"Tetralogy of Fallot"

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Doctor Thompson: Today we are going to discuss the most common form of cyanotic congenital heart disease: tetralogy of Fallot. Doctor Cartaya will present the case. Doctor Vanhoutte did the radiological study on this child. I will discuss diagnosis and the clinical picture. Doctor Williams will discuss surgical management.

Doctor Cartaya: The patient was a two-year-old white girl with a heart murmur known since she was seven weeks of age; she was asymptomatic at that time. By four months of age the child was said to have generalized hypercyanotic spells, especially when crying. The diagnosis of tetralogy of Fallot was confirmed by cardiac catheterization performed when the child was at 14 months of age. Subsequently a right Blalock-Taussig anastomosis was performed. The postoperative course was benign and there was a subsequent decrease in hypercyanotic spells. Definitive surgical repair of the cardiac defect and take-down of the Blalock shunt was done March 1, 1972, while the patient was on cardiopulmonary bypass. She had transient postoperative edema and azotemia which was thought to be secondary to inappropriate antidiuretic hormone (ADH) secretion and slight underhydration. She was discharged 11 days postoperatively (March 12) in satisfactory condition except for some persistent edema and a slight cough. At the time of discharge she was afebrile. Twenty-four hours after discharge, however, the child developed fever, progressive grunting respiration, dyspnea, and cough, and consequently was readmitted with the initial impression of postpericardiotomy syndrome. Physical examination revealed a pale, slightly dyspneic girl with minimal circumoral cyanosis and a cough. Respiration rate was 42/min, and heart rate was 120 to 130/min. Examination of head, ears, eyes, nose, and throat was unremarkable except for the presence of inflamed gingiva. There was a healing, midsternal, Y-shaped incision over the chest. Inspiratory rales were heard at the bases of both lungs, but primarily on the left. There was a grade IV/VI stysolic murmur heard over...
being maintained on digoxin (Lanoxin), 0.1 mg twice a day.

*Doctor Vanhoutte:* Figure 1 is a reproduction of the first roentgenogram of the chest that we have on this child. The child was two months old. The lungs are clear, slightly hyperexpanded, and marginally perfused. The overall size of the cardio-mediastinal outline is well within normal limits. The apex of the heart, however, appears a little elevated. This suggests right ventricular hypertrophy. Combined with the clinical observation of cyanosis, these findings suggest the diagnosis of tetralogy of Fallot. It is not at all unusual not to see the "boot-shaped" heart at this early age.

At the time the roentgenogram of the chest reproduced in Figure 2 was obtained, the patient was 15 months old. Six weeks before, she had undergone a Blalock-Taussig anastomosis on the right side. Because of this anastomosis, the pulmonary perfusion has improved. The slight elevation of the cardiac apex persists. The heart size remains normal.

An angiocardiogram was obtained when the patient was 29 years old. Via the superior vena cava, the injection of contrast agent was made in the right ventricle. On the frontal projection (Fig 3A), during cardiac systole the right ventricle outflow is exceedingly narrow. This is followed by a small infundibular chamber, sometimes referred to as a third ventricle. The main pulmonary artery is not well seen on this projection. On the lateral projection (Fig 3B) it is well-demonstrated and appears hypoplastic. This patient, therefore, has at least two levels of obstruction to the rightventricular outflow. Multiple levels of obstruction are not uncommon in tetralogy of Fallot. Incidentally, a small amount of contrast agent can be seen in the aorta. It reached this locale by crossing the ventricular septal defect.

At the age of 32 months (Fig 4), just prior to total correction, the patient’s pulmonary perfusion has again become marginal. The apparent slight increase in the heart size and the apparent lesser elevation of the cardiac apex are the result of a shallower inspiration, rather than a true change in the heart.

One week after total correction (Fig 5), the pulmonary perfusion is similar to what it was after the Blalock-Taussig anastomosis (Fig 2). The heart is slightly larger than it was preoperatively (Fig 4). This slight increase in cardiac size is not unexpected shortly after a total correction.

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Fig 1. Roentgenogram at two months of age suggesting diagnosis of tetralogy of Fallot.

The entire precordium. Point of maximal impulse (PMI) was slightly shifted to the midaxillary line, and no rub was heard.

