BRITISH MEDICAL JOURNAL

LONDON SATURDAY AUGUST 20 1949

THE SURGERY OF PULMONARY STENOSIS*

BY

R. C. BROCK, M.S., F.R.C.S.

Thoracic Surgeon to Guy's Hospital; Surgeon to the Brompton Hospital

The relief by operation of certain forms of cyanotic congenital heart disease (*morbus caeruleus*), especially those associated with pulmonary stenosis, has aroused widespread interest during the last two or three years, and this seems a suitable time to review the present position. In addition to assessing the results and value of the usual operation for this condition (the Blalock-Taussig operation), a preliminary account will be given of an entirely new operation which has so far proved successful and which may offer certain great advantages.

Human and Clinical Problem of Congenital Pulmonary Stenosis

The problem of the "blue baby" has always been a tragic one in which the doctor has hitherto been powerless to offer relief either to the sufferer or to the distracted parents. Although the plight of these children has been well known, it is only since the prospect of relief by operation has been available that the incidence and full tragedy of the condition has really come to light: those of us to whom these patients are brought for advice have been almost overwhelmed by the numbers and have been very impressed by the large amount of suffering and human misery that has been revealed. Many of these children have been almost hidden away in all parts of the country, not through any deliberate policy of concealment, but because many are necessarily, from their disability, confined to the house or to their room for the whole or the greater part of their lives. A few are able to get about out of doors moderately well under favourable conditions; some can manage to walk a short distance (10-100 yards); a number more must be wheeled in a "pram" or carried by their parents if they are to leave the house at all.

In extreme cases the child is very blue and dyspnoeic even while sitting or lying in bed and cannot move about at all; we have seen children as old as 6 years who have never walked. Some have been able to get about in a restricted fashion for a few years, and with the steady increase in their disability they finally become completely immobilized.

The strain on the parents and on the family life is considerable. The mother can rarely if ever leave the child, who not only occupies most of her attention but may give rise to periods of intense anxiety if, as often happens, it is subject to attacks of loss of consciousness and deep cyanosis and distress in which it appears to be near to death, or may indeed die. The burden on the mother is so great that she may abandon any idea of having more children, a situation that only heightens the tragedy when

the blue child eventually dies. It is worthy of remark that the father always accompanies the mother when the child is brought for consultation; much of the carrying about of the child falls on the father, and it is in any case quite impossible for him not to be intimately involved in the management and anxiety of the grave family and domestic problem that the disabled child constitutes.

The more severely afflicted children die young; the mortality in the first few years of life is high. A number of children reach the early or even the late "teens," but usually with increasing disability. It often happens that a bright or intelligent boy or girl who has done well at school and even won scholarships becomes increasingly handicapped by the physical disability that prevents the fulfilment of what might otherwise be a promising and useful career. Only few patients are seen in the twenties who are able to carry out any useful occupation or employment.

The prognosis is poor, and this is strikingly shown by an analysis of the age of patients with cyanotic heart disease who have attended the congenital heart clinic at Guy's Hospital, which is under the care of Dr. Maurice Campbell. An analysis of the age incidence in 340 patients seen by Dr. Campbell over a period of some 15 months shows a rapid falling off which admits of no other general conclusion than that the prognosis is bad and becomes worse as the years pass. Campbell sums it up by stating that only one in two children is likely to survive till the age of 7 years; only one in four is likely to reach the age of 14, and less than one in 10 will reach 21. Moreover, 24 of these patients died while awaiting admission to hospital. These observations of Campbell's not only are of value to us in assessing the gravity and prognosis of the disease, but are also of assistance to the parent in making their difficult decision whether to agree to submit their child to the dangers of an operation.

The Place of Surgery

Although operation on these cases carries a considerable risk, the risk is in most cases far less than that of leaving the condition untreated. Moreover, operation alone offers a prospect of relief, and, as will be shown later in a discussion of the results, it offers a very good chance of striking and dramatic relief. It is now fair to say that surgery has fully justified itself by its results.

It is scarcely necessary to emphasize that operation in these cases is both difficult and dangerous. It is a type of surgery that is not for the lone operator. The problems involved are so numerous and complex that teamwork is essential if good results are to be obtained. It is not enough for the surgeon to acquaint himself merely

*The Alexander Simpson-Smith Lecture, delivered at the Medical Society of London on June 15.

with the technical details of vascular anastomosis and to think that he can then embark upon operations with safety and success. Team-work is of course essential in the operating theatre, where in addition to the surgical and nursing assistants the anaesthetist plays a part of fundamental importance which deserves a special tribute. Rink, Helliwell, and Hutton (1948) have recently published an account of their anaesthetic experiences.

Even more important, however, than team-work in the theatre is team-work before the operation, for much must be done in the diagnosis and selection of the correct cases for operation. Although it is almost a fashion to-day to speak of the need for team-work, it is difficult to over-emphasize the need for it in the problem before us. Close co-operation with a cardiologist is self-evident; good radiology is essential, and we are fortunate at Guy's Hospital in having the valuable and whole-hearted co-operation of Dr. T. Hills in this; the physiologist and clinical research worker can contribute much in the study and application of the various tests and investigations of respiratory and circulatory efficiency; the anatomist and the pathologist are also valuable helpers. At Guy's Hospital there is a body of some 15 people actively engaged in the management of these cases, and all play their part in the efficiency and success of the service.

