,) Marseille Medical 25:418-420 1888. Translated by L. Morissette, Cardiac Classics, F. A. Willia, St. Louis, 1941.

21. Fashena, G. J., Recognition of certain forms of congenital heart disease ammenable to surgical treatment. South, M. J., 39:948-56, (Dec.) '46.

22. Garb, S., Relation of blood viscosity to the intensity of heart murmur, Am. Heart J., 28:568-573, (Nov.) '47.

23. Gibson, S., The cardiovascular system in Brennemanns practice of pediatrics. W. F. Prior Co., Vol. 3, chapter 13, '45.

24. Gross, R. E., and Hubbard, J. P., Surgical ligation of a patient ductus arteriosis, Report of first successful case, J.A.M.A., 112:729-731, (Febr.) '39.

25. Gross, R. E., and Hufnagel, C. A., Coarctation of the aorta, New England J. Med., 233:287-289, 1945.

26. Grishman, A., Steinberg, M. F., and Sussman, M. L., Tetralogy of Fallot: contrast visualization of heart and great vessels, Radiology, 37:178-180, (Aug.) '47.

27. Harper, H. T., and Mathews, M. W., The practical diagnosis of congenital heart lesions amenable to treatment. J. M. Ass. Georgia, 35:280-284, (Oct.) '46.

28. Harris, J. S., and Farber, S., Transportation of the great cardiac vessels with special reference to the phylogenetic theory of Spitzer, Arch. Path., 28:427-502, (oct.) (38.

29. Herndon, R. F., Voss, A., and Donovan, J. J., The Tetralogy of Fallot, Am. Heart J., 17:553-560, (May) '39.

30. Hope, J., A treatise on the diseases of the heart and great vessels. Philadelphia American ed. 1, from London ed. 3, Haswell and Johnson, 449-460, 1842.

31. Kaner, A., Losch, P. L., and Green, H., Oral manifestations of congenital heart disease, J. Pediat., 29:269-274, (Sept.) '46.

32. Leech, C. B., Congenital heart disease, clinical analysis of seventy-five cases from John Hopkins, J. Pediat., 7:802-839, (Dec.) '35.

33. Lev, M., and Saphir, O., Transportation of the large vessels, J. Tech. Methods, 17:126-162, (Sept.) '37.

34. Levy, S. E., and Blalock, A., Experimental observations on the effects of connecting by suture the left main pulmonary artery to the systemic circulation, J. Thoracic Sur., 8:525-530, (June) '39.

35. Lundsgaud, C., and Van Slyke, D. D., Cynosis, Medical Monograph, Vol. 2, Baltimore, Williams and Wilkins Co., 1923.

36. McGinn, S., and White, P. D., Prognosis in the recognition of congenital heart disease, New England M. J., 214:763-768, (April), '36.

37. McGuire, J., and Goldman, F., Apparent increased velocity of blood flow in cases of congenital heart disease with septal defects having right-left shunt. Am. Heart J., 14:230-235, (Aug.) '37.

38. Middleton, W. S., and Richie, G., The tetralogy of Fallot, Am. Heart J., 33:250-253, (Feb.) '47.

39. Miskall, E. W., The tetralogy of Fallot, J.A.M.A., 128:803-804, (July) '45.

40. Olim, C. B., and Hughes, J. G., The Blalock operation for congenital pulmonary stenosis, Southern Surgeon, 13:167-180, (March) '47.

41. Parker, R. L., Symposium on tetralogy of Fallot, Differential diagnosis of tetralogy of Fallot, Proc. Mayo Clin. 22:177-179, (April) '47.

42. Perlman, L., and Meyers, J., A case of tetralogy of Fallot with venucose endocardites. Ann. Int. Med., 22:121-128 (Jan.('45.

43. Pescatore, J. A., Wolffe, J. B., and Digelio, V. A., Tetralogy of Fallot: correlation of clinical roentgenologic, and postmortum findings. Am. Heart J., 17:489-493, (April) '39.

44. Potts, W. J., Smith, S., and Gibson, S., Anastomosis of the aorta to the pulmonary artery. J.A.M.A., 132:627-31, (Nov.) '46.

45. Potts, W. J., Surgery for congenital heart

disease, Lecture - Douglas County Medical Society, Omaha, Nebraska, (Sept.) '47.

46. Potts, W. J., Personal Communication.

47. Pugh, D. G., Roentgenologic diagnosis of certain congenital lesions of the heart and great vessels. Am. J. M. Sc., 212:628-635, (Nov.) '46.

48. Reyes, A. P. de los, Costellanos, A., and Pereiros, R., Angiocardiography and its value, Am. Heart J., 25:298-306, (March) '43.

49. Robb, G. P., and Steinberg, K., Visualization of the chambers of the heart, pulmonary circulation, and the great vessels in man, Am. J. Roentgenol., 41:1-17, (Jan.) '39.

50. Segall, H. N., A case of tetralogy of Fallot: clinicopathological observations; quantative studies of circulation rate and right-to-left shunt. Am. Heart J., 8:628-629, (June) '33.

51. Sussman, M. L., Grishan, A., and Steinberg, M. F., Newer concepts in the diagnosis of congenital heart disease, Am. J. Dis. Child., 65:922-936, (June) '43.

52. Talbott, J. H., Corombs, F. S., Castleman, B., Chamberlain, F. L., Consolozio, W. V., and White, P. D., A record case of tetralogy of Fallot: with comments on metabolism and pathologic studies, Am. Heart J., 22:754-777, (Dec.) '41.

53. Taussig, H. B., Diagnosis of the tetralogy of Fallot and indications for operation. Am. J. Dis. Child., 73:245-246, (Feb.) '47.

54. Taussig, H. B., The tetralogy of Fallot: diagnosis and indication for operation, Surgery 21:145, (Jan.) '47.

55. Volini, I. F., and Flaxman, N., Tetralogy of Fallot, J.A.M.A., 111:2000-2003, (Nov.) '38.

56. White, P. D., Heart disease, 2 ed. New York, Macmillan, 1937.

57. White, P. D. and Sprague, H. B., The tetralogy of Fallot, J.A.M.A., 92:787-791, (March) '29.

58. Wise, S. P. III, Some correctible anomalies of the heart and great vessels, N. Orleans M. & S. J., 99:280-284, (Dec.) '46.