DIAGNOSIS OF THE TETRALOGY OF FALLOT AND MEDICAL ASPECTS OF THE SURGICAL TREATMENT*

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THE tetralogy of Fallot, as described by Fallot,[1] consists of pulmonary stenosis or atresia combined with dextroposition of the aorta, an interventricular septal defect, and right ventricular hypertrophy. It is worthy of note that although Fallot emphasized that such cases constituted a pathological entity, the first case was described more than 100 years earlier by Sandifort.[2]

The pulmonary stenosis usually involves the musculature of the right ventricle below the pulmonary valve. Sometimes the stenosis extends up to the valve itself as in Figure 1. It is, however, common to find the maximum stenosis occurs in the infundibulum of the right ventricle as shown in Figure 2. Although the pulmonary artery is usually smaller than the aorta, the pulmonary artery beyond the pulmonary valve is normally formed. Dextroposition of the aorta means that the aorta, although it arises from the left ventricle, over-rides the top of the ventricular septum and receives some blood directly from the right ventricle. The over-riding of the aorta renders inevitable a high ventricular septal defect. Such is the nature of the "interventricular" septal defect in the tetralogy of Fallot. The pulmonary stenosis and the dextroposition of the aorta both increase the work of the right ventricle. Consequently there is right ventricular hypertrophy.

Clinically a patient with this malformation shows cyanosis and clubbing and a heart of normal size. A systolic thrill and a harsh systolic murmur along the left sternal border are of common occurrence, but neither the murmer nor the thrill are essential for the diagnosis. Indeed if the pulmonary stenosis is extreme too little blood may pass through the pulmonary orifice to produce a murmur. The over-riding of the

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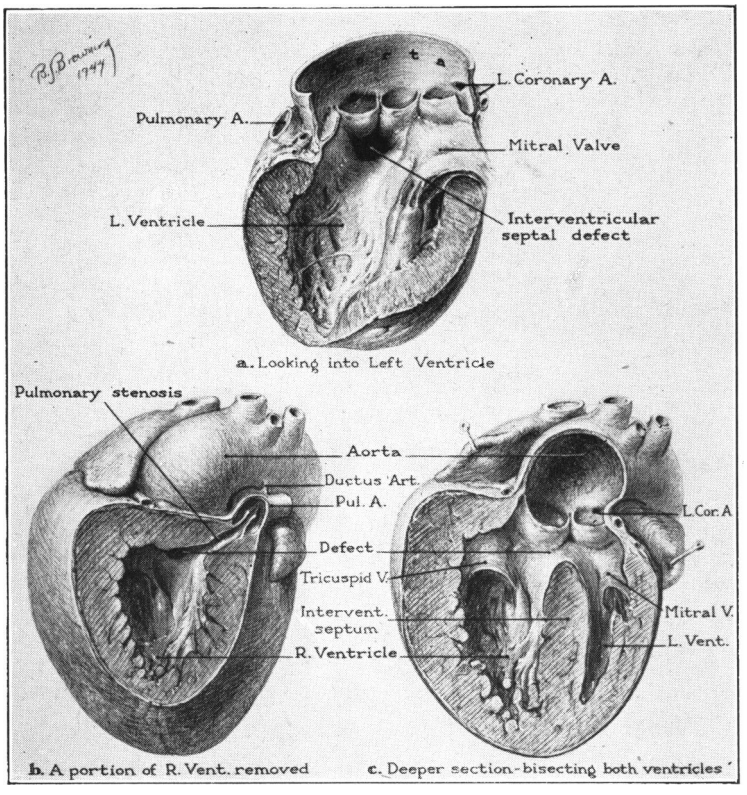

Figure 1: Tetralogy of Fallot.

aorta in a tetralogy of Fallot does not in itself cause a murmur.* The intensity of the second sound varies with the size and position of the aorta. It may be louder to the left than to the right of the sternum. Nevertheless, inasmuch as there is pulmonary stenosis, the second sound at the base is never reduplicated. Indeed the only significant auscultatory finding is the purity of the second sound at the base of the heart.

The blood pressure is narrow and often difficult to obtain.

