THE SURGICAL TREATMENT OF CONGENITAL PULMONIC STENOSIS*

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PRIOR TO THE INITIATION[1] of the work to be discussed in this paper only one attempt had been made by operative means to treat patients with pulmonic stenosis. This attempt was made by Doyen[2] in 1911, the procedure consisting of division of the constricted area with a tenotome knife. The patient died shortly after operation. In patients with congenital pulmonic stenosis the constricted or atretic area is usually not in the valve but is in the pulmonary conus itself. It is doubtful if incision or partial excision of the stenotic area would result in permanent improvement; it appears likely that the area would subsequently become stenosed again. These considerations led to an attempt to treat the condition by a different type of operation.

Many experiments[3, 4] preceded the first attempt to increase the pulmonary blood flow of patients. It was demonstrated first that one can anastomose without great risk the end of one of the arteries arising from the arch of the aorta to the side or to the distal end of one of the two pulmonary arteries of anesthetized dogs. In subsequent experiments, after a high degree of chronic arterial unsaturation had been produced, it was found that the creation of an artificial ductus arteriosus resulted in an elevation in the oxygen saturation of arterial blood.

When the problem was transferred from the experimental animal to the patient, it was thought that a moderate degree of improvement would result in the patient with pulmonic stenosis if the pulmonary blood flow were increased. The improvement in most cases has been much more striking than had been anticipated. Some of the results will be described later in this paper.

The main indication for operation is evidence of an inadequate flow of blood to the lungs. Important in the diagnosis is the absence of visible pulsations in the lung fields as observed under the fluoroscope and roentgenographic evidence that the pulmonary artery is small in size. The typical case of the "tetralogy of Fallot" should not present great difficulties in diagnosis. There are, however, many borderline cases in which it is difficult or impossible to be certain of the true nature of the condition. If under such circumstances the patient has a hopeless prognosis without an operation, Dr. Taussig and I have taken the position that an exploration is indicated. If doubt still exists after the pulmonary artery is exposed, the pressure in the artery is measured by using a needle and a water manometer. When the pressure is greater than 300 mm. of water, it is our opinion at present that an anastomosis is probably inadvisable.

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The limits as to age-groups suitable for the operation have not been established. The operation has been performed successfully on an infant of five months and on an adult of 21 years. The operation was attempted on an infant two months of age, but there was atresia of the pulmonary orifice and the artery was diminutive in size, and an anastomosis could not be performed. It is believed that the age-period of two years to ten years is the most desirable one in which to perform the operation.

Although most of the children have had visible cyanosis when at rest, one should not be misled by the absence of cyanosis if the patient has little or no tolerance to exercise. Under such circumstances the arterial oxygen saturation should be determined under basal conditions and immediately following exercise. If there is definite decrease in the saturation with exercise and if inadequate pulmonary blood flow is believed to be the cause, an operation should be seriously considered. Several of our most gratifying results have been in patients of this type.

There have been a few alterations in operative technic during the past year. Probably the greatest has been in connection with the side of the chest on which the operation is performed. It was held originally that the approach should be made on the left if one wishes to utilize a subclavian artery for the anastomosis. This view has been altered, and it is believed now that one should make the approach on the side on which the innominate is located. This is usually, but not always, the right side. If the aorta descends on the right, the innominate artery is located on the left rather than the right. Such has been the position in more than 20 of our patients. Fortunately the position of the aorta can be determined preoperatively by employing the method of Bedford and Parkinson.[5] If one makes the approach on the side on which the innominate artery is located, one may choose the innominate artery, the subclavian artery, or the carotid artery for the anastomosis depending upon the existing inadequacy of the flow of blood to the lungs and the sizes of the available vessels. The choice of vessels is not always an entirely free one because in some instances the innominate or the subclavian artery may be too short for the desired purpose and the longer vessel must be chosen. Exposure of the innominate artery not only offers a wider choice of vessels but is of advantage in case the subclavian artery is used. After an anastomosis in which the subclavian branch of the innominate is used, the subclavian artery makes an angle of approximately 90 degrees with its parent vessel. The angle is usually a much more acute one when the subclavian branch of the aorta is used. The use of the subclavian branch of the innominate is demonstrated in Figures 1 and 2.

