PULMONARY VALVULOTOMY FOR THE RELIEF OF CONGENITAL PULMONARY STENOSIS

REPORT OF THREE CASES

BY

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The surgery of the heart may be said to have begun in 1896 when Rehn (1897) first successfully sutured a wound of the heart. Until a short time before this a semi-mystical, mediaeval attitude prevailed in relation to the heart, which was generally thought to be untouchable surgically. Thus even as late as 1883 Billroth declared that the surgeon who attempted to suture a wound of the heart would lose the respect of his colleagues. Rehn's epoch-making operation demonstrated conclusively that the heart of man was, in some measure at any rate, tolerant of interference. Surgeons were quick to profit by this lesson, so that within ten years Rehn was able to collect reports from the literature of 124 cases of a wound of the heart treated by suture.

The early promise of the development of direct cardiac surgery engendered by Rehn's success was not fulfilled; development there has been, but most of the progress has been by indirect methods. Operations have been conducted upon the surface of the heart and upon the great vessels near the heart, but very few indeed have been done upon the actual substance or internal structure of the heart except for injury or removal of retained foreign bodies.

The successful ligation of the patent ductus arteriosus by Gross and Hubbard (1939) and resection of aortic coarctation by Crafoord and Nylin (1945) were followed by the brilliant work of Blalock and Taussig (1945) in the treatment of *morbus caeruleus*. By anastomosing a systemic artery to the pulmonary artery Blalock was able to relieve cyanosis in those cases of "blue babies" in which an inadequate supply of blood was going to the lungs—usually those in which a pulmonary stenosis was present. Even this carefully thought out and brilliantly executed operation, which has brought relief to so many hundreds of sufferers otherwise doomed to disability and death, is an indirect operation and not a direct one upon the substance or structure of the heart itself. It is in the nature of a by-passing operation to avoid the stenotic zone.

Earlier Operations for Valvular Stenosis

In 1913 Doyen tried to divide what was thought to be a stenosed valve in a patient aged 20 suffering from pulmonary stenosis. He did this by introducing a small tenotomy knife into the right ventricle; the patient died several hours later, and necropsy showed that a subvalvular rather than a valvular stenosis existed. About the same time Tuffier (1913) dilated a stenosed aortic valve with his finger by invaginating the wall of the aorta, but he did not open the heart or the aorta; the patient was said to be improved. Souttar (1925) used a method of direct digital dilatation of the mitral valve and his patient survived, although it appears from the case report that no true mitral stenosis was present.

The most sustained effort to solve the problem of relief of valvular stenosis by direct attack on the valves was made by Cutler and his associates (Cutler and Beck, 1929); they used a tenotome in their first three cases and a cardio-valvulotome in four others. Their first patient survived four and a half years and was thought to have been improved; six other patients died soon after operation, as also did single cases reported by Allen and Graham (1922) and Pribram (1926).

These early efforts were succeeded by a long period of inaction, and in the minds of many these operations of a quarter of a century ago seem to have proved that direct surgical attack on the diseased valves of the heart is too dangerous to be practicable.

Now, we must remember that these attempts were indeed made nearly a quarter of a century ago. Since that time there have been great advances in surgery and anaesthesia. Enormous developments have been made in the whole field of intrathoracic surgical technique; great strides have been made in pre-operative and in post-operative care, particularly in regard to blood transfusion and oxygen administration; and, more recently still, chemotherapy has heralded a new era. With our present knowledge and experience of intrathoracic operations we can understand the difficulties in the way of these early pioneers. It is no discredit to them that they did not succeed; they deserve only the greatest credit and recognition for their courageous efforts. At the same time it is important to realize that this accumulated experience and development of the last 25 years has entirely altered the prospects of success, and he would be blind who did not realize that the time has now come to try the matter again. When one examines the heart post mortem in patients dying of valvular disease and contemplates the minute size of the orifice through which the whole life-blood of the body has to be forced it is impossible not to feel that this simple mechanical obstruction must be capable of relief by surgery. The relief of such mechanical derangements is one of the oldest functions of surgery, and surgery is wanting in some part until it has been able to relieve cardiac valvular stenosis. It should require no special imagination to grasp this—even a student in training could postulate it.