First and second heart sounds were muffled. The liver was thought to be depressed four cm; the spleen was not palpated. There was no clubbing or cyanosis of the extremities, but there was 2+ to 3+ pitting edema. Neurologic findings were within normal limits. Roentgenograms revealed a slight increase in cardiac size, with indistinct apex and prominent pulmonary vascularity. Laboratory data revealed the hematocrit was 32.2%, WBC was 8,700/cu mm (no differential given), and the urine was normal. On the day after admission, a friction rub compatible with postpericardiectomy syndrome was heard over the precordium. Treatment consisted of bed rest and the administration of aspirin. The patient was discharged five days after admission. Two follow-up examinations in the OPD revealed gradual improvement in her condition. She is

Fig 2. Roentgenogram at 15 months of age, six weeks after Blalock-Taussig anastomosis on right side.
by the cardiac silhouette on this reproduction is suggestive of pericardial effusion. The typical picture of pericardial effusion is altered in this case by postoperative adhesions between the pericardium, the pleura, and the anterior chest wall. At this time, minimal thickening of the inter-lobar fissures was the only evidence of pleural effusion. Six weeks after the surgical correction (Fig 7), the cardiac outline is well on its way to assuming a normal configuration.

In summary, I have shown the classical evolution of the roentgenographic findings in tetralogy of Fallot. A relatively benign form of the "post-pericardiotomy syndrome" was the only complication this child had.

**Doctor Thompson:** This child did have the combination of abnormalities usually referred to as tetralogy of Fallot. We will go into some detail in this discussion because of the fre-
Fig 6. Roentgenogram 24 days postoperatively.

Fig 7. Roentgenogram six weeks postoperatively.

abnormalities do not survive their first year unaided by surgery. The cause is unknown, as it is in most cases of congenital heart disease. It is known, however, that there is a higher incidence of patients with tetralogy of Fallot with increasing maternal age. The reported sex incidence varies somewhat; most give a 6:4 ratio of males to females. From an embryologic viewpoint, the malformation is best explained by maldevelopment of the bulbus cordis, the structure from which the outflor tract of the right ventricle is derived, and which contributes to formation of the membranous portion of the ventriculum septum. Defects in the development of the bulbus cordis occur between the fifth and seventh weeks of intrauterine life, implying that the insult causing this syndrome occurs during that period. The anatomy of the defect is interesting and quite variable. Classically, as described by Fallot, it includes a ventricular septal defect, pulmonary stenosis, dextroposition of the aortic root, and right ventricular hypertrophy. The conclusion that this particular combination gives rise to the same clinical picture in all cases is not tenable. Modern means of studying cardiac function in the intact human being bring this out sharply.

Individually, the component parts of the abnormality are important and are quite interesting. In patients with tetralogy of Fallot, the ventricular septum is always patent; the ventricular defect is usually large (one to three cm in diameter), including virtual absence of the membranous portion of the ventricular septum. It is located in the membranous portion of the septum with close relationship to the septal leaf of the tricuspid valve and to the aortic root.

Normally, there is a very close spatial relationship of the membranous septum to the aortic valve, which lies in the lowest of the semi-lunar valves and appears to straddle the ventricular septum. Perforation of the membranous portion of the ventricular septum in a normal individual will give the appearance of aortic overriding. Actual malposition of the aorta is probably relatively uncommon in tetralogy of Fallot. The superior margin of the defect from the left ventricular side would be contiguous with the right or posterior aortic cusp. The lower border usually is made up of the muscular portion of the septum. It is thick, saddle-shaped, and is, of course, near the common conduction system, the bundle of His. I am
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sure Doctor Williams will comment on this relationship.

The pulmonary stenosis, which is also a consistent anatomic anomaly in tetralogy of Fallot, is more variable than is the septal defect. The narrowing may involve the infundibulum; it may involve the valve; it may involve both and may be supravalvular in site. There is almost always hypertrophy of the crista supraventricularis, which may produce further stenosis on a functional basis. Complete atresia of the outflow tract is not uncommon and will occur in approximately 20% of all cases. The site of stenosis varies sharply between autopsy and operative cases, but in approximately 50% of patients with tetralogy of Fallot the stenosis will be subvalvular alone; in another 20% to 25%, it will be a combination of valvular and subvalvular. Distal pulmonary stenosis also occurs.