There has been a wide variation in arteries arising from the arch of the aorta. In some instances the four arteries have arisen independently, there being no innominate artery. In several cases the right subclavian artery has arisen on the left and has reached the right arm after passing under the trachea and esophagus. Despite the multiplicity of variations there has been in every case a systemic artery which was suitable for anastomosis to a pulmonary artery.

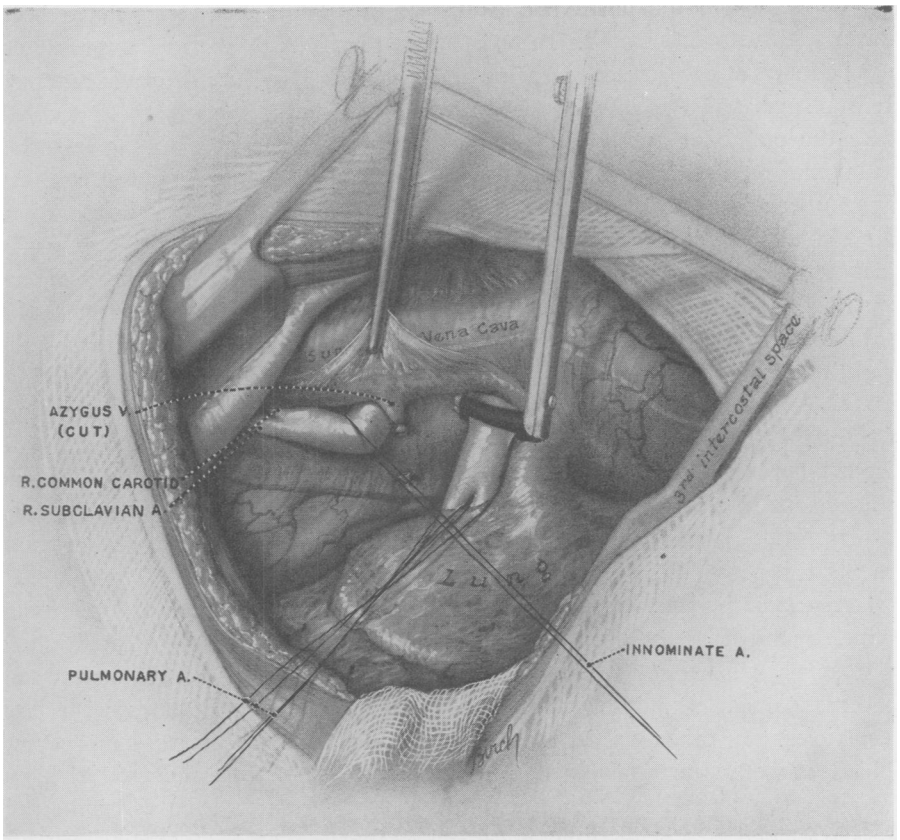

FIG. 1.—Showing the right pulmonary artery, the innominate artery, the right common carotid artery and the right subclavian artery prior to ligation and division.