The electrocardiogram shows a right axis deviation. Almost always the P waves in Lead II are abnormally high; frequently they are 5 mm. in height and occasionally 10 mm.

The diagnosis is established by fluoroscopy. Although in some instances the heart is slightly enlarged, it is usually within normal

* This statement is based upon clinical findings which have been repeatedly checked at autopsy.

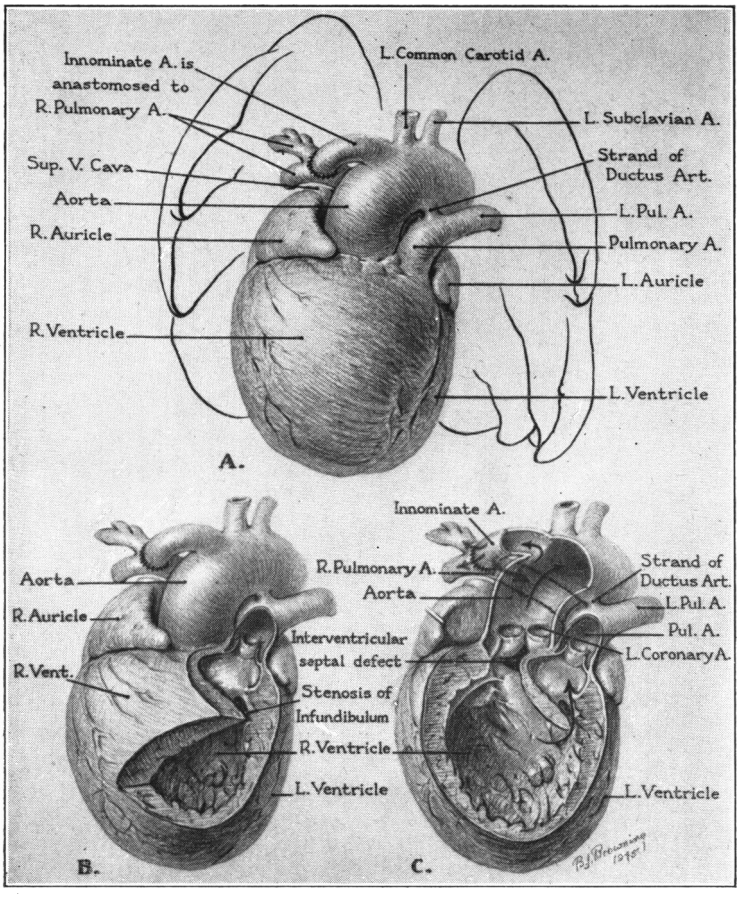

Figure 2: Tetralogy of Fallot, showing Infundibulum Stenosis and Anastomosis of Innominate Artery to Right Pulmonary Artery.

limits. The contour of the heart is characteristic. There is no fullness in the region of the pulmonary conus; consequently the shadow at the base of the heart to the left of the sternum has a concave margin (see Figure 3). Examination in the oblique views is of diagnostic help. In the left anterior oblique position, the heart is but slightly enlarged and the pulmonary window (i. e., the area below the aortic window which is usually occupied by the pulmonary artery) is abnormally clear. In the right anterior oblique position, the upper margin of the cardiac silhouette shows a concavity at the junction of the aorta with the right ventricle.