Objections continue to be raised that even if the stenosed valve is successfully dealt with the secondary alterations in cardiovascular mechanics may prove disabling or fatal. This is no fundamental objection to the operation; it is merely a potential difficulty that has to be assessed or overcome. By timorous meditation we arouse fear and postpone success. Meditation and planning are essential, but in such circumstances as this, faced with a problem beset with so many and obvious hazards, the first step must be to

assess the essentials. The essential fundamental is the stenosed valve which holds up the passage of blood; the relief of this stenosis must be the ideal to be aimed at. Relief by indirect methods has its place and may prove ultimately to be the wisest and best course to adopt, but until proved otherwise, relief by direct attack must be our goal.

The day must surely come when direct operations upon the substance and structure of the heart will be as firmly established and as successfully conducted as operations upon the lungs, brain, and other major systems and viscera that for so long defeated our efforts. There can be little doubt that there will be fundamental changes in technique, such as possibly those associated with temporary discontinuance of the circulation through the heart while it is being operated upon. In the meantime we must attack the problem and begin the development of a technique for intracardiac operations, for unless begun the task can never proceed.

It is with this justification, therefore, that I record three cases in which a direct operation was performed upon the pulmonary valves in an attempt to relieve a pulmonary stenosis; these three patients are alive and their cardiac condition has been improved. It is not suggested that these three cases provide the solution to the problem, but they would appear to be of sufficient importance to deserve description at this time. So far as I know, apart from the earlier attempts mentioned above, they are the only recorded examples of successful direct operations upon the heart to relieve disease within the heart, apart from the repair of injuries and the removal of foreign bodies.

I am indebted to my cardiological colleague, Dr. Maurice Campbell, for his unstinted enthusiasm, co-operation, and medical and moral support in this most difficult venture; the selection of suitable cases would not have been possible without his knowledge, skill, and partnership.

The Problem of Congenital Pulmonary Stenosis

The postulate of the Blalock-Taussig or "blue-baby" operation is that an inadequate supply of blood is going to the lungs and that by joining a systemic artery to the right or left pulmonary artery an additional amount of blood can be delivered to the lungs for oxygenation. The stenosed area is thus by-passed. The immediate results of this operation can be dramatically successful and can usually be described as excellent. It is so far not possible to assess the late results, since Blalock's first operation was done only as recently as November, 1944. Two possible drawbacks to the procedure immediately suggest themselves. First, that the fistulous communication may not grow *pari passu* with the child and thus will prove ineffective in later years: observations upon growing animals and upon patients dying after several years should provide the answer to this. Secondly, an arteriovenous leak or artificial ductus arteriosus is created—a condition for the relief of which, when it occurs as a congenital maldevelopment, an operation has been considered necessary. To this objection Blalock rightly replies that the fear of complications in the future is no justification for allowing a patient to die of anoxaemia in the present; he also believes that the existence of the patent interventricular septum, which is present in most of these patients as a part of the tetralogy of Fallot, may modify the usual drawbacks that a patent ductus arteriosus entails.

The relief of the obstruction itself by direct operation would certainly answer the second and greatest objection: an increased supply of blood would be provided to the lungs without the disadvantages of an arteriovenous leak and the doubtful ultimate prognosis that this condition carries.

It is usually stated that the stenosis in these cases is almost invariably subvalvular or infundibular and is therefore not amenable to relief. In fact this is not so; a proportion of cases of Fallot's tetralogy have a valvular stenosis either with or without a subvalvular stenosis; in addition a further proportion of cases of *morbus caeruleus* occur in which a pure pulmonary valvular stenosis is present. It is not as yet possible to state with certainty the exact proportion of cases in which a valvular or diaphragmatic type of stenosis exists; there can be no doubt, from my observations so far, that the incidence is far higher than has been supposed. Investigations are proceeding to decide this more exactly.

When a valvular stenosis exists it is of a diaphragmatic nature; the fused valves form, as it were, a septum which in time, as the patient gets older, is forced by the blood stream into a nipple-like projection with a tiny hole in its summit. It projects into the pulmonary artery much as the cervix projects into the vagina. Its formation and structure show at a glance that it is eminently suitable for relief by division; moreover, a much larger hole could be made in a very short time, in exactly the right place, than is possible with the Blalock operation, in which a long and tedious dissection is necessary to secure a systemic artery of adequate size.

Before attempting to relieve a valvular stenosis it is clearly desirable first to diagnose its presence. This diagnosis should consist of a pre-operative assessment by clinical and various accessory methods and an actual final assessment at operation. This is not the place to enlarge upon the problem of pre-operative diagnosis, which will be dealt with in a later communication.

A great deal can be learned at operation, although experience is as yet too limited for dogmatism. Time and further experience can alone provide greater efficiency. At present it must suffice to say that much can be learned from direct cardiac examination.