Dextroposition of the aorta probably is not really present. It is certainly of little physiologic significance and has little or no prognostic importance. The amount of this is difficult or even impossible to quantify at autopsy, and as mentioned, does not necessarily imply an abnormal position of the aortic root. It is of surgical importance since damage to the aortic valve can occur in repair of any ventricular septal defect.

Right ventricular hypertrophy is a secondary phenomenon present in all patients who have pulmonary stenosis. It is of little clinical or physiologic importance, except that it may contribute to the degree of obstruction because of hypertrophy of the outflow tract area.

The development of the left ventricle in some very severe cases may be abnormal. This, however, usually is not a significant clinical problem. In the majority of cases the thickness of the left ventricular wall is equal to that of the right ventricle. There is sometimes variation in the chamber volume, but in approximately 50% of the cases the chambers are equal in size.

Other anatomic variations are of interest. The atria usually will show little change, although the right atrium may show mild hypertrophy. Patency of the atrial septum is rare in some series, more common in others, but usually is of no functional significance. The importance of recognition for surgical correction is obvious. Enlargement of the aorta is usually seen, and is a function of the degree of the right-to-left shunt; the larger aortas are found in those patients in whom the shunt is greatest. Approximately 20% to 30% of patients with tetralogy of Fallot will have a right-sided aortic arch. A right-sided aortic arch occurs with such frequency in only one other congenital abnormality: truncus arteriosus. The presence of a right-sided arch is frequently a clue to the particular diagnosis. Collateral pathways for pulmonary blood flow are increased; the most common example is persistent patency of the ductus arteriosus. This has been reported to occur in 20% to 25% of the patients with this abnormality. Although this complicates the situation, chances for survival are more favorable. Dilated bronchial collaterals occur very frequently. They are more severe in extreme cases and are a function of age, being noted in the older patients more often than in the younger ones. Noncardiac abnormalities occur in approximately 20% of patients with tetralogy of Fallot. These are most commonly renal abnormalities of some type.

The physiology of the combination of abnormalities is almost as variable as the anatomy. From a functional viewpoint, the essential components of the abnormality are the presence of the ventricular septal defect and pulmonary stenosis. Obviously, the spectrum can be quite wide, depending on the relative severity of these two lesions. With very small ventricular septal defects, function is essentially the same as in persons with an intact septum. In the typical tetralogy the defect is large, so that there is no gradient in pressure across the defect; the two ventricles function with a common ejectile force and with equal pressures. The pathophysiology is thus dependent upon the degree of pulmonary stenosis. In patients with little pulmonary stenosis, the clinical picture is similar to that of any other patient with a large left-to-right shunt. This grades all the way down to those with complete pulmonary atresia and a total right-to-left shunt at the ventricular level. The clinical picture, of course, will vary. It should be remembered that in all patients in whom the pulmonary stenosis is severe enough so that there is no shunt, or a right-to-left shunt, the total cardiac output is normal. The right ventricular pressure load can never exceed that on the systemic side. Thus, cardiac size is not increased and cardiac decompensation does not occur on the basis of the anatomic abnormality.

In the cyanotic patient the pulmonary blood flow is fixed, since the right ventricular pres-
sure can never exceed the systemic pressure, and thus cannot rise to produce a greater flow through the stenotic area. It is more likely that the degree of pulmonary stenosis will actually increase rather than decrease. In these patients, pulmonary blood flow is maximal and any significant change in venous return is usually associated with changes in the volume of the shunt across the ventricular septum rather than with any significant change in pulmonary blood flow. The pulmonary blood flow is decreased, with a corresponding decrease in the size of the pulmonary vessels. Under the circumstances and with peripheral vasodilation, there will also be an increase in the right-to-left shunt.

The most striking clinical finding in these patients is cyanosis. This is strictly a function of the degree of pulmonary stenosis and adequacy of the diet, and not of the position of the aorta. The onset of cyanosis with tetralogy of Fallot is extremely variable. Approximately one-third of these patients are cyanotic from birth. Another one-third become cyanotic during the first year of life. The remainder may not become cyanotic until the early teen years. The degree of cyanosis varies in the same patient; it varies even more from one patient to another. There is a tendency, however, for it to become more severe with increasing age, size and activity. None of these patients are really asymptomatic. Most of them have a tendency to squat with activity, at least early in life, and they are almost invariably dyspneic with exertion. A history of hypercyanotic spells is obtained frequently, as is the case in this particular child. These episodes may produce unconsciousness, convulsions, and death. They are an indication of the severity of the abnormality. Their occurrence is an indication for early surgical intervention.