DIAGNOSIS AND METHODS OF INVESTIGATION

Until recently there was little general interest in the differential diagnosis of cyanotic heart disease, although a few enthusiasts have made invaluable studies and presentations of these cases. Until the advent of surgery great accuracy in diagnosis was rather an academic refinement of interest to the occasional clinician, as to the pathologist. Once the possibility of relief by operation arises it becomes essential to make the diagnosis as complete as possible before operation so as to avoid the hopeless case and to recognize the ones that can be helped. Careful clinical assessment, aided by routine radiography, is clearly of first importance, and for complete accounts of the clinical presentation it is best to consult the book by Helen Taussig (1947) or the comprehensive article by Campbell (1948). To arrive at a more exact diagnosis, especially in the very difficult cases, it is necessary to use cardiac catheterization and angiocardiology, both of which afford very valuable information.

The Blalock-Taussig Postulate

In considering a case for surgery it is of fundamental importance to realize what one seeks to achieve by operation. Blalock and Taussig (1945) have pointed out that the cyanosis is due to two things—namely, mixing of venous and arterial blood and an inadequate supply of blood to the lungs. The most important element of their contribution is the conception of the part played by the second feature—an inadequate pulmonary blood flow. As a result of this they postulated that the cyanosis and disability could be relieved by improving the blood supply to the lungs by anastomosing a systemic artery to a pulmonary artery. The proof of the correctness of their postulate lies in the good results of their operation: cyanosis is often completely relieved and is always greatly lessened when a satisfactory anastomosis is possible; the disability is also greatly relieved. As the venous-arterial shunt persists it may be impossible, except in the slighter cases, to relieve the cyanosis completely, at any rate after exercise.

It follows from this that the most important matter to be considered in the assessment of a case is in regard to

the pulmonary blood flow; we must try to determine whether this is decreased or not.

Radiography and Radioscopy

Radiography and radioscopy are of first importance, for by their use it is possible at once to exclude those cases in which there is evidence of a plentiful or excessive blood flow to the lungs as shown by the degree of vascularity of the lung fields and visible pulsation of the pulmonary arteries. Atrial septal defect and perhaps Eisenmenger's complex are the common examples of this; transposition of the great vessels may present a similar picture.

In contrast, a typical case of Fallot's tetralogy has clear lung fields due to poor vascularity, and there is no visible pulsation in the hilar vessels. Careful radioscopy should give the necessary information, although in a doubtful case a direct reading of the pressure in the pulmonary artery will be decisive. This can be done during cardiac catheterization.

Cardiac Catheterization

A catheter passed along a vein into the right atrium can also be manipulated into the right ventricle and thence into the pulmonary artery and into its right and left branches. If septal defects are present the catheter may pass into the left atrium and then into a pulmonary vein, or from the right ventricle into the aorta (when the aorta is overriding). Pressure readings are taken in the various positions of the catheter, and also blood samples for estimating the oxygen content. In this way a great deal of valuable information is obtained. For instance, in pulmonary stenosis the pressure in the right ventricle is raised but that in the pulmonary artery is very low. As mentioned above, in Eisenmenger's complex the pulmonary arterial pressure will be high. By noting the exact level at which the drop in pulmonary blood pressure occurs it may be possible to obtain evidence concerning the site of the pulmonary stenosis—for example, whether it is valvular or infundibular.

The information afforded by cardiac catheterization is essentially complementary to other investigations and especially to angiocardiology.

Angiocardiology

This examination is of great value, and our policy at Guy's is to use it on all patients unless there is a grave contraindication. For instance, it is occasionally waived in the case of a small child who has severe cyanosis and in whom it is felt that the examination involves a considerable extra risk. Otherwise we feel that, quite apart from the diagnostic value of the test, the anatomical information it affords the surgeon is indispensable and of a character that is otherwise revealed only by thoracotomy.

It is easy, by angiocardigrams, to demonstrate the presence of a right-to-left shunt, because the aorta and its branches may be opacified after two to three seconds, simultaneously with the pulmonary arteries. Indeed, when there is a large shunt and a severe degree of pulmonary stenosis most of the contrast medium passes rapidly into the systemic circulation and so little enters the pulmonary circulation that the pulmonary arteries may be poorly shown.

By studying the anatomical arrangement of the great vessels the surgeon can form an idea whether they are likely to be suitable for making an anastomosis. It may, for instance, be shown that the aorta is very high and the subclavian artery so short that it could not be brought

down to meet the pulmonary artery; in such a case the surgeon would know that he had to use the aorta itself for the anastomosis (Potts's operation: Potts *et al.*, 1946). It is also valuable to know if both pulmonary arteries are present or whether one is absent or so small as to be useless. Conversely, if there is no deficiency of the pulmonary circulation, or even an excessive blood supply, this also will be revealed by the angiocardiograms.

SURGICAL METHODS

Having satisfied the first postulates—namely, that there are a deficient blood supply to the lungs, a pulmonary artery to receive the blood, and a suitable systemic artery to provide it—it is necessary to consider the most suitable age for operation and the most suitable type of procedure.

