Coronary Arteriovenous Fistula; Clinical, Phonocardiographic, Angiocardiographic, and Hemodynamic Studies in Five Patients. Pediatrics, **25**:3, 1960.

- George, J. M. and D. M. Knowlan: Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery in an Adult. N. Engl. J. Med., 261:993, 1959.
- 16. Jahnke, E. J.: Cited in Case et al.⁸
- Keith, J. D.: The Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery. Brit. Heart J., 21:149, 1959.
- 18. Krause, W.: Ztschr. f. rat. Med., 24, 1865.
- Lampe, C. F. J. and A. P. M. Verheugt: Anomalous Left Coronary Artery. Adult Type. Am. Heart J., 59:769, 1960.
- Lang, E. K. and D. C. Sabiston: Coronary Arteriography in the Selection of Patients for Surgery. Radiology, 76:32, 1961.
- Lemmon, W. M., J. S. Lehman and R. A. Boyer: Suprasternal Transaortic Coronary Arteriography. Circulation, 19:47, 1959.
- Liebman, J., J. Ankenen and H. K. Hellerstein: Anomalous Left Coronary Problem in Older Children. Abstract. Circulation, 22: Part II, 751, 1962.
- Lillehei, C. W., J. R. R. Robb and M. B. Visscher: The Occurrence of Endocarditis with Valvular Deformities in Dogs with Arteriovenous Fistulae. Ann. Surg., 132:577, 1950.
- 24. Rousthoi: Uber Angiokardiographie. Vorlaufige Mitteilung. Acta radiol., 14:419, 1933.
- 25. Sabiston, D. C., C. A. Neill and H. B. Taussig:

DISCUSSION

DR. FRED TAYLOR (Charlotte): Our group in Charlotte has been interested in this operation for some time now. I think it is interesting from an historical standpoint that this procedure developed in widely separated areas independently. In 1950, Carlon, in Italy, first described this procedure as far as we can determine.

Shoemaker, in 1955, was the first to attempt this operation clinically. Unfortunately, he lost both of his patients, apparently because they had pulmonary hypertension. As Dr. Young pointed out, pulmonary hypertension is now known to be a contra-indication to the procedure. The first clinical success was in Russia in 1956 and then in 1958, Glenn and Rasmussen and our own group reported clinical successes (slide).

This is a four-year follow up of a patient who had transposition and pulmonary stenosis, showing that the shunt still works. The patient is The Direction of Blood Flow in Anomalous Left Coronary Artery Arising from the Pulmonary Artery. Circulation, 22:591, 1960.

- 26. Sabiston, D. C., S. Pelargonio and H. B. Taussig: Myocardial Infarction in Infancy: the Surgical Management of a Complication of Congenital Origin of the Left Coronary Artery from the Pulmonary Artery. J. Thor. & Cardiovasc. Surg., 40:321, 1960.
- 27. Sabiston, D. C., E. O. Theilen and D. E. Gregg: Physiologic Studies in Experimental High Output Cardiac Failure Produced by Aortic-Caval Fistula. Surgical Forum, 6:223, 1955.
- Sanger, P. W., F. H. Taylor and F. Robicsek: The Diagnosis and Treatment of Coronary Arteriovenous Fistula. Surgery, 45:344, 1959.
- Sones, F. M. and E. K. Shirey: Cine Coronary Arteriography. Modern Concepts of Cardiovascular Disease, 31:735, 1962.
- Steinberg, I., J. S. Baldwin and C. T. Dotter: Coronary Arteriovenous Fistula. Circulation, 17:372, 1958.
- Taussig, H. B.: Congenital Malformations of the Heart. The Commonwealth Fund. Harvard University Press, Cambridge, Mass., 1960.
- Upshaw, C. B.: Congenital Coronary Arteriovenous Fistula: Report of a Case with an Analysis of 73 Reported Cases. Am. Heart J., 63:399, 1962.
- Williams, J. A., et al.: Coronary Arteriography. II. Clinical Experiences with the Loop-End Catheter. N. Engl. J. Med., 262:328, 1960.

still clinically improved and doing well, attends school, can stand on his head, and put this anastomosis to a test (slide).

We, too, have used this operation in the desperately-ill cyanotic young children with tetralogy of Fallot as a palliative, temporizing procedure.