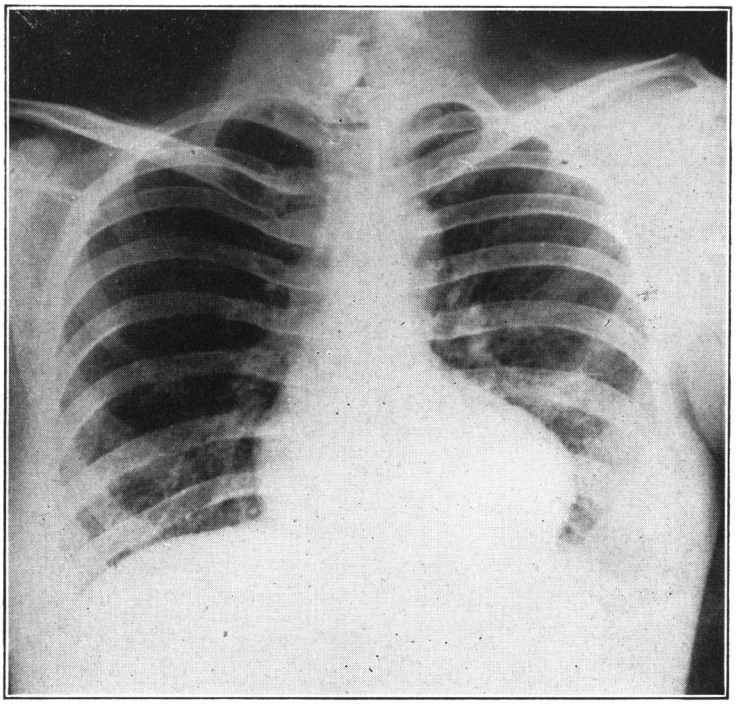

Figure 3: Tetralogy of Fallot with Left Aortic Arch.

The hilar shadows are of prime importance. Owing to the diminished pulmonary blood flow, the hilar shadows are usually minimal. Occasionally, if the principle pathway of the collateral circulation is by way of the posterior mediastinal arteries these shadows become dense.

The contour of the heart characteristic of a tetralogy of Fallot is easily differentiated from that in which there is fullness of the pulmonary conus (see Figure 4). Such a contour is indicative of a large normally placed pulmonary artery and usually connotes excessive circulation to the lungs. Not infrequently upon fluoroscopy a hilar dance is visible. Dancing hilar shadows are caused by pulsations in the pulmonary vessels and are indicative of excessive pulmonary blood flow to the lungs. Hence such shadows are a contraindication for operation.

The x-ray contour shown in Figure 5 is less readily differentiated from that of a tetralogy of Fallot. However, upon fluoroscopy after the observer's eyes were fully accommodated, there were expansile

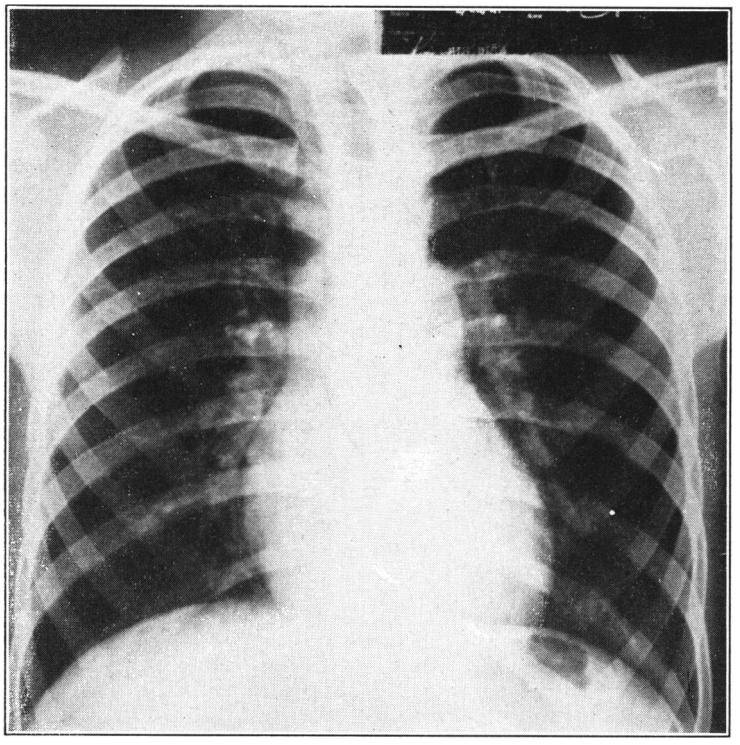

Figure 4: Eisenmenger Complex.

pulsations in both hilar regions. In such cases, even though the patient is cyanotic, operation is contraindicated.