Blalock does not recommend operation before the age of 2 years unless the patient's condition is so grave that he is unlikely to survive long without it. The drawback to operating on young children is that the vessels may be too small to allow an efficient anastomosis to be made, or the anastomosis may cease to be adequate as the child grows up. Our policy at Guy's has been not to operate much before the age of 5 years unless for special reasons. This is partly because we feel that the older children on our waiting-list cannot with safety wait as long as most of the younger children can. The youngest child in our series was aged 2 years 11 months. Blalock also prefers not to operate on a patient over 20 years of age if possible. With this we strongly agree, for in these older patients the operative risks are far greater. The oldest patient we have operated on successfully was aged 29 years.

Certain general technical features of the operation need brief discussion, and for a fuller consideration I would advise reference to the article by Baker, Brock, Campbell, and Suzman (1949). Most of the points raised introduce important practical problems that should concern the surgeon undertaking this work.

Blalock's Operation or the Potts Modification

The Potts-Smith modification of aortic-pulmonary anastomosis has attracted enough attention to make it desirable to discuss its advantages and disadvantages first. It does not introduce a new principle, but is a technical modification based upon the original principles laid down by Blalock and Taussig. They postulated that certain cases of cyanotic congenital heart disease in which pulmonary stenosis existed could be improved by anastomosing a systemic artery to a pulmonary artery in order to increase the blood flow to the lungs, and in their preliminary discussions mentioned the use of the aorta as a possibility in place of one of its branches. It remained for Potts and his colleagues to introduce the ingenious clamp that made this possible. There has been a tendency on the part of a number of surgeons to use the Potts operation because it seems to be quicker and easier than the Blalock operation. It is true that Potts's operation may be preferable in a small child in whom the aortic branches are too small, or indeed in any patient in whom they are too small or too short. Moreover, it is possible to vary the size of the stoma at will. On the other hand, if it is possible to use the subclavian artery to make an effective anastomosis it would appear to be unjustifiable to choose deliberately to use the aorta instead. It cannot be good surgery to interfere with a structure of the importance of the aorta if it can be avoided. If any mishap occurs while the subclavian artery is being used it should be possible to control the situation by ligating the artery. If trouble occurs when

the aorta is used it may not be possible to retrieve the situation.

There would seem to be a very real danger in the size of the stoma that is made when aortic-pulmonary anastomosis is used; if it is made too large a great extra strain is thrown on the heart. Acute pulmonary oedema may appear soon after the operation, or congestive heart failure may supervene later. I have already seen one case of severe congestive heart failure after the operation.

The great advantage of the Potts operation lies in those cases in which the subclavian artery is too small or too short and in which it has been suggested that the carotid or innominate artery should be used. The use of the aorta avoids interfering with these vessels, and thus spares the patient the greatly increased mortality (33%) and also the danger of severe paralysis from cerebral ischaemia. The use of the carotid or innominate artery is no longer justified in view of the high mortality and morbidity that have occurred.

Antero-lateral or Postero-lateral Incision

Blalock's practice of using the antero-lateral intercostal incision was followed almost exclusively in our first 50 cases, but has been departed from since then. In a very ill patient the antero-lateral approach may throw a smaller strain on the lungs and circulation than a postero-lateral. In general, however, the exposure provided by the antero-lateral intercostal incision may be rather cramped, and if the ribs are awkwardly shaped so as to make a high narrow chest the difficulties of dissection and ligation of the highest branches of the subclavian artery may be extreme. In such cases the exposure commits one to a set of circumstances that really constitutes faulty surgery.

My present practice is to use the antero-lateral approach only in those cases in which it may be necessary to operate upon the right ventricle itself. For all other cases a postero-lateral incision is used, with resection of the whole length of the fourth rib from transverse process to costal cartilage. By resecting a rib instead of incising the intercostal space there is much less bleeding at the time and less risk of oozing afterwards; post-operative pain is also lessened by avoiding the need for pericostal sutures. The exposure afforded by the long postero-lateral approach is a great advantage and allows a much more easy, rapid, and safe exposure of the vessels, and inspires the surgeon with much greater confidence that he has more complete control of the situation. The performance of the actual anastomosis is rather easier than from the front. Moreover, the postero-lateral incision allows use of either the subclavian artery or the aorta according to the conditions found. I have even, on one occasion, performed a direct operation on the right ventricle through a postero-lateral incision, but it is unusual to be able to do this with ease and safety; an anterior incision is preferable.

Right or Left Side

Blalock's earlier recommendation was to use the side opposite to the aortic arch in all but adults or patients in the later teens—that is, when the aorta is left-sided a right-sided incision should be made. This is in order to use the subclavian artery as it arises from the innominate artery; when turned down it forms an easy T-junction with the parent trunk, whereas if the subclavian artery is used as it arises from the aorta it may be kinked and flattened. In his latest paper Blalock (1948) states that he uses the right side (when the aorta is left-sided) in all patients between the ages of 2 and 12, but prefers the left approach in children under 2 and in patients over 12 who have attained

most of their growth or who are more than 5 feet (1.52 metres) in height.

After using the right-sided approach in nearly 50 cases I have abandoned it except in exceptional circumstances. In spite of increased familiarity and experience with the operation the dissection of the systemic artery on the right side may be extremely difficult, not a little dangerous, and certainly very exacting; especially so when the superior vena cava is large and dilated. If the innominate artery branches high up, the subclavian may be too short to reach the pulmonary artery; hence an anastomosis is possible only if the carotid artery is divided or the innominate artery is used. As already mentioned, this is no longer considered justifiable.