There is one technical difference we have compared with that of Dr. Young. We have preferred the end-to-end superior vena cava-pulmonary artery anastomosis, leaving the azygos vein open for decompression, while the vessels were clamped off. The azygos vein is ligated after the anastomosis is completed and the clamps removed.

It is very important that the azygos be ligated, as noted on the next slide. The azygos vein was left open in this case of tetralogy of Fallot. The child did not do as well as we had hoped. The cyanosis did not clear as much as we usually see, and she did not gain weight. This may be called an azygos steal syndrome, in which much of the blood is shunted into the inferior vena cava system. DR. WATTS R. WEBB (Jackson): We have used the procedure described by Dr. Young, particularly in instances where we believe it is important not to increase the load or left ventricular strain as is done by the classic systemic-pulmonary arterial shunt. The vena cava-pulmonary artery anastomosis is very effective in that it increases the pulmonary arterial flow without increasing the load on the left ventricle. In all probability it actually decreases the flow and work load of the left ventricle.

There are certain definite problems associated with this particular anastomosis. As this constitutes a low flow, low pressure system, thrombosis is more frequent and this, of course, is usually fatal.

The experimental work in our laboratories, particularly by Drs. Hardy, Cannon and Merck, demonstrated the effectiveness of low molecular dextran in preventing the development of thrombosis.

Before this study was instituted we had one patient who developed thrombosis in the pulmonary bed, not originating at the site of anastomosis, but within the pulmonary arterial bed distally. We do not know all the factors involved in this. In this case it was not due to a pulmonary arterial hypertension, which, of course, is an absolute contraindication to this procedure. There well may have been an unrecognized pulmonary venous hypertension; because of this we believe it is very important that one measure not only the pressures in the superior vena caval system and the pulmonary arterial system but also those in the pulmonary venous system. Low weight molecular dextran postoperatively is infused into the inferior vena caval system at a very slow rate for three or four days to prevent the overloading of the superior vena caval system. We believe that this essentially has solved the problem of early thrombosis.

The increased superior vena caval pressures are likewise a problem, and again we are very careful to monitor these patients' pressures at operation. We do not close the azygos vein until after we have performed the anastomosis, and if at the time we find the superior vena caval pressure is higher than acceptable, we leave the azygos vein open.

We have had to do this in only one patient so far, regarding this as a first stage of a twostage procedure. Fortunately, this patient has improved so markedly—becoming acyanotic and gaining weight—that we have not had to do the second stage. It is entirely possible that we may have to at a later date.

There is one other possible theoretical objection to the Glenn procedure, which is that it is not reversible. This may not be a valid objection at all because I believe it may be possible to leave the anastomosis completely open, even if at some later date it becomes feasible to do a corrective procedure.

DR. ALFRED BLALOCK (Baltimore): Obviously, I do not disagree with Dave Sabiston. I think that he and his associates have now seen something like a dozen patients with this anomalous origin of the coronary from the pulmonary; the demonstration which before was an hypothesis, that by occluding the coronary, which arises from pulmonary, one can improve the patient, I think is a very good and convincing one.

I like the opinions expressed by Julian Johnson. I think most of us are very apt to be too dogmatic in saying that everything should be done by a single method. I think Julian is right and that some of these mitral stenosis patients should be operated upon by the closed technic and others by the open technic.

I enjoyed the paper by Drs. Young, Sealy and Houck. About 15 months ago I was in Moscow. Unfortunately, my visit there followed that of Dr. DeBakey very closely, and very little attention was paid to Glenn Morrow and me, but nevertheless we had an interesting time.

The Russians like this procedure which in this country has been advocated in particular by Dr. Glenn, Dr. Paul Sanger, his associates, and no doubt, by others. In Moscow, in the Cardiovascular Institute, Drs. Bakulev and Kolesnikov and their associates, at the time of our visit 15 months ago, had operated on 77 patients in whom they had anastomosed the right pulmonary artery to the superior vena cava; these patients had either tetralogies, transpositions with pulmonary stenosis, or tricuspid atresia. Dr. Bakulev is probably the most eminent surgeon in Russia. Unfortunately, their mortality is rather high. It was in excess of 30 per cent. I do not know what the mortality in this country is.