Direct Cardiac Examination at Operation

One disadvantage of the Blalock operation, especially if it is done from the right-hand side, is that only very limited examination of the heart is made. In most cases all that is done is that the great vessels are exposed, dissected free, and an anastomosis performed. The completeness of the pre-operative diagnosis can only be assumed, unless the patient dies and the heart can be inspected.

Clearly the best way to assess the state of the valves is to inspect them under direct vision. This formed the first problem that was dealt with. A cardioscope to allow inspection of the interior of the heart was designed and described by Allen and Graham (1922); the principle of contact visualization is used in which the blood is displaced and the instrument rests directly upon the structure to be viewed. Using an operating cardioscope constructed for me by Mr. Schrantz, of the Genito-Urinary Manufacturing Company, with his usual superb skill, I have made a number of direct inspections in living patients.

One problem is the route of cardioscopy. Passage of the instrument through the ventricular wall has such apparent disadvantages that an alternative was sought. Study of the anatomy suggested the possibility of using the left pulmonary artery, for this structure normally forms an almost direct line with the main pulmonary trunk and provides a direct route to the valves. Left pneumonectomy provided excellent opportunities to test this, and it was shown to be a practical procedure. The instrument could be passed without difficulty and the valves inspected, although the vision in a normal patient is naturally limited by the constant movement of the cusps.

The first patient with congenital pulmonary stenosis, a young man of 17, was operated upon in May, 1947. The chest was opened through a left postero-lateral incision, as this approach is necessary in order to introduce the straight instrument into the heart. The valves were inspected and were seen to be normal; the stenosis was clearly subvalvular. A Blalock operation was performed by anastomosing the left subclavian artery to the left pulmonary artery, using the hole made for the passage of the cardioscope. The whole procedure meant that the pulmonary artery was occluded for a very much longer time than is usual in the Blalock operation, and this was largely responsible for the patient's death 12 hours later. Necropsy confirmed the presence of a Fallot's tetralogy with a severe infundibular stenosis and normal valves.

The next patient, a girl of 19, sent to me by Dr. East, was diagnosed pre-operatively as having a pure pulmonary valvular stenosis; the heart was very large, chiefly due to hypertrophy of the right ventricle. At operation in November, 1947, the left pulmonary artery was found to be very small and, owing to the great size of the right ventricle, which had caused gross dilatation, it arose from the main trunk at a right-angle so that it was not possible to pass the cardioscope. Neither was it possible to perform a Blalock operation. No ill effect followed the operation. The patient died of heart failure several months later and, most unfortunately, no post-mortem examination could be made.

A third patient, a girl aged 20, had severe pulmonary stenosis, which was diagnosed clinically as being of a pure valvular type. She had had several attacks of heart failure, had gross albuminuria and oedema, and had a very large heart. She was considered unsuitable for operation, but at the express entreaty of herself, her parents, and her doctor it was agreed to make the attempt. A left postero-lateral thoracotomy was done, the pericardium was opened to permit more direct examination of the heart as in the other cases, and a confident diagnosis of valvular stenosis was made. A short pause was made while the operating cardioscope was assembled, and when one turned again to the heart it was found to have stopped. It was eventually started again by massage and injection of adrenaline, but gradually failed again. It was again revived and again began to fail. It was decided to perform cardioscopy: the instrument was passed and the presence of a diaphragmatic valvular obstruction of characteristic nipple-like shape was easily observed. The heart, however, had again stopped and could not be revived. Necropsy confirmed a pure valvular stenosis with an almost pin-hole orifice of such minute size that it was astonishing that she could have supported life for so many years; very gross hepatic cirrhosis was present from back-pressure; the kidneys showed similar gross changes from chronic passive congestion.

From the experience of these cases, and in particular of the last one, it was felt that the approach through the left pulmonary artery was unsatisfactory, or at any rate was sufficiently unsatisfactory to be discarded for the time being in order to assay the ventricular route. It is clear that in certain of these patients with a valvular stenosis occlusion of the left pulmonary artery for even a short time may not be tolerated; in addition it was felt that the conical shape of the valve made rapid and precise division from above uncertain. These experiences with three patients had shown, however, that it was possible to diagnose pulmonary valvular stenosis with some degree of confidence at operation.

