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# RESULTS OF THE BLALOCK-TAUSSIG OPERATION IN 200 CASES OF MORBUS CAERULEUS

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In 1949 we reported the results in the first 50 patients with morbus caeruleus who had a Blalock–Taussig operation at Guy's Hospital (Baker *et al.*, 1949). We now report the results in those operated on since and a longer follow-up of the earlier patients.

We have included all our 200 patients who have had an anastomosis performed or attempted up to the end of 1951. Most of these (129) have been operated on by Mr. R. C. Brock, but 57 were operated on by Mr. Holmes Sellors and 14 by Mr. Ian Hill. Most of the patients had Fallot's tetralogy, but we use the wider term morbus caeruleus because of uncertainties of diagnosis: even a successful operation proves only that the blood flow to the lungs was inadequate and has been improved. First, the 200 patients are considered together and later in different groups such as tricuspid atresia, pulmonary atresia, and dextrocardia, to assess the operative risk and the improvement to be expected.

*Age and Sex.*—Most of the patients were children, and the age distribution is shown in Table I. Nearly half were between 5 and 9 years of age, over half were

TABLE I.—*Age Distribution of Patients and Operative Deaths*

| | Age in Years | | | | | Total |
|---|---|---|---|---|---|---|
| | 0–4 | 5–9 | 10–14 | 15–19 | 20 and Over | |
| Total No. of cases | 18 | 90 | 59 | 21 | 12 | 200 |
| Deaths with operation .. .. | 3 | 10 | 4 | 1 | 6 | 24 |
| Deaths at each age | 17% | 11% | 8% | 5% | 50% | 12% |
| Very good results among survivors | 40% | 53% | 61% | 80% | 33% | 56% |

less than 10, and over four-fifths less than 15 years: the youngest was 3 and the oldest 35 years. Boys slightly predominated, there being 114 to 86 girls.

*Disability.*—This was graded (Campbell, 1948) as IV in 75, III in 77, II in 44, and I in only 4 of the patients before operation. These last four were mostly older patients, about 20, who were not very disabled but were most anxious to be able to lead a more active life.

*Symptoms and Signs.*—We are not discussing these further, as we have little to add to our earlier remarks (Baker *et al.*, 1949).

## The Role of Special Investigations

All the earlier and many of the later patients have been chosen for operation without any special investigations (angiocardiography, cardiac catheterization, etc.). Where the diagnosis seems straightforward and there is no doubt about the presence of adequate pulmonary arteries this seems a reasonable policy. These investigations all involve at least some discomfort to the patient and sometimes a definite risk.

*Circulation times* were done on most patients at one time, but have now been abandoned, in a centre where cardiac catheterization and angiocardiography are available (Campbell, 1951).

*Angiocardiography* has been used most often—in just over one-third of the patients in this series; it has proved of great value in difficult cases, but has rarely contributed much useful information in the straightforward ones. Briefly, its chief value in selection for anastomotic operations is in confirming the presence of an overriding aorta and in elucidating the anatomy of the systemic vessels and the presence of pulmonary arteries when this is doubtful. It is less effective than might be expected in deciding *in a doubtful case* whether the blood flow to the lungs is diminished or not. The change in the density of the lung fields and the speed with which the diodone passes through the lungs to the left side of the heart are better indications than the time at which the pulmonary trunk first fills (Campbell and Hills, 1950). Like the circulation times, it has no place in post-operative assessment.

*Arterial oxygen saturations* have rarely been estimated except as a part of cardiac catheterization. Alone, they yield little information apart from confirming, perhaps more precisely than the circulation times, the presence of a right-to-left shunt—although they do not, as the circulation times may, distinguish whether the cyanosis is due to a shunt or to some change in the lungs preventing the normal oxygenation of the venous blood. For this distinction a second arterial sample, taken while the patient breathes oxygen, is necessary. If the patient is frankly cyanotic the arterial oxygen saturation is superfluous. Its use as a means of assessing the severity of the shunt or the degree of improvement we have not felt to be justified; its apparent objectivity gives it a spurious appearance of reliability, for the lability of the cyanosis in these patients is well known. Significant increases in the arterial oxygen saturation are nearly always appreciable clinically as lessened cyanosis.