Although not essential for the diagnosis of a tetralogy of Fallot, prior to operation it is important to ascertain the direction of the aortic arch. A right aortic arch occurs in 20 to 25 per cent of all cases of a tetralogy of Fallot. Under such circumstances, the innominate artery lies to the left of the sternum. Furthermore, the subclavian artery is given off the innominate artery at a better angle for anastomosis than it is from the arch of the aorta. Therefore the operation is best performed on the opposite side to that upon which the aorta arches.

The position of the aortic arch is determined by x-ray and fluoroscopy.[3] With a normal left aortic arch, the aortic knob is usually visible upon the left. Upon the delineation of the esophagus with a barium

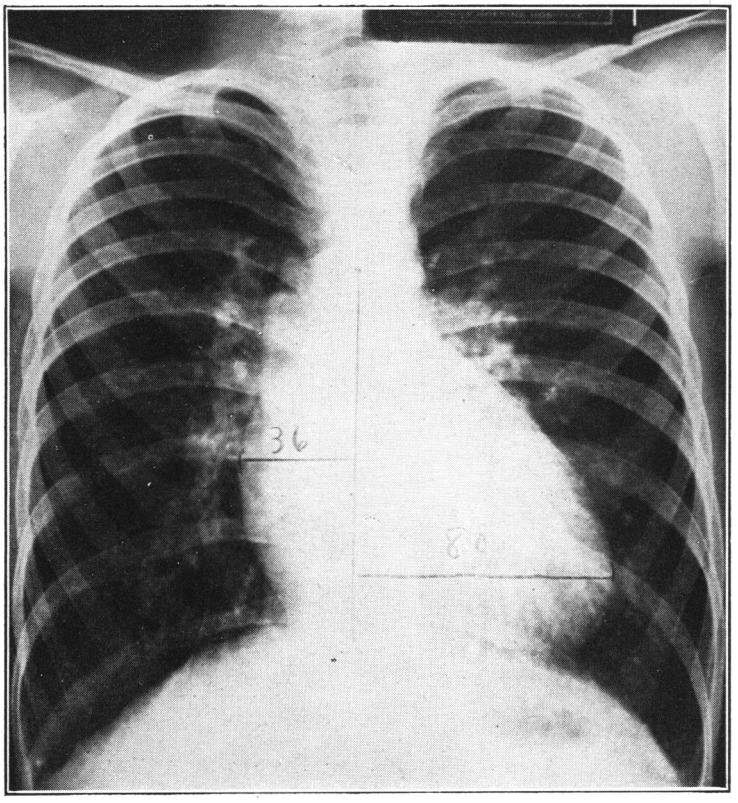

Figure 5: Heart with a Contour similar to Tetralogy of Fallot, but with Pulsations in the Hilar Regions.

opaque mixture, the esophagus descends in the mid-line and is indented by the aorta on its left margin to the right. Examination in the oblique positions aids in determining the course of the aorta. In the right anterior oblique position, the esophagus huugs the cardiac shadow and is slightly displaced posteriorly by the arch of the aorta; in the left anterior oblique position, the esophagus descends independently from the aorta. When the aorta arches to the right and descends upon the right, the aortic knob lies within the shadow cast by the superior vena cava; consequently in the A-P position the aortic knob is seldom visible. The esophagus is deviated to the left. It usually lies at the extreme left margin of the great vessels and its right border is indented to the left by the aorta (see Figure 6). If the esophagus is displaced backward by