On physical examination, these patients are cyanotic. The cyanosis is central in type and involves the lips, mucous membranes and nail beds. It is frequently associated with suffusion of the conjunctiva, a malar flush and, not uncommonly, clubbing of the nose. Physical development in these patients is often retarded, although not necessarily so. Poor muscular development is almost certain. There is no precordial bulge, the precordial activity is normal, and clinically, cardiomegaly is not present. Cardiac findings include a systolic ejection murmur over the outflow tract and pulmonary valve area; it is a murmur of pulmonary stenosis. There may be a thrill and, usually, a relatively pure basal second sound. The degree of obstruction obviously affects the murmur, as in other patients with pulmonary stenosis, and the murmur is completely absent in those who have pulmonary atresia. With the presence of ductus arteriosus, or very marked development of the bronchial collateral patency vessels, or both, continuous murmurs are common. The murmur of the blood flow will be noted over the typical area. An increase in anteroposterior diameter of the chest is fairly common. There usually is no evidence of venous engorgement or anything else to suggest heart failure. The electrocardiogram will show evidence of right axis deviation and unless right ventricular hypertrophy is quite severe, there usually is no evidence of atrial hypertrophy. In patients with the same degree of pulmonary stenosis with an intact septum, evidence of right atrial hypertrophy is usually seen.

Doctor Vanhoutte has commented on the radiograph picture of this abnormality. I would like to emphasize again that right-sided aortic arches occur fairly often with this abnormality, and with pure tetralogy of Fallot, cardiac enlargement is not part of the picture.

Cardiac catheterization in these patients is of little help. It will reveal evidence of right ventricular hypertension at systemic levels. There usually is a very slight left-to-right shunt at the ventricular level, and of course, a right-to-left shunt will be present. There will be a gradient across the outflow tract with normal pulmonary arterial pressures. Peripheral arterial unsaturation will be present. The catheter frequently can be manipulated across the ventricular septal defect and into the ascending aorta. Selective angiocardiography, as noted, will reveal simultaneous filling of both the pulmonary artery and the aorta, and will reveal details of the anatomy as shown by Doctor Vanhoutte.

Prognosis in patients with tetralogy is variable. Rather remarkable longevity has been reported on occasion, with survival up to the fifth and sixth decades. Most patients, however, do not survive the teenage period. The signs and symptoms and prognosis, unaltered by surgical intervention, vary with the degree of pulmonary stenosis.

Patients with tetralogy of Fallot are usually grouped into three categories. Group 1 comprises the most severe forms of the disease, and includes the patients with pulmonary atresia. These patients are cyanotic from birth. Not uncommonly they are dependent upon the ductus
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arteriosus or bronchial collaterals or both for the major portion of their pulmonary blood flow. Cyanosis, however, is quite severe. Almost always the hypercyanotic spells begin to occur at the age of three or four months and usually increase in frequency and severity. Unless these patients are operated upon, they rarely survive the first year of life. Death usually occurs with a major hypoxic spell. If they survive the first year on collateral flow, they may live for several years, but they are markedly cyanotic and severely disabled. They seldom survive the second decade of life. In this group of patients, squatting is rare. They are usually very irritable, but mentally alert.

Group 2 patients are considered the most typical of this particular combination of abnormalities. They have the murmurs and are cyanotic, although not as severely. Hypoxic spells begin around six months of age, but they occur less frequently and with less severity than in Group 1. These children are also very irritable, intolerant of exercise, and usually mentally alert. A loud systolic ejection murmur and thrill are present over the precordium. They do not tolerate extremes in temperature. Symptoms become more severe as the children grow older and become more active. Their difference from other children, which grows more marked, becomes a major problem both for the child and for the parents. Survival into the second decade of life is common. Rarely, survival into the third decade occurs. They usually meet their demise in trying to cope with the stresses of adolescence. Death in these patients is associated with hypoxic episodes, cerebral accidents, brain abscesses, and endocarditis. Congestive heart failure can be seen in patients with severe anemia or with endocarditis, particularly if they develop aortic regurgitation.