In addition to direct inspection of the valves much other valuable and significant information can be obtained by direct critical external examination of the heart. Even before the pericardium is opened a tentative diagnosis can be made. When a valvular stenosis is present the stem of the pulmonary artery immediately distal to the obstruction is often dilated and forms a thin-walled aneurysm-like bulge; this can be seen through the intact pericardium and a thrill of characteristic nature can be readily felt. If the pericardium is opened a much more useful examination can be made. The sac should be incised longitudinally for

some 5-8 cm. just anterior to the phrenic nerve, and by means of an additional transverse incision at each end a flap is turned forwards; by grasping the corners of this flap with haemostats it is also possible to manipulate the heart. Attention should first be directed to the region of the sinuses of Valsalva; if the sinuses are present and fully formed it is almost certain that the valve cusps are normal. Conversely, when a valvular stenosis is present there is either a continuous collar of dilated artery just above the valve-level, or the artery shows a larger, aneurysm-like dilatation resembling the bulb-like swelling of a young onion. The lesser degree of dilatation is usually associated with a smaller pulmonary artery of lower tension, and I suspect that it implies the existence of a subvalvular stenosis as well, although as yet insufficient observations have been made to confirm this. The greater degree of dilatation suggests a more powerful jet of blood coming through the stenosis. Palpation of the first part of the artery is most suggestive of this; the thrill is quite characteristic, and is of a fine high-pitched frequency, very strictly localized at a level just above the origin of the artery, and, most significant of all, can actually be felt to be extremely localized within the central portion of the lumen. Indeed, it feels as if it were caused by a thin but very powerful jet of fluid being forced through a tiny hole; and this is exactly how it is formed. If the thin-walled dilated artery is gently compressed between the fingers the precise, localized nature of the thrill becomes more obvious, and it is even possible to feel the conical valve momentarily between each heart-beat. If the finger is now passed down on to the surface of the right ventricle it will be found that the fine, purring thrill rapidly fades.

When a subvalvular stenosis is present no such characteristic localized thrill is felt in the first part of the pulmonary artery. The thrill is coarser, as if of a lower frequency, and although it can be felt by a finger placed on the artery this is because it is conducted towards it; it is felt more strongly just below the level of the valves, and altogether its localization is less precise; it is felt over a somewhat wider area. The infundibulum may also present as a thin-walled, aneurysm-like dilatation with a coarse thrill palpable at its base and conducted distally along the pulmonary arteries. Further information should be obtainable by means of direct pressure readings at different levels, but so far this has not been done successfully on the unopened heart.

Direct external examination of the heart in this way, coupled with pre-operative assessment, should provide enough information to decide whether a valvular stenosis exists.

The final observation should be direct valvulotomy through the wall of the ventricle, and it is intended to do this. It had been planned in the three cases next to be described, but at the actual operation it was felt to be wiser to omit it, chiefly on account of the uncertainty of how the heart would tolerate the extra manoeuvre. It is, after all, a serious and anxious step to perform a cardiomyotomy, and until one is more familiar with what these abnormal hearts will tolerate in the way of manipulation and interference it seems wiser to proceed cautiously step by step. With an operation of this sort one makes certain plans beforehand, but in the tension of the most critical phase of the operation it may be found easier not to attempt too much. It is nevertheless definitely desirable to include direct cardioscopy in the operative assessment.

Although the cardioscope is provided with a cutting knife which can be advanced under direct vision it was decided to use a special valvulotome for division of the stenosed valve in these first cases. This enables the stenosis

to be located and divided almost instantaneously, whereas the operating cardioscope might well entail considerable fumbling and uncertainty. The valvulotome used has a gently curved shaft ending in a blade shaped like a spear-head and carrying a short probe-end; the two edges proximal to the probe-end are cutting; the shoulders and the retreating edges are blunt. This instrument was also made for me by Mr. Schrantz. A pair of gently curved dilating forceps with fine blades, rather like a curved sinus forceps, are also used after the valvulotome.

Case 1

Miss D. N., aged 18, had been cyanosed from birth. She walked and talked at a normal age, but always had great disability in getting about, and during the winter could not walk at all outside the house; in the summer, unless it was too hot, she could sometimes walk as much as 200 to 300 yards, although in recent years she could rarely manage more than 50 yards. She squatted as soon as she learnt to walk, and continued the habit when older, although her mother tried to stop it. She found that nothing relieved her discomfort so quickly after walking, and needed to do this in the x-ray room after feeling exhausted when standing up for screening.