*Cardiac catheterization* has been performed on many patients with morbus caeruleus, but not all these have had anastomotic operations. About 30 in this series were so investigated, but as the results are in no way

peculiar to them we have reviewed all who were studied by this technique. We have not used catheterization to study the results of the anastomotic operations. The only change that could be detected is the shunt of arterial blood into one of the pulmonary arteries; by the nature of things it is not possible to assess its volume by catheterization and there are simpler criteria by which its mere presence or absence can be judged.

*Fallot's Tetralogy.*—In 57 catheterizations on patients with this diagnosis there was only one failure to enter the right ventricle. The pulmonary trunk was intubated in 45 and the aorta as well in 9 of these; *in no instance was the aorta entered when the pulmonary trunk was not.* A defect of the atrial septum was demonstrated by the passage of the catheter in three patients. The systolic pressure in the right ventricle was within the normal range for the systemic arterial pressure. Whenever they were determined at the same time they showed no significant difference. This contrasts with pure pulmonary stenosis, in which the right ventricular systolic pressure often exceeds the systemic. The pulmonary arterial pressure was generally between 17/6 and 10/3 mm. Hg, and nearly always below the normal value of about 23/8 mm. Hg, measured from the mid-axillary line.

In the patients accepted as having Fallot's tetralogy a right-to-left shunt was always present but varied greatly in size; the arterial oxygen saturation fell within the range 47–92%, but in over half was between 70 and 85%. In keeping with the observations of Deuchar and Knebel (1952), the systemic blood flow at rest has always fallen within the normal range, whereas the pulmonary blood flow has been reduced by an amount dependent on the size of the shunt.

Among the straightforward examples of Fallot's tetralogy there was evidence of a left-to-right shunt into the pulmonary artery in one only, and at operation the surgeon found a small persistent ductus arteriosus. A few patients with a provisional diagnosis of Fallot's tetralogy have been found to have left-to-right shunts into either the right atrium or the right ventricle. The former has been observed only where anomalous pulmonary veins were present and the latter where post-mortem control or further study (angiocardiographic, etc.) nearly always proved the presence of some more complex lesion than Fallot's tetralogy—for example, a single ventricle or pulmonary atresia. The finding of a shunt into the right ventricle is therefore a strong indication for reconsidering a diagnosis of simple Fallot's tetralogy.

*Pulmonary Atresia.*—We have studied 14 patients believed to be examples of this condition, but were able to intubate the right ventricle in 10 only. Naturally the pulmonary trunk was not entered in any, but the aorta was entered in six: its seems that entry of the aorta with failure to enter the pulmonary trunk may help in distinguishing pulmonary atresia from Fallot's tetralogy. A defect of the atrial septum was demonstrated in 8 of the 14, including those in whom the right ventricle was not entered. The arterial oxygen saturation always fell within the range 60–84%. Basing blood-flow calculations on the assumption that the blood to the lungs has the same oxygen saturation as the systemic arterial, the pulmonary blood flow was less than, equal to, or more than the systemic in about equal numbers of cases, although radioscopy gave a different impression in the last group: again it was the pulmonary blood flow that was changed when it differed from the systemic.

*Tricuspid Atresia.*—Here cardiac catheterization has little to offer, and the findings, being essentially negative, have little significance. It can contribute useful information only when by intubating the right ventricle it disproves the suspected diagnosis of tricuspid atresia. In six cases studied the atrial septal defect was demonstrated in three; the arterial oxygen saturation fell within the range 56–79%, and in all except one the pulmonary blood flow was less than the systemic.

## Nature of Operation

Most of these patients had end-to-side subclavian-pulmonary anastomosis, as this was regarded as the operation of choice. At first, following the original practice of Blalock and Taussig (1945), the operation was performed on the side opposite to the aortic arch; but after the first 50 Mr. Brock decided on surgical grounds that the left side was the best in all cases (Baker *et al.*, 1949). Provided the anastomosis is a good one, there is no evidence that the side on which it is done makes any difference to the result. Departures from this operative technique were dictated by the anatomical findings at operation.