As mentioned, both of these groups of patients have episodes that are usually referred to as hypercyanotic spells or blue spells. The spells tend to occur after exercise, after feedings, or upon arising in the morning. Many theories as to the cause have been put forth. These include episodes of spasm of the outflow tract resulting in an increase in the degree of cyanosis. The efficacy of morphine in treating spells was postulated as due to the effect of decreasing myocardial contractility, thus decreasing the degree of pulmonary stenosis. Studies have shown, however, that morphine given in doses that are effective increase myocardial contractility and, if anything, actually increase the degree of obstruction. The most logical explanation of these episodes is that they are related to hyperpnea. In these patients, for one reason or another, the right-to-left shunt increases and arterial oxygen saturation decreases. The normal response to this is an increase in the rate and depth of respiration. In normal persons, this results in an increase in the degree of oxygenation of the blood. This cannot occur in patients with tetralogy of Fallot, since their pulmonary blood flow and oxygenation are maximal at all times. Thus, with an increase in hyperpnea and breathing effort, more oxygen is consumed. Arterial saturation drops further, and a vicious cycle is set up that eventually results in unconsciousness and convulsions. The treatment in these patients is the administration of morphine, because of its depressant effect on respiratory activity. In contrast to what occurs in a normal person when morphine is given in respiratory-suppressant dosages, the arterial saturation in these patients rises rather than drops. In addition, since an increase in the venous return produces a greater right-to-left shunt, decreasing the venous return in these patients tends to improve the arterial saturation. The immediate tendency of almost all concerned with the care of these patients is to administer oxygen. Fortunately, as long as the oxygen is cool, it will do no harm.

The Group 3 patients are those with the so-called pink tetralogy or atypical tetralogy. These patients have a lesser degree of pulmonary stenosis without a gross right-to-left shunt with cyanosis. These patients commonly survive into the third or fourth decades of life, although rarely beyond age 40. The treatment of this particular condition, as of most other types of structural abnormalities of the heart, is surgical.

There are many types of procedures employed for correction of this abnormality. Careful consideration of the physiology of this abnormality leads to the concept of partial physiologic correction, resulting in amelioration of the abnormality rather than complete correction. The historical importance of this particular procedure is tremendous, because its success led to the development of the whole field of cardiac surgery and pediatric cardiology.

In the adult patient with tetralogy of Fallot
there usually are marked fibrotic changes in the outflow tract, which further increase the difficulty of total correction. Proper management of the adult case is not well documented. Complications become more frequent with advancing patient age. The incidence of cerebral abscesses in these patients is high, as it is in any patient with cyanotic abnormalities. Such abscesses are rare before the age of two. They frequently follow a respiratory tract infection; symptoms at onset are headache, vomiting, personality changes, and focal neurologic findings. Fever may or may not be present, but usually is. It is important to remember this greater frequency, since the diagnosis is difficult. The importance of making an accurate diagnosis and undertaking proper treatment is obvious. Polycythemia, and the resulting increase in blood viscosity, is of importance. With polycythemia, bleeding and clotting problems become significant, presumably on the basis of a consumption coagulopathy. This is a phenomenon based on time, becoming of increasing incidence and severity with advancing age. Bacterial endocarditis, another significant danger, probably occurs more frequently in patients with tetralogy of Fallot than it does in almost any other congenital anomaly of the heart. Hemoptyis is common in the older patients due to rupture of extensive collateral vessels in the lung; it also can be associated with thrombosis and pulmonary infarction. Gastrointestinal bleeding, probably due to thrombocytopenia, can occur. Gout does occur in the older patient; I am not familiar with reported cases in children. Tetralogy of Fallot may protect individuals from the complication of coronary heart disease, since in the patients that survive into adulthood, huge coronary arteries are seen that are not significantly involved with atherosclerosis. Perhaps the alternative is worse than the disease that is cured.

Doctor Williams will now comment on the surgical management.

Doctor Williams: The tetralogy of Fallot is interesting to surgeons for several reasons. Foremost, it is a common and disabling abnormality. Doctor Thompson has given the incidence figures. A second reason for interest is that very effective surgical treatment has been developed within a relatively short period of time, and it now seems possible to restore these incapacitated children to a near-normal life expectancy in the majority of instances. A final reason is the considerable historic interest in this form of congenital heart disease.