Another disadvantage of the right-sided approach is that it does not permit a proper examination of the heart and origin of the pulmonary artery and aorta in order to make as complete a diagnosis as possible. When the left side is used for the approach a very thorough examination can be made if the pericardium is opened.

In no case that I have so far done from the left side has the subclavian artery been so kinked or flattened that it jeopardizes the efficiency of the anastomosis. In any case in which the artery is found to be too short to use without tension, kinking, and flattening I either employ direct end-to-end anastomosis between the subclavian artery and the pulmonary artery or use the Potts operation of aortic-pulmonary anastomosis.

Post-operative Haemorrhage

We have found that there is a great tendency for bleeding to occur into the chest after operation in older patients who are severely cyanosed and polycythaemic and in younger patients with complete pulmonary atresia who have been submitted to exploration. This tendency is extremely dangerous, and severe bleeding occurs even after every care has been taken with haemostasis. The patients may appear to be progressing satisfactorily, and then collapse and die within a few minutes. The condition is insidious and rapid in its development. The best safeguard is the use of an intercostal drain which enables the bleeding to be recognized early and spares the patient the cumulative mechanical effects of the haemothorax. Even frequent examinations and radiography may not prevent rapid deterioration, especially during the night hours.

In the average young patient we have not, until recently, used a drainage-tube, because usually one or two aspirations are all that is needed and often aspiration is never wanted. However, our 99th patient, a boy of 7 years in whom operation was straightforward, died suddenly six hours later from the same cause; we therefore now make it a rule to drain every case.

These operations can be difficult and very exacting, demanding all one's patience, skill, and endurance; they may be made more anxious by deterioration of the patient's general condition. Nevertheless, we have found that it is best to press on quietly even when things seem almost desperate, and not to give up until it becomes quite clear that success is impossible. This applies not only to anatomical difficulties but to physiological ones as well. On several occasions the anaesthetist has reported the patient's condition as very grave, and the temptation to abandon the operation has been great. In almost all of these cases continuance with the operation after a short wait has been rewarded by final success.

The most striking example of the soundness of this policy came in the case of a very ill child of 5, who was deeply

cyanosed, with severe recurrent pain in the chest even when resting in bed; she was incapable of any activity at all. During induction of anaesthesia her breathing stopped, and her heart also. The heart's action was restored after intracardiac injection of adrenaline. Spontaneous respiration did not return for over 40 minutes. Her condition was of course desperate. At the end of an hour her condition had begun to improve and it was decided to proceed with the operation. This was completed with success. The child made an excellent recovery and has continued to do well; she walks and runs about easily, a thing she was quite unable to do before.

Results

In his latest paper Blalock (1948) reports 610 cases treated by operation in which the overall mortality was 17%. The mortality ranged from 9% when the subclavian artery was used to 33% with the carotid or innominate. In the first 50 cases from Guy's (Baker, Brock, Campbell, and Suzman, 1949) seven deaths occurred, a mortality of 14%. The total number of cases in which the Blalock or Potts operation has been done or attempted at Guy's is now 83, and 13 of these patients died—a mortality of 15.5%. Among these 83 were many severely ill patients who were very poor risks; also included were a number in the late teens and the twenties.

Blalock states that the result was excellent in about 75% of his cases. The first 50 of our cases have been carefully followed and assessed by Campbell, who states that, in addition to the seven who died, in three there was no improvement and in four the result was only "fair." "This leaves 33 (66%) in which the result was almost perfect. The patients' capacity was enormously increased, up to walking several miles instead of a few hundred yards, or getting about all day instead of being an invalid mainly in the house. To the parents the improvement seemed almost miraculous."

The results in the good cases are indeed strikingly dramatic; in addition to the banishment of disability the cyanosis is usually changed to a good normal pink, although in some a mild cyanosis remains or can be noticed after exercise. It should be realized that the right-to-left shunt persists, and so it is not surprising that some degree of cyanosis may remain in the more severe cases, especially after exercise.

It would be possible to cite case after case of really dramatic improvement in the amount of activity possible. A typical case may be mentioned of a boy of 14 years who could walk only some 20 yards before becoming distressed and squatting down. Within a few months of operation he was able to walk several miles, could ride a horse, was learning to ride a motor-cycle, and is hoping to become a dirt-track rider.

It is a regular and rather heartening experience to receive letters from parents who state that they feel they must write and say what a joy and relief it is for them to watch their child setting off happily to school or playing with other children without distress, whereas before the operation such activities were quite impossible. In not a few cases a steady deterioration with increasingly frequent attacks of deep cyanosis and unconsciousness and the probability of death within a few months has been changed for apparent good health with no cyanosis or disability and complete cessation of the alarming attacks.

DIRECT ATTACK UPON THE STENOSIS

Although the Blalock-Taussig operation is brilliant in its conception and results, it has certain possible disadvantages, at any rate in theory. It is, in effect, a short-circuiting operation in which an obstruction is by-passed,

and in this respect is comparable in the soundness of its conception and the excellence of its results to the other classic short-circuiting operations in surgery such as gastrojejunostomy for pyloric obstruction.

It differs from these operations, however, in that by creating an arteriovenous leak it actually imposes a new burden upon the body; in effect it gives the patient an artificial ductus arteriosus, a condition which, when it occurs naturally, we prefer to treat by operative closure.