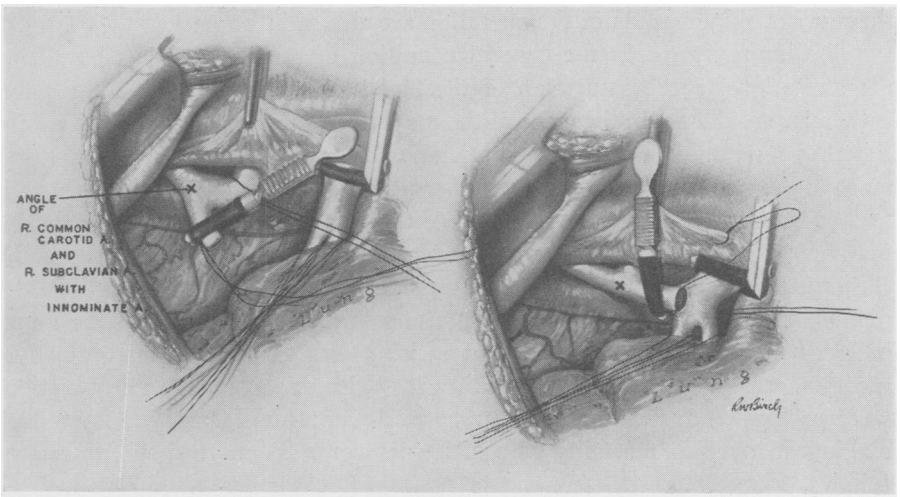

FIG. 2.—Showing the anastomosis of the proximal end of the right subclavian artery with the side of the right pulmonary artery. The transposed subclavian artery makes an angle of approximately 90 degrees with its parent vessel.

Variations in the position and size of the pulmonary artery have been fewer than was anticipated. In one patient the right pulmonary artery could not be found at the time of operation and was located with difficulty at autopsy. It was small and was inferior and posterior to its usual position. In another case the pulmonary artery was lying inferior and posterior to the superior pulmonary vein. Unfortunately, the vein was mistaken for the artery and the anastomosis was performed. Death occurred a short time after operation. In a third patient, an infant of two months with atresia of the pulmonary orifice, the right pulmonary artery was diminutive in size. The lumen of this vessel was only about one-tenth the size of that of the subclavian artery and a satisfactory anastomosis could not be performed. In a number of cases the right main pulmonary artery has been very short because of early branching. In two of the earlier operations the branch to the right upper lobe was mistaken for the main right pulmonary artery. In an occasional case in which the main right pulmonary artery is very short it may be necessary to ligate and divide the vessel and to use the distal end for an anastomosis to the end of the systemic artery. If, however, the pulmonary artery is freed of its pericardial attachments, this procedure will very rarely be necessary.

In evaluating the alterations in the oxygen saturation of arterial blood which result from the operation one should bear in mind that in addition to pulmonary stenosis or atresia, patients with the tetralogy of Fallot have an interventricular septal defect and an aorta which communicates with both ventricles. In other words, the aorta probably always receives some venous blood as well as arterial blood, and one would not expect the arterial blood to reach the normal saturation of 95 per cent, or greater, even when the pulmonary blood flow is definitely increased by the creation of an artificial ductus. In view of the septal defect and the overriding aorta it is rather surprising that the arterial saturation has risen so greatly in many of the patients. Some of the more striking results of the operation are shown in Table I.

The improvement in the general condition of the patients has paralleled the alterations in the saturation of the blood and the decrease in the polycythemia. A change in the color of the mucous membranes is usually apparent shortly after completion of the operation. Cyanosis of the nail beds disappears more slowly and the regression or disappearance of clubbing of fingers and toes requires still longer. A number of the children who could walk only a few feet prior to the operation can now walk a mile or more. Some of the children engage in fairly strenuous exercise.

Operation has been performed upon a total of 110 patients. This figure includes all patients upon whom an incision was made and hence some in whom an anastomosis was not performed. With one exception all operations were performed during the last 14 months. A total of 25 patients have died, making an over-all mortality rate of 23 per cent. A more detailed analysis follows.

In the 110 operations an anastomosis was performed between the end of a systemic vessel and the side of one of the two pulmonary arteries in 91