The first successful operation for congenital heart disease was the successful ligature of a patent ductus arteriosus in 1939 by Dr. Gross. This was a significant surgical achievement, but perhaps because the child with patent ductus really does not appear to be sick, it did not attract a tremendous amount of attention. The introduction of a procedure which dramatically changes a scrawny cyanotic baby to a pink, normal-looking child, however, did generate world-wide attention, and I believe this was the greatest stimulus for the interest in cardiovascular surgery that has resulted in enormous progress in the three decades since Doctor Blalock's operation. Doctor Alford Blalock was a young surgeon who had come from Vanderbilt to be Chief of Surgery of the Johns Hopkins Hospital. Principally interested in the surgical laboratory and in surgical physiology, Doctor Blalock had developed in the dog lab a method of producing pulmonary hypertension by anastomosing a brachiocephalic artery to the pulmonary artery. This was a nice operation, but it did not produce pulmonary hypertension and was a failure as far as Doctor Blalock was concerned. Doctor Helen Taussig became interested in the heart while she was a medical student in Boston. After graduation, she came to Johns Hopkins to work in the Department of Pediatrics under Doctor Edwards Park. Doctor Taussig did for pediatric cardiology what Doctor Maud Abbott had done for the pathology of congenital cardiac lesions some decades before. She systematically attempted to make an accurate diagnosis and to categorize both the physical findings and the physiologic effects in the various forms of congenital heart disease. In the process, it occurred to Doctor Taussig that a method of surgically improving pulmonary blood flow ought to greatly improve children with tetralogy of Fallot. She mentioned this to Doctor Blalock, who thought of his experiments attempting to produce pulmonary hypertension. The two of them decided that this could and should be tried in children. The first operation was performed. The procedure went very well and the child became pink. Interest and enthusiasm rapidly diffused throughout the institution and, although the early experience was difficult and the mortality rate high, the saving of these infants started a distinct trend.

Today, proper management of the tetralogy...
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depends on the age at which the child becomes symptomatic. In the 30% of patients with tetralogy of Fallot who become significantly symptomatic and begin to have hypercyanotic spells in early life, we feel that palliative procedures are still the operations of choice. There is an arbitrary dividing line at three years of age. If children have to have something done before three years of age, we recommend a palliative operation.

Palliative operations for tetralogy at the present time are four in number. The oldest of the palliative operations is the Blalock-Taussig procedure, in which the subclavian artery on the side opposite the arch is simply divided and the proximal end of the subclavian artery brought down and attached by vascular anastomosis to the side of the pulmonary artery. This perfuses both lungs with systemic unsaturated blood. It is not an easy operation, but it has now been done in large numbers and produces very satisfactory palliation in the majority of patients in which it is carried out successfully. The operation does not work very well in the small infant, because the structures are so small that an appropriate anastomosis which will remain patent is difficult or impossible to construct.

Not long after Doctor Blalock's operation was described, Doctor Willis Potts in Chicago described a procedure that could be applied more successfully in infants. The second operation, known as the Potts procedure, is a direct anastomosis between the descending thoracic aorta and the side of the left main pulmonary artery. Where the left main pulmonary artery crosses the aortic arch, it is possible, using a set of special instruments, to make a longitudinal anastomosis between the two. It is technically simpler than the Blalock procedure, and it has the great advantage of being applicable to infants. Its disadvantage, however, is that it is extremely difficult to take at a later date when total correction is done, and this difficulty has led to the virtual abandonment of the Potts operation. What most surgeons do now when shunting is indicated was described by David Waterston at Great Armond Street Hospital a number of years ago and is known as the Waterston operation. It consists of an anastomosis between the ascending aortic arch and the right main pulmonary artery behind the aorta. This operation is also known by a number of other surgeons' names (Cooley) and has the great advantage of being very simple to correct at later repair of the defect.

The fourth operation that can be done in the infant that requires palliation is the so-called Broek operation, in which a blind attempt at opening the stenotic pulmonary outflow tract is performed. I mention it because there are two or three centers in the United States in which it is done. We have not used this procedure in a tetralogy here.