In about one-twelfth of the cases it was found necessary to divide a pulmonary artery and perform an end-to-end anastomosis. This was mainly in patients with pulmonary atresia in whom the pulmonary artery, if present, was often small and thin-walled. The less good results may therefore be due as much to the original defect as to the end-to-end anastomosis. There is no doubt that it can sometimes produce a very good result, and, strangely enough, the anastomotic murmur resulting from it is indistinguishable from the other.

Pott's modification of the Blalock–Taussig operation was carried out in three patients only. In two the result was very good, but one shows some evidence of the shunt being too large. The third, despite the surgeon's fear that the anastomosis was too large, had no anastomotic murmur and no benefit from the operation, which suggests that the stoma failed to function (Case 1).*

*Innominate and Other Arteries.*—The innominate artery was used once with an ideal result. The right common carotid artery was also used once; the stoma formed was thought to be rather large, and the patient died of pulmonary oedema. Another patient, in whom the carotid had to be divided to mobilize the subclavian sufficiently, died, apparently from the interference with the cerebral blood supply. After this experience Mr. Brock decided that the use of the carotid or innominate arteries was a gamble that should not be taken (Baker *et al.*, 1949). The internal mammary artery was successfully used for an anastomosis in two cases: one yielded a very good result, though the reduction in cyanosis and haemoglobin was only slight, but the other showed little improvement and had no anastomotic murmur. An attempt to use a large intercostal artery on one occasion failed, and the patient recovered from the operation unchanged.

## Method of Follow-up

All these patients were seen by one of us before operation and nearly all by both of us afterwards, generally several times. Often the category in which they should be placed was decided in discussion with others who were working in the clinic. We have tried to see the patients annually near to the anniversary of the operation, but could not always adhere to this strictly. We have therefore grouped the visits between 10 and 21 months as one year, between 22 and 31 months as 2 years, between 32 and 43 months as 3 years, and 44 months or over as 4 years.

Nine patients have gone abroad since their operations, but three had already been followed for 18 to 24 months. We have received recent reports from all except two who were doing well when they left the country 19 and 24 months after operation. Of those in this country none has been lost sight of, but we have accepted reports of several who

*Some cases have been given numbers to save repetition when they are referred to again.

live some way from London, especially when we had seen them previously and have been told that their condition is as good as before.

The period of follow-up of the 165 who left hospital after a completed anastomosis was as follows (the patients who died later have been counted as being followed until the date of death): 4 years, 28 patients; 3 years, 37; 2 years, 42; 1 year, 53; less than 1 year, 5. Some of the patients have now been seen at the five-year follow-up, and none of them has lost ground.

At each visit we have assessed the patient's capacity and progress and have recorded the degree of cyanosis and clubbing, the site of the anastomotic murmur, and the blood pressure. Getting the patients to jump up and down for a short time has proved a simple and useful way of helping to assess the disability and the cyanosis after exercise. Haemoglobin estimations and teleradiographs of the chest have been done as a routine. Electrocardiograms were done often at first, but as it became apparent that there was little if any change, even when the heart became larger, we have been less particular about repeating them. The role of the special investigations has already been discussed. In a few cases all the data outlined have not been obtained or we have had to rely on reports forwarded by their doctors.

*Assessment of the Degree of Improvement.*—This is not always easy. Some patients before operation could walk only a few yards, whereas others could walk a mile or more, so that the improvement cannot be judged simply by the amount they can do after. At first, with the vivid memory of the severe disability, the result seems miraculous, and for the parents often remains so; later, as the memory of this fades, there is more tendency to compare them with a normal child, and in this way it may seem less dramatic, but the examples given will indicate how great the change is. Even moderate improvement may mean a lot to the patient, but our good results mean much more than this, as such patients can often earn a living and lead a life that is nearly normal instead of being almost completely disabled and a drag on their families. The classification of the results is shown in Table II. We have taken particular care not to overestimate the improvement, placing the patient in the lower rather than the higher category whenever there was any doubt.

TABLE II.—*Results of the Operation*

| Group | Results of Operation | No. of Patients |
|---|---|---|
| I | Very good | 99 |
| II | Good | 37 |
| III | Some improvement | 17 |
| IV | Good or improved at first, but died later | 6 |
| V | No improvement | 6* |
| VI | No anastomosis possible | 11 |
| VII | Death at or shortly after operation | 24 |

*Two other patients who were originally in this group showed some improvement after a second operation.