On the other hand, there can be little doubt that in many cases this is the lesser of two evils, and, as Blalock and Taussig themselves say, there is no justification for letting a patient suffer and die from anoxaemia to-day just because he may develop trouble from his artificial ductus arteriosus in later years. Moreover, they suggest that in the presence of the interventricular septal defect and the overriding aorta the arteriovenous leak may not have the same significance as in an otherwise normal heart.

Whatever the answer to that question is, and time alone can tell, it is certain that surgery is not static, and that we must try to assess the feasibility and efficacy of direct operations upon the pulmonary stenosis in an attempt to relieve it. When one contemplates the small hole through which all the blood to the lungs must pass, whether the obstruction be at the pulmonary valves or lower down, it seems as if surgery should be able to offer relief to this mechanical obstruction.

Valvular Stenosis

In its simplest form the pulmonary stenosis may consist of a pure valvular stenosis with no interventricular septal defect, a condition that can be recognized clinically and can be confirmed by appropriate investigations. In most of these cases cyanosis is absent or slight in early life and appears later, usually during the teens; this is probably due to the effects of an increasing obstruction which in the presence of a patent foramen ovale causes a right-to-left atrial shunt to develop. In this type of stenosis it is certain that Blalock's operation is wrong, because it merely imposes an extra strain on the left ventricle and does nothing to relieve the strain on the right ventricle.

In a certain number of cases of Fallot's tetralogy (the exact proportion as yet being unassessed) the stenosis is also valvular in type.

The correct treatment for this condition is relief of the valvular obstruction by valvulotomy, an operation which I recorded a year ago (Brock, 1948) and which I have performed with success in five cases. In one of these not enough time has elapsed to assess the final result, although so far it would appear to be good; in two the result is moderately satisfactory and in two it is excellent. The valve is divided by inserting a special valvulotome through the wall of the right ventricle and passing it up into the pulmonary artery; haemorrhage from the heart is controlled by two rows of interlocking mattress sutures.

The actual valvulotomy is formidable, perhaps alarmingly so, but is not technically very difficult. The difficulties in the way of successful operation lie in the liability of the heart to fail quite suddenly soon after the chest is opened or as soon as it is touched. This is probably directly due to the extremely small size of the pulmonary outlet; while the heart is beating efficiently the circulation is maintained, after a fashion, but as soon as any fall in peripheral pressure occurs, with a corresponding fall in cardiac output, too little blood passes through the pinhole valve orifice to maintain the circulation. The coronary circulation suffers; the heart's action deteriorates further and soon ceases altogether.

Pulmonary valvulotomy was attempted in 10 cases, but in three the heart failed soon after the chest was opened and all three patients died. In the last of these three, as it was clear that the heart was making no response to all efforts at resuscitation, valvulotomy was performed in order to relieve the strain on the right ventricle and to enable blood to pass to the lungs. The effect was dramatic, for the heart at once revived and its powerful action was fully restored. Unfortunately the circulation had been arrested too long for the brain to have escaped injury, and, although the blood pressure was well maintained, death occurred six hours later from cerebral damage. From my experience with these three cases I am convinced that when a pulmonary valvulotomy is planned one must be prepared for the heart to fail soon after it is exposed, and if this occurs it is essential to perform valvulotomy *at once* and then to concentrate on reviving the heart; for unless this is done the heart is not likely to respond at all, or not before fatal cerebral damage has occurred.

Of the remaining two deaths, one occurred on the table from heart failure after the valve had been divided, but it was a badly chosen case of a woman in the twenties with a rigid calcified valve; the other patient (a woman aged 27) died from a haemothorax some hours after operation.

One of the patients in whom the result is classed as excellent is one of the three cases previously reported (Brock, 1948; Case H 107, Case 2). This patient, a girl aged 11, is a normal colour and leads a normal life. One other of the successful cases is worthy of special mention.

The patient, a married woman aged 26 (Ref. No. 0207), was deeply cyanosed and could walk only a few yards; her condition was deteriorating rapidly and she obviously had little time to live unless something could be done to help her. She was even unable to eat an adequate amount of food because of the severe dyspnoea that followed a meal. All her symptoms have been relieved; she is an excellent colour and leads a normal life; she has gained over a stone in weight since her operation in December last. Also significant is the fact that, although she had been married for several years and wanted children, she had never conceived. Although warned against an early pregnancy, she has become pregnant. In view of her good general condition the pregnancy is being allowed to continue. It will indeed be gratifying if she is safely delivered of a live child.

Infundibular Stenosis

Although valvular stenosis does occur in Fallot's tetralogy it is certain that infundibular stenosis is more common, and the more cases one sees and operates upon the clearer it becomes that if direct relief of the obstruction is to be at all widely applicable to these cases it must include direct attack upon the infundibular stenosis itself. This is, of course, a far more complicated matter than relief of a valvular stenosis by valvulotomy, since it involves resection of a portion of the actual wall of the active ventricular muscle and not simple division of a narrowed valve at the exit of the ventricle: it has been said that even if it were possible to operate upon the infundibular obstruction it would probably recur after the operation. It has also been suggested that since we have such an effective procedure as the Blalock-Taussig operation it is scarcely justifiable to embark upon a more elaborate intracardiac one.

It has already been pointed out that surgery is not static and therefore the feasibility and efficacy of direct relief of the infundibular stenosis must be considered carefully and critically. A moment's reflection shows that if it were possible to relieve this infundibular stenosis it would be far better than short-circuiting it as in Blalock's operation. The obstruction would then be relieved directly and no

new strain would be thrown upon the heart by the creation of the artificial ductus arteriosus.