Her cyanosis was fairly severe, Grade III, though it did get less with complete rest on a warm day. It would get worse with emotion as well as with exertion, and was changed greatly by a minor effort like undressing. The nails showed Grade III clubbing. She had fairly severe cyanotic attacks as a child after feeding; and at 15 she spent a period of many months in bed, when her colour was exceptionally bad and her doctor thought she was not likely to survive.

The heart was not enlarged; there was a rough systolic murmur down the left side, loudest in the pulmonary area, and a faint systolic thrill; the pulmonary second sound was faint. Screening showed clear lung fields, a left-sided aorta, and no increased pulsation in the lung roots; the heart was not generally enlarged. Electrocardiograms revealed extreme right ventricular preponderance. Blood pressure was 110/85. A blood count showed: red cells, 7,400,000; Hb, 127%. The arterial oxygen saturation at rest (Dr. Zak) was 90.9%.

A diagnosis of Fallot's tetralogy was made.

Operation (Feb. 16, 1948)

Intratracheal cyclopropane was given by Dr. E. H. Rink, and an intravenous infusion of plasma (one bottle) was administered. A left inframammary incision was made and the chest was opened through the second left interspace with division of the second costal cartilage; many collateral vessels were met with. As soon as the chest was opened the heart's action became very feeble and slow—less than 50; the general condition was poor and gave considerable anxiety. The left pulmonary artery was small, thin-walled, and of low tension, and the anaesthetist felt that the patient would not tolerate its occlusion for a Blalock operation.

Palpation of the pulmonary artery area revealed a thrill suggestive of a valvular stenosis. The pericardium was opened and the pulmonary artery was seen to be slightly dilated in its first part and showed no sinuses of Valsalva. The thrill was of a jetlike character and was situated exactly in the position of the pulmonary valves; moreover, what appeared to be a conical valve could be felt through the vessel wall. No thrill could be felt lower down. It was decided to perform valvulotomy.

Two rows of three interlocking mattress sutures of linen thread were inserted in the wall of the right ventricle, which was then incised between them; the pressure within the ventricle was 24 cm. of saline. The wall was 1.25 cm. thick. The mattress sutures were crossed and held by an assistant so as to control haemorrhage from the incision. The curved valvulotome was inserted and passed effortlessly into the pulmonary artery; a dilating forceps was then passed and opened fully. Very little blood was lost. The heart wound was closed by two linen-thread mattress sutures and the pericardium sewn to it; the heart was frequently moistened with 1% procaine solution throughout the operation.

The thrill persisted, but its character changed from a fine jetlike nature to a coarser one of lower frequency; the pulse-wave could be clearly seen within the pulmonary artery. The colour was poor at the end of the operation in spite of a good respiratory exchange.

Post-operative Course

The patient's condition was grave after the operation and her colour was bad even with oxygen. She showed little improvement until Feb. 20, when her colour was better and her blood pressure had slowly risen to 120/80. The heart was very large on Feb. 18, suggesting grave dilatation, possibly due to profound alteration in mechanics; it was much smaller by Feb. 21 and her colour had improved further. A systolic murmur could still be heard.

She made steady improvement as regards her heart, but on the evening of Feb. 24 she complained of sudden severe pain in both legs with loss of movement and sensation. Unfortunately the full significance of this was not appreciated, and I was not informed until more than 24 hours later. By this time the right leg had improved, but the left leg was cold, pulseless, pale, painful, and insensitive. It was clear that an arterial occlusion was present, probably embolic, but it was considered to be too late to justify exploration. Further improvement occurred in the legs, and all peripheral pulses could be felt on the right but not on the left. Two or three superficial ulcers appeared around the left ankle; these have since healed. The condition of the legs delayed her getting up, but her colour remained good except for occasional mild cyanosis with emotion and when she first began to get out of bed. She was allowed home on March 18, at which time her red cell count was 6,200,000, Hb 119%, and arterial oxygen saturation 79.6%.

She was seen on April 10 and had made good progress. There was no cyanosis, the finger-nails were pink, and she walked about the room without dyspnoea although there was a slight suggestion of cyanosis. She no longer squats. Her relatives are most emphatic that the relief of cyanosis and of dyspnoea is impressive. They emphasize especially the absence of dyspnoea, which had always been a marked feature even at rest.

Comment.—It is as yet too early to assess the results of operation completely in this case. The condition of the left leg has prevented full test of activity. It would appear that a sharp watch must be kept in these cases for symptoms suggesting arterial embolism and prompt action taken. There has been definite improvement in the cardiac condition, although less than hoped for. The prognosis in these older patients is less certain, and from her behaviour under anaesthesia she might not have tolerated a Blalock's operation. She would appear to be an example of Fallot's tetralogy with a valvular stenosis, although probably some degree of subvalvular stenosis exists as well.