## Results of Operation

### Group I. Patients with Very Good Operative Result (99)

We have described a result as very good when the patient can earn his living or attend an ordinary school and can walk three to four miles. He is living an almost normal life except for the more vigorous activities that are not essential. If he had been so disabled that he could not walk 100 yards we have classified the result as very good even if he has not reached this standard but was able to walk at least a mile. The following examples will make the position clearer.

A boy of 3 who could walk only 10 yards was soon running about almost like a normal child. A boy of 7 who could walk 400 yards on a very favourable day was soon at school leading an almost normal life except for football: three years later he persuaded a friend to run away from school with him, and was found by the police 12 miles away. A young man of 18 who had found it hard to walk across the out-patient hall was able to walk 6 miles and to work at light engineering. A boy aged 12 who could walk only 20 yards had walked 9 miles two years later; and another who could walk only a few hundred yards at a normal pace or half a mile slowly has walked 12 miles. One girl of 12 was carried into the room on her first visit and could walk no more than 20 yards in good weather: after operation she was enormously improved, and the day she was seen had walked round the zoo for three hours with one rest. Another girl, aged 15, had been getting so much worse that even dressing provoked attacks in which she nearly lost consciousness: now, two years after operation, she is working regularly as a machinist and can walk four miles and do everything she wants except dancing; her cyanosis has not improved as much as most (see later) and her haemoglobin has fallen from 135 only to 119% (Case 2).

There were other patients who were much less disabled before operation. For example, one girl of 13 could walk half a mile fairly easily and was not very cyanosed: after operation she was walking five miles, cycling in a hilly district, and doing anything she wanted.

The percentage of very good results increases from the youngest patients to those between 15 and 19, in whom it is 80% of the survivors, but then falls off again in those over 20 (see Table I).

### Group II. Patients with Good Results (35)

We have been conservative in including cases in the highest class. When a patient leads a normal quiet life but falls short of this high standard in some particular we have recorded the result as good. For example, a girl of 6 was getting worse before operation and walked only the length of the room; she rarely went outside, and was having frequent cyanotic attacks, in which she was unable to stand. A year after operation she is a different child, almost a normal colour and without attacks, but was not put in the highest class because she was not yet going to an ordinary school or walking more than half a mile.

A boy aged 10, with tricuspid atresia, found it difficult to walk a mile and was still moderately cyanosed, with a haemoglobin of 130%. But his mother called the result a miracle, as he was able to attend an ordinary school and go and return alone, whereas before he had walked only 100 yards (Case 3).

Some cases which would otherwise have been classed as very good have developed another disabling lesion, and therefore have been classed as improved or good. This applies to two patients who have developed tuberculosis of the lungs and to others with neurological lesions from which they have not recovered completely.

### Group III. Patients Somewhat Improved (17)

This relatively small group contains all those who were still seriously limited in activity but showed real improvement, which sometimes meant a lot to them. Thus one girl who could walk only 20 yards can now get about the house freely and walk 200 yards outside—a tenfold increase—but we regard this as disappointing, though the change it has made to her inside the house seems to her parents enough to justify the operation.

### Group IV. Patients who were Improved but Died Later (6)

All these patients were improved at first, the result being very good in two, good in two, and improved in two, but they died 8 to 37 months after operation. The first two were especially disappointing cases.

The first, a boy aged 3, was very disabled, and the large heart—cardiothoracic ratio (c.t.r.) 62%—made us anxious about the chance of success. He was almost normal 15 months after operation and the heart was no larger. Eighteen months later, in Kenya, he developed weakness of one side and was sent back to England: the condition of his heart was still good, but a diagnosis of cerebral abscess was made, and, though operation for this seemed successful, he died suddenly six hours later (Case 4).

The second, a girl aged 7, was able to walk only 50 yards on a good day and had frequent attacks in which she nearly lost consciousness: afterwards she could walk a mile to school and run 20 yards. Three years after operation she was leading an ordinary life except for no games at school; her heart had not

enlarged unduly (c.t.r. 47 to 53% quickly and 54% three years later). A month later, on holiday, she seemed as well as usual, but woke up one night with symptoms of acute pulmonary oedema and died before a doctor could see her (Case 5).

The third, a young woman aged 20, was classed as a good result but died suddenly in the street 17 months later; the inquest did not reveal the cause of death.