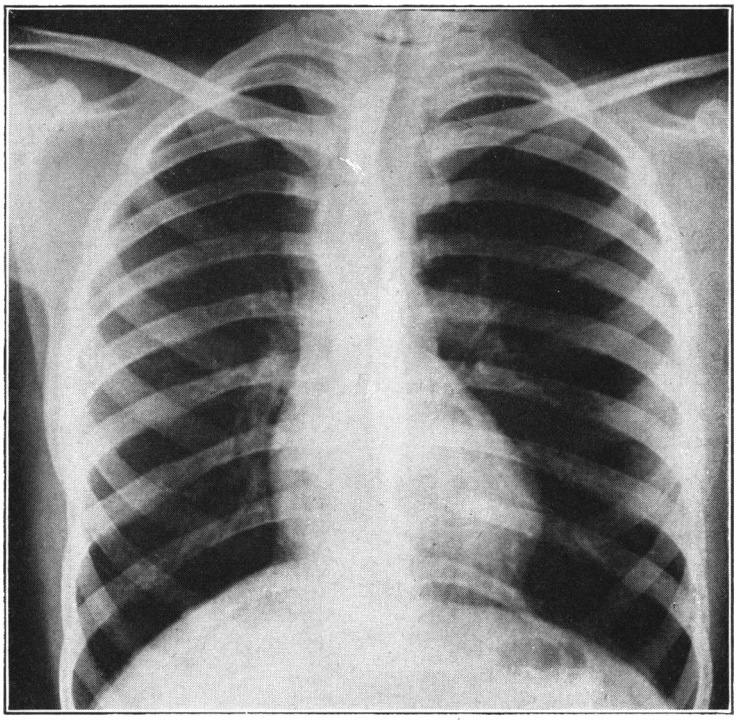

Figure 6: Tetralogy of Fallot with a Right Aortic Arch (A.P. View).

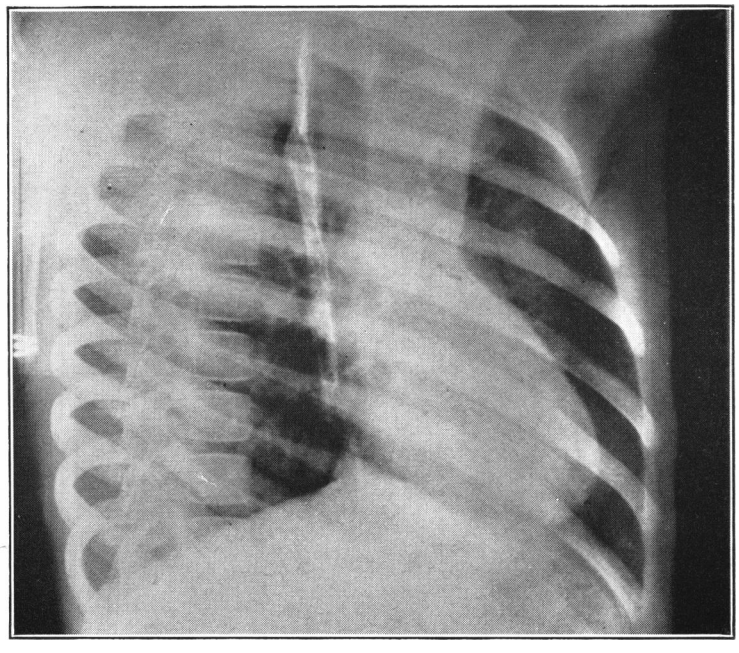

Figure 7: Tetralogy of Fallot with Right Aortic Arch (Right-Anterior Oblique View).

the aorta the displacement is visible in the left anterior oblique position. Even if this does not occur, the esophagus in the right anterior oblique position descends independently from the heart and the aorta (see Figure 7).

The operation is designed to increase the circulation to the lungs by the creation of an artificial ductus arteriosus.[4,5] This is accomplished by the anastomosis of the proximal end of either the subclavian artery, the innominate artery, or the common carotid artery to the side of the right or left main pulmonary artery. Such an operation is described as an end-to-side anastomosis. Collateral circulation is relied upon to carry the blood to the part which has been deprived of its normal circulation. In our experience the collateral circulation to the arm is always adequate: no difficulty has resulted from the sacrifice of the subclavian artery.* When the innominate or the common carotid artery is used there is danger of cerebral thrombosis. Usually, however, this can be overcome. Whenever possible it is preferable to use the subclavian artery. Occasionally the right pulmonary artery is abnormally short and consequently it is necessary to sever the pulmonary artery and to anastomose the proximal end of the systemic artery to the distal end of the right pulmonary artery. Such an operation is spoken of as an end-to-end anastomosis.