Moreover, another important advantage would be conferred by the operation. In Fallot's tetralogy the cyanosis and disability are due to two things—the pulmonary stenosis which allows too little blood to go to the lungs, and the septal defect with an overriding aorta which allows a mixing of venous with arterial blood. The Blalock operation increases the blood supply to the lungs but does not relieve the right-to-left intracardiac shunt (except in so far that it pays back into the pulmonary circulation imperfectly oxygenated blood that has failed to enter the pulmonary artery and has passed into the aorta). I wish to emphasize that these two factors are to a great extent interrelated, a fact to which insufficient attention has been given.

If the degree of pulmonary stenosis is severe, the blood, in ventricular systole, cannot pass quickly enough into the pulmonary artery and must be diverted into the overriding aorta; if the degree of pulmonary stenosis is slight, a far greater portion of the blood ejected during ventricular systole can pass into the pulmonary artery and a correspondingly smaller amount passes into the aorta (Fig. 1).

FIG. 1.—Diagrams illustrating the importance of the degree of pulmonary stenosis in determining the amount of right-to-left shunting that occurs in the presence of an overriding aorta. In *a* there is complete pulmonary atresia, and all the blood passes into the aorta. In *b* there is a high degree of pulmonary stenosis and most of the blood from the right ventricle enters the aorta. In *c*, in which the pulmonary stenosis is less or has been relieved by operation, more blood enters the pulmonary artery and correspondingly less escapes into the aorta. In *d*, in which there is no pulmonary stenosis and only a small inter-ventricular septal defect, there is no appreciable mixing.

Nature itself provides us with instances of the two extremes: on the one hand we have the example of complete pulmonary atresia in which the exit of blood from the right ventricle into the pulmonary artery is completely prevented (Fig. 1*a*); on the other hand we have pure inter-ventricular septal defect with no pulmonary stenosis ("maladie de Roger"), in which no apparent right-to-left mixing occurs (Fig. 1*d*). It is true there is no overriding aorta.

In the case of complete pulmonary atresia, if we imagine opening up even a small lumen to the pulmonary artery, instead of all the blood passing through the right-to-left

shunt some could pass into the pulmonary arteries. Conversely, if we imagine a pulmonary stenosis supervening in the "maladie de Roger," it is clear that a proportion of the blood normally ejected by the right ventricle into the pulmonary artery would pass through the septal defect, the exact amount being directly proportional to the degree of the pulmonary stenosis; if the pulmonary artery were narrowed down to complete obliteration the only exit for the right ventricular blood would be through the septal defect and thence into the aorta.

It is not unreasonable to assume that in Fallot's tetralogy there is a similar correlation between the severity of the pulmonary stenosis and the severity of the right-to-left shunt. As the years pass by, the stenosed channel becomes more rigid, warty vegetations appear, and the lumen is further diminished. As the passage to the lungs becomes more difficult, correspondingly more blood escapes into the systemic circulation, and cyanosis and disability become worse. The pulmonary obstruction functions rather like the sluice-gates of a river, regulating the amount of flow that occurs in the main stream or is diverted to a side stream. The effect of direct operation upon the pulmonary stenosis and of relieving the obstruction to the outflow of the right ventricle will be not only to increase the amount of blood going to the lungs but to lessen the amount passing through the shunt and causing venous arterial mixing (Fig. 1*c*).

The same mechanism is seen, although it functions in a more roundabout manner, in pulmonary valvular stenosis without an inter-ventricular septal defect but with a patent foramen ovale. There may be little or no cyanosis in early life, but as the stenosis becomes more severe the pressure behind the obstruction rises and the effect is eventually felt on the right atrium, with consequent passage of blood from right to left through the interatrial septum. By dividing the stenosed pulmonary valve one not only allows more blood to go to the lungs but the back-pressure effect is also relieved and the interatrial right-to-left shunt is corrected. The cyanosis and disability are thus relieved by a dual mechanism.

This correlation between the degree of the pulmonary stenosis and the amount of right-to-left shunt appears also to offer a very reasonable explanation for the severe attacks of cyanosis, often with loss of consciousness, that are apt to afflict these patients. They occur with slight exercise or even when the patient is at rest. Although in older patients the walls of the stenosed infundibular canal may become very fibrous and rigid, in most patients, and certainly in young children, there is much more muscle than fibrous tissue surrounding the narrowed channel. One of the functions of muscle, in addition to that of contraction, is to exhibit tonus, and this tonus can vary. It is not improbable that variation of tonus of the encircling muscle may affect the functional lumen of the already narrowed infundibular channel, diminishing further the blood flow to the lungs and also causing more blood to be diverted into the systemic circulation through the intracardiac shunt.

This concept of variations in the tonus of cardiac muscle resulting in corresponding variations in the distribution of blood through the various anomalous channels, even though largely theoretical, provides a satisfactory explanation for the dramatic variations in the patient's cyanosis and general condition. It is, in fact, difficult to picture a more satisfactory explanation. Moreover, in the heart of Fallot's tetralogy, as the cardiac output increases with exercise, the narrowed pulmonary canal is too small to take advantage of the greatly increased blood flow, most of which is

inevitably diverted into the systemic circulation, thus rapidly increasing the cyanosis and disability.