Dr. Galankin at the Vishnevsky Institute had performed such a procedure in quite a few patients. It is his favorite operative procedure for tetralogies. I think he originated the operation in Russia. We have not had a great experience with it, and in our hands it has been limited to the treatment of tricuspid atresia.

In a good many patients I unfortunately have had to perform an end-to-end subclavian pulmonary anastomosis, because of the small size of the pulmonary artery, and those patients present a real difficulty, as far as subsequent corrective procedures are concerned.

I think that these patients with tetralogies, reported by Dr. Young, Dr. Stephenson and others, are going to present a similar difficulty. It is very easy to close a subclavian pulmonary anastomosis when doing a corrective procedure, but an endto-end anastomosis between any two vessels, whether it be the superior vena cava and pulmonary artery, or the subclavian and the pulmonary, presents a real difficulty if total correction is attempted.

DR. EDWARD F. PARKER (Charleston): I want to congratulate Dr. Johnson for the very sensible and keen analysis of his work, and for his sensible approach to the desirability of deciding whether or not to use cardiopulmonary bypass at the time of operation, rather than beforehand, in cases of secondary operation for mitral stenosis.

I also wish to comment on the paper of Dr. Young and his associates, concerning the use of the operation of superior vena cava-right pulmonary artery anastomosis in cases of tricuspid atresia. Unless or until we get to the stage of homotransplantation of the heart, this lesion at present is a totally incorrectible one. Therefore, the operation can be classed only as palliative. Since the nature of the physiologic defect is a deficiency in pulmonary blood flow, the purpose of the operation is to increase this. For this purpose, the performance of a left to right shunt is contra-indicated in many of these cases because it puts an extra load on the left ventricle. This is especially true of patients who may have been already in failure. Inasmuch as a left to right shunt may be contraindicated in many cases, and no intracardiac procedure is available, only one technic remains and that is the so-called *right-to-right* type of shunt, of which the superior vena cava anastomosis to the pulmonary artery is one type. It has always seemed to me that it would be preferable to transport the entire return to the right side of the heart to the pulmonary artery rather than just half of it. This could be done by anastomosis of the right atrial appendage directly to the pulmonary artery. The right atrium is frequently large. Of course the pulmonary artery should be of normal size, in order to be able to accommodate the increased flow. The chance of the anastomosis remaining open would seem to be small unless the flow were obligatory and this could be done by closing the atrial defect present in these cases at the same time, by the Sondergaard technic. I have performed this operation only once, but without the closure of the interatrial defect. The patient expired but even so the anastomosis was patent at the time of autopsy. With the closure of the interatrial defect, the operation of complete diversion of venous return to the heart to the pulmonary artery has always seemed a reasonable one to me.

I would like to ask Dr. Young, if he has considered the use of this technic or if he knows of its use.

Another phase of the operation for tricuspid atresia has been especially troublesome. The cause of death in these cases is again deficient pulmonary blood flow and in the operative improvement of this, regardless of what method is used, the pulmonary artery has to be partially occluded for the performance of any type of anastomosis to it. This further compromises the already inadequate flow. Therefore, in many cases, one has the sad experience of having performed a perfectly satisfactory type of procedure but the patient expires. We are now considering the use of a partial cardiopulmonary bypass in such cases in order to try to avoid this. I would like to ask Dr. Young if he has considered this and if he thinks the mortality might be lessened by its use.

DR. DENTON A. COOLEY (Houston): The use of cavopulmonary anastomosis for treatment of tetralogy of Fallot has several disadvantages which should be emphasized. The operation is destructive in the sense that it prevents subsequent total repair of the lesion. Not only is the right pulmonary artery sacrificed, but also the superior vena cava is occluded at the atrial level. This would make difficult the cannulation for subsequent open repair using cardiopulmonary bypass.

Dr. Young has stressed the point that systemic pulmonary arterial anastomosis as devised by Blalock and modified by Potts leads to enlargement of the left ventricle. If these operations are done properly, this enlargement is not extreme. Moreover, we believe that this is advantageous for subsequent two-staged repair. Our experience indicates that patients with severe cyanosis and tetralogy of Fallot are actually a better risk if they have had a previous systemic pulmonary anastomosis. We believe that development and enlargement of the left ventricle and the pulmonary vascular bed accounts for this improved condition for total correction.