Case 2

Gwenda B., aged 11, the youngest of eight children, had been obviously cyanosed at 6 months, though blue lips had been noted before this. She walked at 1 year and squatted as soon as she walked; she was never able to walk more than 100 yards without distress, although she could sometimes manage as much as half a mile by getting very breathless. Cyanosis was extreme, ranging from Grade III to Grade IV; the fingers showed Grade III clubbing. She was well developed physically and could walk about the ward without getting breathless.

The heart was sabot-shaped, with the apex raised off the diaphragm. The aorta was right-sided. A faint thrill was felt in the pulmonary area on one occasion; a loud systolic murmur was maximum in the pulmonary area and widely conducted. The blood pressure was 95/65; electrocardiograms showed gross right ventricular preponderance. The lung fields were clear and there was no pulsation. Red cells, 8,700,000; Hb, 156%; haematocrit, 77.5%.

A diagnosis of Fallot's tetralogy was made.

Operation (Feb. 19, 1948)

Intratracheal cyclopropane was given by Dr. Helliwell, and one bottle of plasma was infused intravenously. A left inframammary incision was made and the chest was opened through the second left intercostal space with division of the second costal cartilage. There were numerous collateral vessels; the left pulmonary artery was small. The pericardium was opened and the main pulmonary artery was also found to be small, being about 1.25 cm. in diameter; it was of low tension and there were no sinuses of Valsalva. A fine faint thrill could be felt in the position of the valves but not in the subvalvular position. After some deliberation it was decided to perform valvulotomy.

The right ventricle was opened and the valvulotome passed; it met with definite resistance at the valves, and slight pressure carried it into the pulmonary artery; dilating forceps were passed and opened widely. Free bleeding occurred from the heart, and during manipulations the pulse became poor in quality but soon improved. The cardiomyotomy incision was closed with four sutures; the pericardium was sewn down over the suture line. The general condition was fair, although the colour was poor; it had been poor at the beginning of the operation. The blood pressure was 85/55, as opposed to 90/60 at the beginning.

Post-operative Course

The patient was very blue, even in the oxygen tent, for the first 24 hours; her colour then began to improve very slowly but steadily, and by Feb. 27 was reasonably good. Her progress was then rapid, and in fact by March 1 she was out of bed and visited the next ward, where she sat at table for a birthday party with other children without any distress. She was very soon able to walk freely about the ward and even down to the x-ray department, and maintained her good pink colour. This was in marked contrast to her constant cyanosis before operation. The arterial oxygen saturation (March 22) was 81%, but she was crying and distressed during the arterial puncture.

She was discharged home on March 25, the blood count at that time being: red cells, 6 millions; Hb, 116%; haematocrit, 56. She had ceased to squat.

She was seen on June 4, when she looked well and was a good colour; her mother said that her exercise tolerance was greatly improved and she was now able to run, a thing she could never do before the operation. The mother particularly remarked upon the normal colour of the nails, which had always been blue. The child walked normally to the x-ray department for screening, but although she showed no dyspnoea a slight tinge of cyanosis could be seen in the cheeks. This is not uncommon on exercise after a successful Blalock operation.

Comment.—This would appear to be a case of Fallot's tetralogy with a valvular stenosis; the hypoplasia of the pulmonary artery strongly suggests that a degree of subvalvular stenosis is present as well. Nevertheless, the result of valvulotomy has been most encouraging. The colour and clinical improvement are as good as after a successful Blalock operation.

Case 3

Miss R. C., aged 23, had been cyanosed since birth and did not walk until 3 years old. She was never allowed to go to school and dyspnoea had always been extreme. As a child she was able to play with other children, but had to rest frequently by squatting. She has always squatted, and even sleeps propped up with her legs curled up. Until the age of 18 she was able to walk perhaps 50 yards without stopping and could go considerably further with frequent rests. About five years ago, however, she became much worse quite suddenly, and found she could do very little without extreme distress. In fact, until October, 1947, she remained almost wholly in bed or on a couch, moving only from bedroom to sitting-room; any effort, even moving in bed, provoked dyspnoea and pain across the chest. In October, 1947, she spent five weeks in the National Heart Hospital, and with encouragement found she was able to do a little more; for instance, she could just about walk the length of the ward and could sit up for meals.