If these arguments are true it follows that by a direct operation on the stenosis, which relieves the obstruction to the blood flow to the lungs, we increase the flow of blood to the lungs and at the same time diminish the escape of venous blood into the systemic circulation through the intra-cardiac shunt. During exercise the pulmonary circulation is further able to avail itself of the increased cardiac output. The procedure, therefore, would appear to be superior to a simple short-circuiting operation that neither relieves the obstruction nor lessens the shunt. The only way to stop the venous-arterial shunt completely would be to close the interventricular septal defect and correct the overriding of the aorta. It would seem very difficult to achieve both these things by operation; at present we can only hope to lessen the degree of the shunting by opening up the pulmonary channel.

Observation of the types of patient presenting themselves for treatment has impressed me that it is possible to separate out two rather rough groups. Not all patients fall into these two groups; some are intermediate, but they do reveal two extremes. In one group are the very blue and disabled patients, in particular the younger children with severe heart lesions; the other group is composed of older patients, in the middle or late teens, who have less cyanosis and disability and are often bright and intelligent, have done well at school, and are held back by their cardiac condition. In the first group it is of first importance to save life. Most of the patients in it are threatened with an early death unless they are relieved in some way by operation; they are quite unable to make any use of educational facilities and are unlikely to become useful citizens. Nobody can question the justification for using the Blalock-Taussig operation to relieve these patients; the operation is life-saving and confers great benefits in the relief of severe disability.

I am always more worried about the use of the Blalock-Taussig operation in the second group—not because it will not lessen cyanosis and disability and enable the patient to make full use of his educational and mental attainments, but because of the uncertainty of the later prognosis attached to the artificial ductus arteriosus. It is for this reason that I have always felt a lurking doubt when viewing the present good result after a Blalock's operation on these patients. It would be so much more satisfactory if there was not in the background the uncertainty that in another five to ten years serious effects of the artificial ductus arteriosus will appear. It would be much better if one could feel that a bright and intelligent boy or girl of 10–18 could be spared this doubtful future.

By direct operation on and relief of the obstruction in the pulmonary channel one might be able to achieve this, and I feel that this provides another powerful argument in favour of operative relief of the infundibular stenosis; certainly so in the older and less disabled group, but by no means excluding the more severe or most severe cases.

If these arguments are correct the advantages of direct operation upon the infundibular stenosis are manifest. I am happy to be able to state that the operation is certainly possible, because I have already performed it successfully in five cases. It is as yet too early to know whether there will be a tendency for the stenosis to recur, but up to now there seems to be progressive improvement rather than regression with the passage of months. The newly made wider channel seems to be progressively increased by the improved circulation; it is of course possible that the

passage of years may tell a different story, but so far the results have been extremely encouraging.

The development of an operation of this sort must be based, among other things, on a clear conception of the surgical and pathological anatomy of the condition. A complete description of this is impossible here and must be dealt with in a separate report. It must suffice to say that there are considerable variations in the position and character of the stenosis, which may be placed a short distance below the pulmonary valves or as much as several centimetres lower down in the ventricle. Fig. 2 shows diagrammatically the position and form of most of the usual types; and Fig. 3 shows two types similar to those in Figs. 2c and 2d but with a muscular outer wall to the infundibular chamber.

Especially important is the variety seen in Fig. 2d, in which the stenosis is situated low in the ventricle and there is a large infundibular chamber separated from the main ventricular chamber by a small ostium.

This type is usually easily recognized radiologically before operation by a well-marked convex projection on the left border of the heart shadow just below the pulmonary artery; it is thrown into greater prominence in the right anterior oblique view, and the chamber itself is clearly demonstrated by angiocardiology.

In view of the ease of recognition, and the considerable clearance below the pulmonary valves afforded for operative manipulations, this was the type chosen for the first resections. This resection was done by inserting a special punch through an incision in the wall of the right ventricle below the stricture. In some cases a finger has been passed into the heart, both to examine the local state of affairs more exactly and to dilate further the pulmonary canal. In one case finger dilatation was all that was done, and a good result followed.

A complete clinical account of the five successful cases will be given elsewhere in conjunction with Dr. Maurice Campbell.

In addition to these five successful cases there was one patient who died. This was a very blue woman aged 35 who, like all these patients over the age of 20, was an

FIG. 2.—Diagrams illustrating certain forms of the commoner types of infundibular stenosis. In *a* there is a high degree of pulmonary hypoplasia with a bicuspid valve and stenosis lying just below the valve in a narrow muscular channel. In *b*, the commonest type, there is a high infundibular stenosis. In *c* there is a small infundibular chamber with a very thin fibrous wall. In *d* the thin-walled infundibular chamber is large.

FIG. 3.—These two types are similar to those shown in Figs. 2c and 2d, except that the outer wall of the infundibular chamber is muscular.

extremely bad risk; an operative diagnosis of a valvular stenosis was made and a valvulotome was passed, although as soon as it was introduced it became clear that a more complicated condition existed. She died, and post-mortem examination revealed an infundibular stenosis situated just below the valves. Although not strictly an example of infundibular resection, it was felt that this fatal case should be included in the series.