In all children, whether they required a palliative operation before age three or not, we think that correction of the tetralogy of Fallot should be carried out before they enter school. The operation is technically as easy at that time as any other. The adjustment problems are somewhat less and, actually, the procedure seems easier on the child at that age than it does if the child is not operated on until a later age. The correction of the tetralogy of Fallot is carried out with the patient on cardiopulmonary bypass. The operation has two components: the first problem is to close the ventricular septal defect, and the second is to relieve pulmonary stenosis. The ventricular septal defect of the tetralogy of Fallot is anatomically different from any other ventricular septal defect, and it is a little harder to close. The other concern with closure of the ventricular septal defect is the section of the bundle of His. The conduction bundle passes close to the ventricular septal defect. Since there is, as yet, no reliable method of detecting the presence of the bundle, the production of complete heart block during closure of this ventricular septal defect is always a possibility, and one which has dire consequences for the patient. The second part of the problem is opening the pulmonary outflow tract. Variations in the anatomy of the outflow tract are common. Stenosis may be infundibular, valvular, or both. Opening the valve is extremely simple, but often infundibular stenosis requires a fairly complicated dissection of the musculature of the outflow tract of the right ventricle. When results of the operation to correct the tetralogy are poor, the most common cause by far is failure to completely relieve the ventricular outflow obstruction.

Multiple postoperative problems may follow correction of the tetralogy of Fallot. These include the bleeding abnormalities that are a threat in any cyanotic patient. Of course, such problems are somewhat magnified by the fact...
that one does have to suspend the clotting systems of patients who go on cardiopulmonary bypass. Heart block may require the use of a pacemaker. Postoperative bleeding not related to clotting difficulties is a problem because of extensive collateral vessels. Finally, failure to adequately perform the two mechanical parts of the procedure, that is, closure of the ventricular septal defect and relief of pulmonary stenosis, adds to postoperative problems.

The mortality for tetralogy of Fallot repair was very high all through the 1950s and even into the early 1960s. The mortality now is below 10%. The percentage of satisfactory anatomic and physiologic corrections probably exceeds 85%, and it looks as if these recovered children will live relatively normal lives.

Somebody is going to ask about the post-pericardiomyotomy syndrome this child presented. This is an as-yet-unexplained phenomenon that can occur after any procedure in which the pericardial cavity is opened. It consists of fever, splenomegaly, splinter hemorrhages, and pericardial effusion or at least pericardial friction rubs. The only nice thing about it is that it goes away with the use of salicylates. We do not know the cause. It is not common after surgery for the tetralogy of Fallot; surprisingly it is most common after closure of an atrial septal defect.

Repair of the tetralogy of Fallot is one of our more successful areas in surgery. It does restore these children, with a life expectancy of a decade and one half at best, to what we think will probably be a normal life span.

**Doctor Seely:** I want to ask Doctor Williams about the duration of follow-up available on a reasonable number of children and about their growth.

**Doctor Williams:** The first total corrections of tetralogy of Fallot were performed in 1954. No one has very many cases and even fewer survivors for about the subsequent five years. So, at the present time, we are talking about a 12-year follow-up in any appreciable number of patients. I cannot give you the figures in relation to growth; perhaps Doctor Thompson can. Most of the children grow at a normal rate, granted that they started low on the scale.

**Doctor Thompson:** Statistically, most of the children are smaller than other children of their age and at the same degree of development. There are a significant number of patients who do have a growth spurt. Most, as Doctor Williams mentioned, never reach normal size, but they grow at an essentially normal rate after the operation. Apparently they lose much of their growth potential in the first year or so of life, before correction, and never really catch up. Frequently, these patients have been classified as having a significant central nervous system problem with low intelligence because of the cyanosis and cerebral hypoxia. In general, this is not severe and I doubt the accuracy of the previous classification. There is a tendency to operate, with the idea that they may be able to achieve more potential of total growth and development. This is true regardless of the abnormality you are dealing with. It is of interest to note that nobody commented on the place of digitalis in the treatment of these patients. They do not require digitalis prior to surgery, but most of these patients show very poor right ventricular function for 12-to-18-months postoperatively. For this reason, we usually digitalize all tetralogy of Fallot patients after the operative procedure.

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**ANNOUNCEMENT**

Application for Research Grants-in-Aid and for Postdoctoral Research Fellowships, for the year beginning July 1st, 1977, are now being accepted by the American Heart Association, Oklahoma Affiliate, for review by the Research Policy Committee.

Application forms may be obtained from the American Heart Association, Oklahoma Affiliate, Inc., 800 N.E. 15th Street, P.O. Box 11376, Oklahoma City, Oklahoma 73111. These applications must be received or postmarked no later than November 1st, 1976.