She was poorly covered; finger clubbing was Grade I and cyanosis Grade I (i.e., not cyanosed at rest in bed); there was

extreme dyspnoea with rapidly developing cyanosis on walking; the blood pressure was 115/70. The heart was not enlarged, although the apex was raised a considerable distance from the diaphragm, suggesting right ventricular hypertrophy; a definite rounded shadow in the region of the "conus" suggested a minor degree of aneurysmal dilatation of the pulmonary artery. Perhaps a slight thrill could be felt in the pulmonary area; a harsh systolic murmur, loudest in the pulmonary area, was audible all over the praecordium and also at the back of the left chest. On screening, the lung fields were clear and showed no pulsation. Red cells, 6,500,000; Hb, 128%; haematocrit, 56. Circulation time (Dr. Allanby): arm to tongue, 10.5 seconds; arm to lung, 8.4 seconds (indicates right to left shunt).

Cardiac catheterization (Drs. Zak and Hollings): pressure in right ventricle, 53 mm. Hg; pressure in pulmonary artery, 13 mm. Hg. Oxygen saturation in superior vena cava, 56%; in right pulmonary artery, 67%; in femoral artery, 77%. This was taken to indicate pulmonary stenosis with a right to left shunt.

A diagnosis of Fallot's tetralogy was made, although the presence of an associated valvular stenosis was considered probable.

Operation (March 23, 1948)

Intratracheal cyclopropane was given by Dr. Helliwell. Two bottles of plasma were infused, followed by one bottle of blood. A left inframammary incision was made and the chest opened through the second interspace, with later division of the second and third costal cartilages; a considerable number of collateral vessels were encountered.

As soon as the heart was exposed the smooth globular dilatation of the pulmonary artery within the pericardium was seen; a fine jetlike thrill could also be felt. The pericardium was opened; the pulmonary artery was now seen to be dilated to a diameter of about 2.5 cm. and was thin-walled, with dark blood easily seen through it. The artery beyond the dilatation was small, lax, showed no visible pulsation, and was rather less than 1.5 cm. in diameter. There were no sinuses of Valsalva. Careful palpation again confirmed the jetlike nature of the thrill, which seemed to arise from the level of the valves; moreover, at times it seemed as if one could actually see the white cone of the valvular stenosis through the thin wall.

In addition to applying swabs wet with procaine solution to the heart 1 ml. was injected into the wall of the ventricle. A double row of mattress sutures was inserted and a small incision made to allow a curved cannula to be introduced. The pressure in the ventricle was only 5 cm. of saline, and was the same in the pulmonary artery; it was felt that this low pressure was the result of manipulation of the heart. A probe 3 mm. in diameter was now passed into the ventricle, and was felt to catch on the valves, suggesting a diaphragmatic stenosis. It was decided not to pass a cardioscope in view of the low blood pressure.

The valvulotome was now inserted, and was felt to catch on the valves and then to pass on into the pulmonary artery. After a pause the valves were further dilated with forceps. A pressure reading was again taken; this showed a mean of 15 cm. of saline in both the ventricle and the pulmonary artery, with obvious fluctuation in systole and diastole. The incision was now closed with three mattress sutures; the pericardium was not sewn to it.

The general condition at the end of the operation was satisfactory; the systolic blood pressure was 70 mm. Hg.

Post-operative Course

Although the patient began to come round at the end of the operation she became very drowsy the same evening, and on the next morning was still drowsy and had a complete left-sided hemiplegia. She was slightly blue even in the oxygen tent. Dr. McArdle, who saw her, thought the hemiplegia might be due to an embolism rather than to thrombosis.

Her general condition steadily improved and her colour became and remained a good pink. The paralysis showed no sign of improvement for over three weeks. By April 22 she was able to move the upper arm, elbow, hip, and knee, but hand and foot movements were still absent. At that time she was up most of the day in a chair with no distress. Red cells,

4,600,000; Hb, 92%; haematocrit, 48. On April 27 arterial oxygen saturation was 88.5%.

Comment.—There seems little doubt that there has been considerable improvement in the cardiac condition, and the fall in blood count and haematocrit reading (both of which are now normal) and the rise in the oxygen saturation percentage are most encouraging. Unfortunately the persistence of the hemiplegia makes it impossible to assess the actual functional improvement. It is thought that the paralysis will soon improve enough to allow the patient to move about by herself.