The fourth, a boy aged 4 with tricuspid atresia, was classed as a good result, and there would have been no anxiety had he not been seen after five months and found to have an unusual increase in the size of his heart (c.t.r. 61 to 69%). He died three months later with congestive failure: it is true that it followed bronchitis, but this would probably not have proved fatal had the heart been smaller (Case 6).

The fifth, a boy aged 4, was thought to have Fallot's tetralogy with complications, as the heart was large (c.t.r. 59%). Operation was decided on as he was losing ground and seemed unlikely to live long: he was better for a year, but then lost ground and died in his second paroxysm of ventricular tachycardia 17 months after operation. At necropsy pulmonary stenosis with a single ventricle was seen (Case 7).

The sixth, a man aged 21, was classed as improved only, as he had not been very disabled. A pulmonary abscess had troubled him before his operation, but was thought to be quiescent and healed. A year later, however, he was admitted with recurrent haemoptysis; aspergilli were cultured from the sputum, and in spite of treatment he died a few weeks later. Fallot's tetralogy had been diagnosed, but necropsy showed a pulmonary valvular stenosis with only a small patent foramen ovale and an unusual, small, low ventricular septal defect (Case 8).

These deaths give a rough indication of the mortality that may be expected after an operation that has been successful. Of 159 patients, 6 (3.8%) died during the period of follow-up, but two at least from causes quite unconnected with their original operations—a cerebral and a pulmonary abscess. As the average period of observation was 2.5 years the annual mortality rate from all causes was 1.5%.

### Group V. Patients Without Improvement After Operation (6)

There were six patients who were little if at all improved by their operation, and three of them have died since. One was submitted to operation after much doubt, as the diagnosis was uncertain. Only an end-to-end anastomosis was possible, and her colour and capacity were somewhat improved, but she developed signs of congestive failure, which persisted in varying degree over the next two years (Case 9). In a second the surgeon was doubtful if he had been able to achieve a functional anastomosis, and the absence of any benefit and of continuous murmur after the operation supported this. A third had a Pott's operation (Case 1). The fourth seemed better at first but became worse again a few days later, probably from a thrombosis at the site of the anastomosis. She died at home a year later from "meningitis." The fifth, with tricuspid atresia, was better in colour and was able to do more, but this was only at the expense of heart failure, and his liver remained enlarged and pulsating. In spite of a period of treatment in hospital his congestive failure progressed, and he died after ten months (Case 10). The sixth, a girl aged 8, should probably not have been operated on, as she had never walked and had chronic nephritis, but her outlook without seemed hopeless. Her dyspnoea and cyanosis were improved after operation, but a month after leaving hospital she developed gross oedema and died suddenly a few weeks later with an exacerbation of her nephritis. At necropsy severe pulmonary stenosis with a single ventricle was found (Case 11).

There were two others who were at first in this group. However, second operations one or two years later have been followed by some improvement, so they have been included in that group.

Four patients, in all, have had a second operation, and none of them has achieved a good result. This may be bad luck, and often the surgeon has thought that he had not been able to make a good anastomosis, but it hardly encourages one to advise a second attempt when the first has not succeeded.

### Group VI. Patients in whom Anastomosis was not Possible (11)

In 11 of the 176 patients who survived the operation no anastomosis was possible. They recovered from the operation unchanged, though one or two later claimed that they were somewhat improved. Two of them had transposition of the main vessels. In retrospect, this mistake should have been easy to avoid: the lung fields may look very dense from a heavy collateral circulation, but the markings are finer and more concentrated round the hilum or in the upper right mediastinum. It is, however, important not to make the opposite mistake of thinking that a collateral circulation indicates an increased blood flow to the lungs, and this has been one of the main medical reasons for angiocardiography.

Taussig (1948), in setting down the criteria for a successful anastomosis, included the presence of a suitable pulmonary artery; lack of this has been the most common cause of failure to complete an anastomosis. In four patients who are discussed in the section on pulmonary atresia no pulmonary artery could be found. In a fifth it was thought wiser to stop the search for the pulmonary artery because of the mass of collateral vessels: in retrospect he was thought to have pulmonary atresia though pulmonary arteries could be seen on the angiocardiogram. In a sixth the pulmonary arteries were too small: the left was so minute that no anastomosis could be made, and at a second operation the right, although apparently adequate on angiocardiography, soon divided into three and proved too small even for an end-to-end anastomosis. In the last three, technical difficulties prevented an anastomosis being made, and a further operation has been advised.