Although an end-to-end anastomosis is technically easier than is an end-to-side anastomosis, wherever possible an end-to-side anastomosis is preferable. An end-to-side anastomosis permits the flow of blood to both lungs whereas an end-to-end anastomosis places the entire load upon one lung. Furthermore, with an end-to-side technique the two vessels used for the anastomosis do not need to be of the same size. If the end of a large vessel is anastomosed to the end of a small vessel, thrombosis is liable to occur at the site of the anastomosis. The end-to-side anastomosis has the additional advantage that if the vessel is not sufficiently large or does not increase in size with the growth of the child, it should be possible, at a future date, to perform a similar operation on the opposite side. Finally, if thrombosis occurs following an end-to-side anastomosis, the resulting condition is the same as it was prior to operation whereas in case of thrombosis of an end-to-end

* The author has heard of one case in which the arm was injured by the sacrifice of the subclavian. In that case the child had a hemiatrophy and the radial pulse was not palpable before operation. At operation the subclavian artery was of good size. Evidently if a good-sized subclavian artery does not give a pulse at the wrist, collateral circulation is not sufficient to compensate for the sacrifice of this vessel.

anastomosis, the circulation to the entire lung is cut off; this is almost inevitably fatal.

The indications for operation vary with the age of the patient and depend upon the severity of the pulmonary stenosis and upon the height of the compensatory polycythemia. Children are a better operative risk than are infants or adults. Therefore, whenever possible it is advisable to postpone operation until the patient is over two years of age and to perform it before he is fifteen years of age.

Early operation is indicated if an infant suffers from repeated attacks of paroxysmal dyspnea or the anoxemia is of such severity as to cause loss of consciousness. If, however, the general condition of the infant is good and the attacks are not severe, it is wiser to postpone operation until the child is older. Delay in sitting, crawling, or walking is not sufficient indication for operation under two years of age.

In children and young adults the indications for operation depend mainly upon the degree of incapacity of the individual and the height of the red blood cell count and the hematocrit reading. A person with an extreme degree of polycythemia, as for example, a red blood cell count of 8.5 million or above, or a hematocrit reading of over 80, is in danger of cerebral thrombosis. Successful operation causes the red blood cell count, the level of the hemoglobin, and the hematocrit reading to return to normal and thus eliminates this danger. Children who are markedly incapacitated, even though they do not have a polycythemia, can be greatly benefited by operation. Such children have a very low oxygen saturation of the arterial blood or the oxygen saturation of the arterial blood drops markedly with exercise. A child with an arterial oxygen saturation of 30 per cent usually can walk only a few feet. If the oxygen saturation of the arterial blood drops markedly with exercise it is a definite indication for operation. In one instance, that of a four year old boy, the oxygen saturation of the arterial blood dropped from 41.2 to 15.2 per cent upon climbing two steps, three times. Indeed if the oxygen saturation drops from 80 to 50 per cent on exertion, that, too, is an indication for operation.

If there is evidence that, in the future, operation will be necessary, it is advisable, whenever possible, to operate before 12 or 14 years of age. The tissues of an adult are less resilient than those of a child and the lungs do not adjust as readily to the increased circulation.

The choice of the vessel to be used at operation depends upon the

size of the vessels and the severity of the reduction of the pulmonary blood flow. In infants the subclavian artery is a tiny vessel; therefore the use of the innominate artery is almost invariably indicated. On the other hand, in patients over fourteen years of age the use of the innominate artery is almost always contraindicted; it places too great a strain upon the lungs. Therefore it is only in children that the selection of the vessel is of concern. Whenever possible the use of the subclavian artery is preferable to the innominate artery because as previously mentioned, the former entails less danger of cerebral thrombosis. Recent experience has shown that in children over two years of age, the subclavian artery is usually sufficiently large to raise the oxygen saturation of the arterial blood to 70 or 80 per cent. If the child is of frail build the subclavian may be a small vessel. It is advisable under such circumstances to postpone the operation, if possible, until the child is six or seven years of age in order that the subclavian artery will be sufficiently large to give the desired increase in the pulmonary circulation.