Comment upon Direct Cardiac Operation

These direct operations upon the heart involve considerable anxiety, and many difficult and complex problems arise. This is not surprising, as they constitute virtually an entirely new field of surgical endeavour. Even more than in the case of the Blalock-Taussig operation they are definitely not for the lone operator, but demand the work and attention of an enthusiastic, loyal, and courageous team. Quite apart from the technical details of the actual operation upon the heart itself, there are the abstruse problems that arise in connexion with the avoidance or treatment of cardiac arrest or of the various severe arrhythmias that may occur during the operation; in addition there is the ever-present danger of temporary or permanent damage to the brain occurring as a result of arrest of, or interference with, the cerebral circulation. The various problems are being elucidated, and answers to them are being sought and found; progress is being made all the time, and we hope that the final results will justify the efforts. It is certain that by this means alone can we settle the question of the feasibility or otherwise of direct operations upon the heart for the relief of pulmonary stenosis.

It has been suggested that all this hard, anxious, and difficult work is scarcely justified for patients with congenital cyanotic heart disease, since so many of them are poor lives in any case and do not merit the effort. Morally, of course, this attitude is quite indefensible, but apart from this it must be emphasized that the issue is a far larger one and should be recognized as such. Although at present most of our efforts are being directed towards the surgical relief of congenital heart abnormalities, there is also the much vaster and more important field of the acquired heart lesions. Up to now very little has been achieved in their surgical relief, but the knowledge and experience gained by direct cardiac surgery for the congenital cases must help the development of successful operations for acquired heart lesions. We have, in fact, already at Guy's one successful case of the operative relief of post-rheumatic mitral stenosis.

Both these operations upon the heart itself and the indirect operations of the Blalock-Taussig type are tiring and anxious procedures, involving much care, responsibility, and hard work. Fortunately they have their happier side, and we are in large part recompensed for the long and difficult hours by seeing the almost miraculous change in the children themselves and in witnessing the joy and relief of the parents when they see their children running about happily and without effort like other children.

REFERENCES

- Baker, C., Brock, R. C., Campbell, M., and Suzman, S. (1949). *Brit. Heart J.*, **11**, 170.
 Blalock, A. (1948). *Surg. Gynec. Obstet.*, **87**, 385.
 — and Taussig, H. B. (1945). *J. Amer. med. Ass.*, **128**, 189.
 Brock, R. C. (1948). *British Medical Journal*, **1**, 1121.
 Campbell, M. (1948). *Guy's Hosp. Rep.*, **97**, 1.
 Potts, W. J., Smith, S., and Gibson, S. (1946). *J. Amer. med. Ass.*, **132**, 627.
 Rink, E. H., Helliwell, P. J., and Hutton, A. M. (1948). *Guy's Hosp. Rep.*, **97**, 48.
 Taussig, H. B. (1947). *Congenital Malformations of the Heart*. New York.

SOME ASPECTS OF POLIOMYELITIS IN NEW ZEALAND

BY

J. E. CAUGHEY, M.D.N.Z., F.R.C.P., F.R.A.C.P.

(From the Auckland Hospital)

From the study of poliomyelitis in different parts of the world it seems apparent that the clinical picture of the disease varies from country to country and from time to time in a given locality. The occasional case of endemic poliomyelitis with a minimal prodromal illness and sudden onset of paralysis, as is common in the United Kingdom, is poles apart from the type of case which occurs in epidemic form in New Zealand, where there is a high mortality and the prodromal illness is often an acute influenza-like affair with marked muscle spasm. As a result, concepts of the disease and methods of treatment in the U.K. and those of New Zealand and the North American continent are widely divergent, and we have the paradox of skilled observers in the U.K. stating that they have never seen muscle tightness in poliomyelitis, whereas competent observers in other countries aver that muscle tightness occurs in every patient with poliomyelitis. Treatment has varied accordingly.

In the United Kingdom in the past, apart from occasional minor epidemics, the disease has been endemic, but there has been a rising incidence in the last two years, and it seems probable that poliomyelitis may be about to shed its cloak of endemicity and appear in full-dress epidemic form. Whether such a change is a result of a rising virulence or is due to the advent of a new strain of poliomyelitis is not yet apparent, but if severe epidemic poliomyelitis does occur in the U.K. it seems probable that the clinical picture will be different from what it has been in the past. In view of this it might be of value to place on record some aspects of an epidemic of poliomyelitis which occurred recently in New Zealand, where poliomyelitis has been endemic since 1887 but where in addition there have been major epidemics at regular intervals, as in 1916, 1925, 1937, and 1948 (Department of Health, New Zealand, 1938).

The New Zealand Epidemics

Seasonal Incidence.—In the recent epidemic in Auckland (population 354,000), 250 patients were admitted between November, 1947, and September, 1948 (Hughes, 1949), and thereafter occasional cases occurred up till April, 1949. The onset and main intensity of the epidemic occurred in the summer, in the first three months of the outbreak. Thereafter the epidemic smouldered for 14 months. Previous epidemics in New Zealand have occurred in the summer months, whereas in an epidemic among New Zealand troops in Egypt (Caughey and Porteous, 1946) the incidence was highest in the months of January and April, unlike that of other epidemics in the Northern Hemisphere, which tend to fall in the summer months, between July and October.

Age Incidence.—The 250 patients admitted comprised 188 children and 62 adults. A study of the age incidence of notified cases in this and previous epidemics (Table I) shows a steady increase in the incidence of patients over 16 years and a fall in the 0–5 age group, in keeping with the definite shift to the higher age groups noted in the poliomyelitis statistics of New York City and elsewhere.