### Group VII. Patients who Died at or Shortly After Operation (24)

Four of the 24 patients who died were in the groups that are discussed separately. Two (Cases 12 and 13) had a single ventricle and pulmonary stenosis, one with isolated dextrocardia and one with isolated laevocardia (Campbell, Reynolds, and Trounce, 1953). The third (Case 14) had isolated laevocardia, a bilocular heart, and pulmonary atresia, and the lungs were supplied through a persistent ductus (Campbell, Gardner, and Reynolds, 1952); the fourth also had pulmonary atresia with a persistent ductus. A fifth, a woman aged 24, had complete transposition of the great vessels and pulmonary stenosis.

Of the other 19, there were 5 (in addition to the two just referred to) who had pulmonary atresia. Surgical treatment is more dangerous in this subdivision of Fallot's tetralogy. The patient's life is dependent on the collateral arteries that have developed, and the division of some of these produces an extra risk of haemorrhage during operation and of lessening a pulmonary blood supply that is already scanty: in addition, the small and often thin-walled pulmonary artery is poor material for an anastomosis. Even after these difficulties were appreciated and special efforts made to avoid them, three patients with pulmonary atresia have died.

The remaining 14 patients had Fallot's tetralogy: 5 were 19 or older, this being nearly half the patients of that age who were operated on. Our figures show that the operation is more dangerous in the older patients (Table I), though the good results justify some extra risk. Those in whom operation has been advised were often losing ground, for the others are usually reasonably satisfied with the quiet life they can lead.

Even among the nine younger patients (all under 12 years) there was sometimes a special feature to account for the fatal issue: one had a single pulmonary artery and died when it was clamped for the anastomosis; a second had an aorta that arose entirely from the right ventricle; and a third had a large heart and almost a single atrium. A satisfactory feature of this analysis, therefore, is that among those who died there were only six young patients with uncomplicated Fallot's tetralogy.

Patients who leave the operating theatre in fair condition can be expected to do well, for nearly all those who died were already causing anxiety by the end of the operation. Of the 24 patients, 6 died during the operation, generally from cardiac arrest; three of these had Fallot's tetralogy and three had more complicated lesions. Another 13 died within the first 24 hours—two early cases from collapse of the lung after a prolonged operation, one from pulmonary oedema, and most of the others from intrathoracic haemorrhage (especially in pulmonary atresia) or from the effects of cerebral anoxia, usually the result of the low blood pressure and defective cerebral circulation during operation.

Only 4 of the 24 who died left the operating theatre in reasonably good condition with a completed anastomosis. These four died on the second or third day after operation —one from intrathoracic haemorrhage, the second suddenly and without any obvious reason, and the third and fourth after developing thrombosis at the site of the anastomosis. Thus, with one exception, all the patients who have died have done so within 72 hours of operation. This one, soon after a difficult operation, had such a severe haemorrhage of about 6 pints (3.4 litres) that she developed cerebral symptoms: after her death on the eighteenth day widespread pontine haemorrhages and thrombosis of the basilar artery were found. In 5 of the 24 patients who died it had not been possible to complete an anastomosis.

## Objective Effects of the Operation

Most of the patients who were improved by the operation show a number of objective changes, which are briefly as follows: Less cyanosis at rest and on exercise, and less clubbing of the fingers and toes; some widening of the systemic pulse pressure; the development of a continuous murmur, similar to that of a persistent ductus arteriosus, arising from the anastomosis; a reduction in the polycythaemia; and an increase in the size of the heart as measured by the cardiothoracic ratio.

All these changes are not found in every case, and any one or more may be absent, except the anastomotic murmur, which is rarely so. They vary in degree in different patients, but, generally speaking, the greater the subjective improvement the greater these changes: it is reasonable to deduce that the greater the changes the larger the shunt through the anastomosis. While this is the picture gained by reviewing the group as a whole, individual patients show many exceptions to it. Each of the changes is discussed separately.