The benefit derived from increasing the circulation to the lungs is immediately apparent. When the patient is first anesthetized and given a high concentration of oxygen, his color improves greatly. After the opening of the chest and the collapse of one lung, he becomes cyanotic. When the circulation to that lung is cut off, the child usually becomes deeply cyanotic. This is of no great concern provided the heart action remains strong. After the completion of the anastomosis, just as soon as the clamps are removed and blood flows to the lungs, one can watch the patient's color improve. As the lungs are re-expanded the color continues to improve. By the time the chest is closed and the drapes are removed and oxygen has been discontinued, the patient's lips are usually of normal color. Indeed, the color of the patient's lips and cheeks at this time generally gives an indication of the amount of benefit derived from the operation. Although the maximum arterial oxygen saturation is not immediately obtained there is an abrupt increase in the oxygen saturation of the arterial blood at the end of operation. In one case it rose from 30 to 69 per cent; in another from 24 to 63 per cent.

If the polycythemia is marked or if the cerebral circulation has been disturbed, venisection at the end of operation is a sound physiological procedure. Just as soon as the circulation to the lungs is increased, the need for the polycythemia has been removed. Venisection at this time lessens the load upon the lungs and lessens the danger of cerebral throm-

bosis. A normal adult can give 500 cc. of blood without difficulty. A child with a marked polycythemia is greatly benefited by the withdrawal of 100 to 250 cc. of blood depending upon the size of the child and the degree of the polycythemia.

The routine postoperative care includes the use of oxygen, penicillin, sedation and the careful regulation of the fluid intake. The last mentioned is the only one of these procedures which requires special consideration. The maintenance of correct fluid intake before, during and after operation is of great importance.

Inasmuch as patients with polycythemia are liable to develop cerebral thrombosis it is always important to prevent dehydration. Prior to operation these children should always receive 1500 cc. of fluid per day and young adults over 2000 cc. Care should be taken not to dehydrate the patient the night before operation. Twelve hours without fluid is liable to cause cerebral thrombosis even though the circulation to the brain is not disturbed. If the innominate artery or carotid artery is used at operation, the danger of cerebral thrombosis is increased.

There is, however, less danger of postoperative pulmonary edema and pulmonary effusion if a low fluid intake is maintained during and immediately after operation. During the operation the patient is given slow continuous intravenous fluid so that in case of hemorrhage the blood loss can be promptly replaced by plasma. Except in cases of hemorrhage, the infusion of plasma should be small in order to lessen the danger of pulmonary edema upon the release of the clamps after the completion of the anastomosis. During the first twenty-four hours (midnight to midnight) of the day of operation, most infants require 750 to 900 cc. of fluid; children two to ten years of age require approximately 1000 cc. of fluid and adults should not receive over 1800 to 2000 cc. of fluid. This total fluid intake includes the fluids given during the operation (except that which is required to replace blood loss from severe hemorrhage): the fluids given intravenously, and that which is taken by mouth. Therefore, if shortly after operation fluids by mouth are well tolerated, the intravenous fluid should be decreased or discontinued.

The most serious postoperative complications are cerebral thrombosis, pleural effusion, pneumothorax, cardiac failure and suppression of renal function and thrombosis at the site of the anastomosis. The last mentioned nullifies the benefit to be derived from operation.

Cerebral thrombosis and *hemiplegia* are especially likely to occur when the circulation to the brain has been disturbed by the use of the innominate or the common carotid artery. The danger is increased by the marked polycythemia and by dehydration. The period of greatest danger is four to twenty hours after operation. The development of a hemiplegia or of paresis of any of the extremities calls for prompt treatment. Heparin causes an immediate prolongation of the clotting time and therefore is of far greater benefit than is dicoumarol. The initial dose of heparin (0.5 mgm. per kilogram of body weight) is given intravenously; this is followed by the slow continuous administration of heparin. This is best accomplished by the addition of heparin to the plasma, glucose, or saline, which is given by continuous intravenous drip. The objective is to prolong the clotting time of the blood to twenty minutes. Approximately the same amount of heparin per hour is required to maintain the increased clotting time as is initially required to raise it to a given level. The estimated amount of heparin should be combined with the amount of fluid required per hour. For example, if the patient weighs 20 kilograms and is to receive 100 cc. of fluid per hour for the next few hours, 10 mgm. of heparin is added to 100 cc. of the fluid and the clotting time is checked at the end of one half hour. Then if the fluid is to be reduced to 75 cc. of fluid the next hour, 10 mgm. of heparin should be added with the next 75 cc. of fluid provided the desired clotting time is obtained. If the clotting time is not sufficiently prolonged, more heparin is added to the solution; if it is excessively high, more fluid is added and the concentration of the heparin is thereby reduced. It is advisable to continue to use heparin for 12 to 48 hours depending upon the condition of the patient. If treatment is promptly instituted it is often possible to overcome the paralysis and prevent the development of any residual hemiplegia.