TABLE I

THE EFFECTS OF AN ARTIFICIAL DUCTUS ARTERIOSUS IN THE TREATMENT OF PULMONIC STENOSIS

Systemic Artery Used	Patient	Age	Art. Saturation Per Cent		Red Blood Cell Count		Hematocrit Reading (Wintrobe)		Result
			Preop.	Postop. (Time After)	Preop.	Postop. (Time After)	Preop.	Postop. (Time After)	
Innominate	A. B.	2	28.1	85.3 (111 days)	5,640,000	4,620,000 (111 days)	57.5	38.6 (55 days)	Good
Innominate	J. B.	3	45.9	74.3 (55 days)	7,530,000	5,300,000 (55 days)	68.0	56.0 (13 days)	Good
Innominate	C. C.	4	35.6	77.1 (19 days)	9,630,000	6,170,000 (19 days)	82.0	55.2 (19 days)	Good
Innominate	M. E.	26 Mos.	29.9	71.9 (20 days)	10,010,000	5,950,000 (20 days)	64.3	46.2 (20 days)	Good
Innominate	R. L.	6	22.1	74.8 (17 days)	11,260,000	6,870,000 (17 days)	73.2	57.0 (17 days)	Good
Innominate	M. M.	6	23.4	83.7 (2.5 mos.)	10,120,000	5,600,000 (2.5 mos.)	81.0	38.0 (2.5 mos.)	Good
Innominate	B. R.	12	36.3	86.7 (5 mos.)	7,660,000	4,980,000 (5 mos.)	71.0	50.0 (5 mos.)	Good
Innominate	L. S.	6	38.6	89.9 (3.5 mos.)	9,970,000	4,640,000 (3.5 mos.)	81.0	46.0 (3.5 mos.)	Good
Innominate	B. S.	2	34.0	73.2 (20 days)	9,680,000	7,530,000 (20 days)	72.2	50.2 (20 days)	Good
Carotid	R. L.	3	26.1	81.9 (15 days)	9,080,000	5,200,000 (15 days)	62.4	50.0 (15 days)	Good
Subclavian	S. B.	21	57.7	83.7 (61 days)	7,960,000	6,580,000 (61 days)	81.2	62.5 (61 days)	Good
Subclavian	R. H.	3	32.7	74.0 (18 days)	5,660,000	5,340,000 (18 days)	58.0	50.5 (18 days)	Good
Subclavian	W. L.	3	65.6	84.3 (4 mos.)	6,390,000	4,610,000 (4 mos.)	57.0	41.8 (4 mos.)	Good
Subclavian	S. V. H.	7	49.3	81.1 (3 mos.)	8,160,000	6,050,000 (3 mos.)	78.8	56.5 (3 mos.)	Good
Subclavian	C. W.	3	39.3	75.7 (35 days)	7,480,000	6,690,000 (35 days)	77.0	64.3 (35 days)	Good

patients. There were 16 deaths in this group, a mortality rate of 18 per cent. The subclavian artery was used in 46 patients, with four deaths. One of these was due to hemoptysis on the fifth day which was believed to have been caused by the use of dicumarol. The innominate artery was used in 36 patients and there were 11 deaths in this group. One of these patients was found at autopsy to have a single ventricle with insignificant pulmonary stenosis and another had a transposition of the great vessels which was combined with pulmonary stenosis. The commonest cause of death in this group was cerebral anemia or thrombosis. The end of the carotid was anastomosed to the side of one of the pulmonary arteries in nine patients. There was one death in this group, which occurred nine months after operation. Anastomosis had been performed despite the fact that the patient had a large pulmonary artery with vigorous pulsations. It is believed that this patient had the Eisenmenger complex.

An end-to-end anastomosis was attempted between the end of a systemic artery and the end of one of the two pulmonary arteries in ten patients. This method was used because the pulmonary artery was very short or small or because the patient was extremely ill and haste was necessary. There were four deaths in this series. These four patients had atresia of the pulmonary

orifice and the vessel was too small for a satisfactory anastomosis. In the future an attempt will be made under such conditions to perform the anastomosis between the end of the systemic vessel and a longitudinal opening in the side of the pulmonary artery.

In two patients the right main pulmonary artery was very short and the branch to the upper lobe was mistaken for the main vessel. The subclavian artery was anastomosed to the end of the small artery to the upper lobe. Both of these patients died. In retrospect, it is apparent that the main right pulmonary artery might have been divided and its distal end used for the anastomosis if the condition had been recognized.

Anastomosis was not performed in six patients. In four of these the pulmonary arterial pressure was high and it was thought that anastomosis was not indicated. These patients survived the exploratory operation. The remaining two patients died. In one of these the pulmonary artery was not found at operation and it was located with difficulty at the time of autopsy since it was small and in an abnormal position. In a second patient the chest was simply opened and closed because it was apparent that the patient would not withstand the operation.