In regard to Dr. Sabiston's paper our surgical experience has not included a case of anomalous origin of a coronary artery from the pulmonary artery. We have, however, had six cases of anomalous distribution of the coronary artery in which the coronary artery opened into the right ventricle. In five cases in which surgical correction was performed no fatalities resulted. One patient was a small infant in which the coronary to right ventricular fistula was not recognized and expired without surgical correction.

The technic of repair of these fistulas is of importance. If the fistula occurs relatively close to the origin of the artery from the aorta, ligation of the vessel proximal and distal to the fistula will cure the fistula but may lead to myocardial ischemia. We were not convinced of this until we encountered evidence from two patients treated by this method. The first was a child who had electrocardiographic evidence of right ventricular ischemia after operation but who recovered completely. The other patient was a woman who several hours after operation developed cardiac arrest and ventricular fibrillation while in the recovery room. Fortunately cardiac resuscitation was successful, and she has also recovered.

On the basis of this experience we now advocate a method in which the continuity of the parent vessel is maintained while the fistula is repaired. In this technic a series of mattress sutures are placed behind the coronary artery, closing off the fistula completely. The patients who have had this method of repair have had normal recovery after operation without electrocardiographic evidence of myocardial ischemia.

DR. PAUL W. SANGER (Charlotte): I rise only to comment on Dr. Sabiston's paper. It is not actually necessary to have angio movies to demonstrate this condition. Think of it as a heart typical of A-V fistula. It is a rare instance that a fistula does empty into the left ventricle. I do not know of any congenital condition where the left ventricle is in failure. There is a very high oxygen saturation on the right side and only moderate or no elevation at all, the pressure in the right ventricle.

DR. W. GLENN YOUNG, JR. (closing): I knew this was a controversial subject, and I presented it with that in mind. The discussions of Drs. Blalock and Cooley emphasize the unsettled role of the venous shunt in the tetralogy of Fallot.

I should stress again that our use of this procedure in the tetralogy patients has been limited to those cases where we do not believe we can do an open correction with a reasonable survival rate. In those patients meeting the criteria of *high risk* the anatomic anomaly may actually be so severe as to be termed non-correctable. Whether a first stage systemic artery-pulmonary artery anastomosis will change the poor risk patient into a good risk remains to be demonstrated conclusively.

In answer to Dr. Parker, I believe the complete right heart bypass has been unsuccessful in animal experiments. The inferior vena caval hypertension resulted in splanchnic congestion and ascites. Whether this could be accomplished by a staged procedure I do not know.

In performing the caval pulmonary anastomosis we temporarily occlude the right pulmonary artery for several minutes. If the patient withstands well the period of temporary occlusion, we believe we can go ahead with the anastomosis. I believe the use of extracorporeal circulatory support during the period of occlusion has been reported.

Dr. JULIAN JOHNSON (closing): It is certainly true that many patients who have thrown emboli

before their operation for mitral stenosis are found to have no clot in the left atrium at the time of operation. Of the 16 patients upon whom we have operated under direct vision because of a history of a previous embolus, residual clot was found in only six. The other ten patients, therefore, could have been operated upon quite as well perhaps by the closed technic. If one were set up so that the heart-lung machine could be utilized on a moment's notice, it might be possible to prepare to operate upon patients with previous emboli by the closed technic and then change to the open technic in the event that some clot was found to be present in the left atrial appendage. In this fashion the ten patients in our group would have had a simple closed mitral commissurotomy and the expense of the bigger procedure would have been avoided.

It is also quite true that occasionally one finds a very bad left atrial appendage and if it should be torn the surgeon is apt to back out before the mitral valve has been opened as widely as he might like. If we were in a position to use the machine on a moment's notice, the heart lung machine could then be attached to the patient and the operation completed in a more satisfactory manner. This would, of course, necessitate operating upon the mitral valve from the left-sided approach. Many people prefer this approach. However, we are inclined to prefer the right-sided approach to the mitral valve under direct vision primarily for the reason that it gives us a little better idea of regurgitation at the level of the mitral valve without having to close the heart first to determine the degree of regurgitation.

DR. DAVID C. SABISTON, JR. (closing): I agree completely with Dr. Cooley concerning the technic of closure for the coronary fistula which is proximally located. This is an important point and bears emphasis.