Pnemothorax may result from injury to the lung at the time of operation or from the use of too great positive pressure at the time of the re-expansion of the lung. The latter may occur on the side opposite to that of operation. Unless there is a tension pneumothorax, simple aspiration usually suffices. A tension pneumothorax may require continuous suction.

Pleural effusion is a common complication. The fluid is usually hemorrhagic. Pleural effusion occurs so frequently that a portable x-ray plate the evening after operation is a wise precaution. If there is no

demonstrable fluid eight to ten hours after operation, it is reasonably certain that there will be no serious embarrassment from a pleural effusion before morning; it does not however mean that no fluid will accumulate. Indeed, aspiration of a pleural effusion is often necessary one to two days after operation. Not infrequently repeated aspirations are needed. While there is any tendency for fluids to accumulate in the chest, care should be taken that the patient does not receive an excessive amount of fluid by mouth.

Slight increase in the size of the heart after operation is relatively common. Usually, however, once the heart has adjusted to the load placed on it by the altered circulation, there is no further cardiac enlargement. Slight engorgement of the liver frequently occurs during the first week after operation. Digitalis is helpful at this stage, but if the child responds promptly, it is seldom necessary to continue it over a period of weeks. Diuretics, especially theocalcin, are of value in cases of engorgement of the liver and are strongly indicated if the urinary output is low.

Suppression of kidney function is a common complication. The cause is not clear. Diuretic and digitalis are both helpful.

Hypertension in the early postoperative period is of frequent occurrence. It usually causes no concern; on the contrary, it improves the circulation to the brain and lessens the danger of cerebral thrombosis. It is important to remember that even though the blood pressure may be difficult to obtain before operation, it should be readily obtainable within two to three hours after operation. If the blood pressure does not rise promptly, plasma should be given or the rate of the flow of plasma should be increased.

Although there are many possible serious complications, it is remarkable how smooth the postoperative course usually is and how rapid and how great is the improvement. A liquid diet may be allowed on the day of operation and the following day a soft diet is often desired. The child is usually free of pain and comfortable without oxygen in four days time and ready for discharge in two to three weeks.

The results of the first three hundred operations are briefly summarized as follows: 54 (18 per cent) have died at, or shortly after, operation; 9 cases (3 per cent) were found to be inoperable but survived an exploratory thoractomy. Ten children (3.5 per cent) were unimproved by operation; in most of these cases thrombosis occurred

at the site of the anastomosis. Three of the patients who were unimproved, died within six months of operation. Eleven or 4.5 per cent were helped but not brilliantly improved. Two hundred and fifteen patients (71 per cent) were greatly benefited by operation. An excellent result means that the cyanosis virtually disappears; that the lips are of normal color, there may be slight cyanosis of finger tips but clubbing recedes. The oxygen saturation of the arterial blood rises to 80 per cent or above and the red blood cell count, the hemoglobin level, and hematocrit reading return to normal values. The heart, is, of course, not normal. On the contrary, murmurs are louder than before operation; and in addition to the systolic murmur, there is a loud continuous murmur heard throughout the chest, front and back. The amount of increase in the heart size has varied considerably but after the initial increase, in only one instance during the first postoperative year, has there been evidence of progressive cardiac enlargement.

Although the heart is not normal and the danger of subacute bacterial endocarditis is not eliminated, both the danger of dying from anoxemia and the danger of cerebral thrombosis have been overcome. The child is able to measure his exercise tolerance not in terms of feet but in terms of miles. Although it is too early to determine whether the child's life may or may not be prolonged by operation, it is certainly happier.

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