In the remaining patient of those who died the right pulmonary artery was inferior and posterior to the superior pulmonary vein. The vein was mistaken for the artery and its side was anastomosed to the end of the carotid artery. The patient died shortly after completion of the operation.

It is of interest that none of the patients had empyema or mediastinitis. Severe bleeding from the anastomosis did not occur postoperatively in any case. There was no significant interference with the circulation of the arm on the side on which the subclavian artery was sacrificed or used for the anastomosis in any of the patients. Weakness or paralysis of the opposite side of the body in patients in whom the innominate or carotid artery was used either had cleared or was diminishing in all who survived the operation.

Although some of the operations were performed too recently to allow an evaluation, it appears that all of the patients with one exception who have survived the performance of an anastomosis are improved. The subclavian artery of this patient was injured by the too vigorous application of the constricting device, and it is probable that thrombosis occurred. It is believed that the artificial ductus in the remaining patients is patent although a murmur cannot be heard in at least one of them. Heart failure or *Streptococcus viridans* endarteritis has not developed thus far in any of the patients.

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DISCUSSION.—DR. ALLEN O. WHIPPLE, New York City: It is a privilege for me to pay tribute to this magnificent presentation and to the outstanding leader in the new field of surgery of shunting operations, and I cannot express this too strongly. There is one matter I want to mention, that is, the very interesting opportunity for studying the physiology of the patient with the shunting operation; whether they be cardiovascular, or whether they be thoracic or abdominal. It is a new field, one in which the disarranged physiology, in many instances total disability, is altered immediately by the shunting procedure, and I am sure that in the future this field will be explored extensively; but we shall always look back, those who have seen this, to this very remarkable presentation today.

DR. CLAUDE S. BECK, Cleveland, Ohio: I should like to congratulate Doctor Blalock and Doctor Taussig on this work. I have been trying to think of something in the surgical literature that might be related to this operation. The only suggestion that has any similarity to this operation was published by Jeger.* He proposed an operation for aortic stenosis consisting of anastomosis of the severed proximal end of the innominate artery to the cavity of the left ventricle, using a segment of jugular vein to connect these two structures. He also proposed an operation for mitral stenosis consisting of anastomosis of the distal end of a severed pulmonary vein to the cavity of the left ventricle using a segment of jugular vein to connect these two structures. These proposals were not applied to patients. The Blalock-Taussig operation is original in its conception. It was worked out in the experimental laboratory and, finally, it was applied to patients. The operation does not cure this cardiac abnormality. The circulation is not restored to normal. It provides definite benefit. I think it is one of the nicest contributions to surgery made in my lifetime.

Cardiovascular disease, in general, is responsible for about one death in three. This is the most important group of diseases that the physician treats, and yet the physician who treats diseases of the heart and blood vessels seldom if ever sees these organs in the living patient. We know that direct vision and direct manipulation can contribute to knowledge and understanding such as cannot be accomplished by any of the indirect methods of approach. I believe that progress in this field will be made by the direct approach to these structures. The Blalock-Taussig operation is based upon direct approach. There are funds available for cardiovascular research, and I hope that the internists who have charge of these funds will not forget that the direct surgical approach has something to offer and that surgical exploration should be encouraged.

DR. ELLIOTT C. CUTLER, Boston, Mass.: I rise to add my congratulations to the authors of the present paper. About 22 years ago, in a rather bungling fashion, I presented a direct attack upon the stenosed mitral valve. In the intervening time a great many signposts have been laid down making cardiac surgery far more safe than it was in those days. I have watched the work of Robert Gross, who first successfully accomplished the ligation of the patent ductus arteriosus. This new procedure in the field of cardiac surgery for children with other congenital lesions offers great stimulation to those who work in the field of cardiac surgery, and we are indebted to Doctor Blalock for his magnificent contribution.

*Jeger, Ernst: Die Chirurgie der Blutgefässe und des Herzens, Berlin, Hirschwald, 1913.