Conclusion

These three cases are by no means precise, but the results are considered to be encouraging enough to be recorded in this preliminary communication. They serve, at any rate, to show that pulmonary valvulotomy is a feasible operation, and all three patients have survived the procedure. The result in the first may be classed as moderately successful so far; in the second the result is good; in the third it would appear to be good, but is as yet indeterminate functionally.

Two of the patients were of an age when the prognosis is ordinarily poor and when any operation carries a grave risk; both were very severely disabled. There can be no doubt that far better results are to be anticipated in younger patients, but it has not yet been possible to extend the operation to the younger age groups, although it is intended to do so.

The vascular complications in the first and third cases are disturbing and disappointing. The first case teaches us that a careful watch must be kept for signs of peripheral arterial embolism after cardiomyotomy so that immediate embolectomy can be considered. Arterial thrombosis, even to the extent of hemiplegia, occurs with the Blalock operation just as often. Presumably it is most likely to occur when the blood pressure falls to a low figure for some time, especially in the presence of polycythaemia.

Much still remains to be done in the development of this operation in the treatment of "blue children" suffering from pulmonary stenosis, but the results in these three cases are encouraging enough to justify proceeding further with it. It should be made clear that it is not suggested that this operation is meant to supplant the Blalock operation. As it stands at present it is of use in only a small proportion of cases; until a method can be found to relieve the subvalvular stenosis (if indeed this is possible), the Blalock operation must continue to take precedence. However, unless the Blalock operation is considered to have solved the problem of *morbus caeruleus* completely, it is essential to try to develop new and alternative measures.

I am indebted to Dr. Maurice Campbell, under whose care all these patients were, for most of the case histories, which are made from his own notes.

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The British Red Cross Society has published a neatly illustrated pamphlet (price 1s., by post 1s. 1½d.) by Air Marshal Sir Harold Whittingham, entitled *ABC of First Aid Treatment*. It is suitable as a short manual of reference or for refreshing the first-aiders' memory.

B.C.G. VACCINATION: IS IT OF ANY VALUE IN CONTROL OF TUBERCULOSIS?

BY

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Several eminent German physicians who are about to introduce B.C.G. vaccination in Germany, thanks to the Scandinavian Red Cross, have called my attention to the article on this subject contributed to the *British Medical Journal* by Dr. G. S. Wilson (1947). From various quarters, including England, I have been requested to answer his criticism of B.C.G. vaccination, which has given rise to a certain hesitation and concern, since Dr. Wilson, as a member of the Medical Research Council, is a critic whose opinion carries considerable weight. As I introduced the most widely practised and, by Wilson, criticized method of B.C.G. vaccination and am largely responsible for its present widespread use in Sweden, and as, furthermore, I am prepared to help in introducing it to combat tuberculosis in the devastated countries of Europe, I feel prompted to test the value of Wilson's criticism.

I have no reason to disbelieve Wilson when he says that he is not prejudiced against B.C.G. vaccination; my faith in his objectivity is, however, put to a severe test by the manner in which he argues against B.C.G. In several points the criticism is undoubtedly justified, although correct mostly on unessentials; in many respects it is erroneous and unjust, not giving the impression of being sufficiently unbiased; and in all respects it overshoots the mark.

Investigations on Children

Wilson's quotation of my investigation (Wallgren, 1934) is entirely correct; I pointed out in that paper that it was not feasible to establish how large a part B.C.G. immunity had played in the favourable results reported. Other factors were contributory. Although many children were isolated until 3 months of age before being subject to exposure, after this period they nevertheless were exposed to virulent infection without falling ill from primary tuberculosis, save in a single case. This is a result which surely the whole battery of prophylactic measures known to us cannot parallel when dealing with children of this age that have not been vaccinated before exposure to infection. Further experience has shown me that cases of clinical tuberculosis occurring after an adequately performed B.C.G. vaccination are extremely uncommon and remarkably benign. While primary pulmonary tuberculosis in a child unvaccinated with B.C.G. does not show any tendency towards radiographically demonstrable regression during the first six months, in a child who has been vaccinated with B.C.G. and who was still tuberculin-positive at the time of infection radiographs reveal that within this period the condition has virtually resolved. It is also remarkable that I have not seen a single adequately vaccinated child die from tuberculosis after having been infected, nor have I observed a single case of tuberculous meningitis among vaccinated children. Is, then, the scientifically calculated and statistically reported degree of immunity of such great import if the effect of the vaccination is satisfactory?

Rosenthal and his co-workers (1945) have used a different method of vaccination (multiple punctures) from that with which I have had experience (intradermal). Wilson is admittedly right in stating that the results of their experi-