### Cyanosis

In many of these patients the cyanosis changes greatly from time to time, and the degree present at any moment is determined by a large number of variables—for example, temperature, emotion, and recent exercise. In addition its assessment depends on the subjective judgment of the observer, so that although we have graded the cyanosis at each visit we have not thought it worth while to attempt any numerical analysis. Our general impression, confirmed by study of the data, is that the lessening of the cyanosis and of the disability are well correlated. When the initial cyanosis is only slight at rest, there is less room for improvement, and so the change is less striking, except perhaps after exercise.

Most patients still show cyanosis even at rest, although it may pass unrecognized by the relatives. About a quarter of the very good results and an eighth of the good results showed no cyanosis at rest, but only two patients showed no cyanosis after exercise. One, a girl of 6 with Fallot's tetralogy, had grade IV cyanosis and severe disability. At three and at four years after a successful operation she could do almost anything, and showed no cyanosis even after exercise. Her haemoglobin had fallen from 150 to 70%; the blood pressure had widened from 105/75 to 120/60; and the cardiothoracic ratio had increased from 52% to 65% at three years and to 67% at four years. There was a very loud anastomotic murmur, and there is little doubt that this patient has a shunt that is larger than the optimum (Case 15).

There are also a few exceptions to the general rule at the other end of the scale—that is, a good result but yet with deep cyanosis. A boy aged 9 is a good example. Before operation he was severely disabled and cyanosed. Two years after operation he was regularly at an ordinary school and could walk two miles; but his cyanosis was still moderate, and his haemoglobin had fallen only to 125 from 141% (Case 16; see also Case 2).

### Clubbing of the Fingers

Before operation the degree of clubbing usually corresponds with the degree of cyanosis. It seems to reflect the anoxaemia present during activity, as when it is more than would be expected for the cyanosis at rest the latter often increases more than usual on exercise. Baker *et al.* (1949) found that after operation the clubbing improved for about six months and then reached a stable level; longer follow-up has disclosed no reason to alter this impression.

The regression of the clubbing has generally been striking, but it is less consistent than the reduction of cyanosis. Only 10 patients had no clubbing after operation; all but one of these showed very good results, and none had more than moderate clubbing originally. In no case has severe clubbing been wholly abolished by a successful operation. It has always looked symmetrical in the two hands before operation, but afterwards the clubbing on the side of the division of the subclavian artery has sometimes been less than on the other side. The difference has been slight except for one patient, in whom it was specially noted at both visits, one and two years after a successful operation. These observations seem to be in keeping with the findings of Lovell (1950) and of Wilson (1952), who was able to demonstrate an increased finger blood flow in patients having clubbing from a variety of causes, and also changes in the clubbing in individual patients associated with corresponding changes in the blood flow.

### Changes in Pulse Pressure

After subclavian-pulmonary anastomosis the peripheral pulse in the affected arm inevitably disappears. In every case the collateral circulation has developed well and there has been no trouble in the arm, but even four years after operation the radial pulse has rarely been detected and the blood pressure has never been measured by the sphygmomanometer.

Significant changes in pulse pressure due to the anastomosis must be caused by the blood flowing from the systemic to the pulmonary circuit through this. Generally the change is not great: this is hardly surprising, as even with a persistent ductus the pulse pressure is often not very wide and after subclavian-pulmonary anastomosis the flow is not usually expected to be very large. Satisfactory readings were available for comparison in only 103 of the 158 patients showing improvement (Table III), largely because the diastolic pressure was often difficult to determine before.

The blood pressure in these patients is probably not very different from that of normal children of the same age: the systolic pressures are mostly below 115 and often about

TABLE III.—*Increase of Pulse Pressure after Subclavian-pulmonary Anastomosis*

| Result of operation | No Significant Change | Increase Between 15 and 30 mm. | Increase of 30 mm. or More | Total |
|---|---|---|---|---|
| Very good .. .. | 53 | 11 | 9 | 73 |
| Good .. .. .. | 14 | 4 | 0 | 18 |
| Somewhat improved .. | 11 | 0 | 1* | 12 |
| Total .. | 78 | 15 | 10 | 103 |

* The objective changes—the improved co'our and capacity when in hospital—show that the operation had succeeded, but ten years in bed prevented her gaining corresponding improvement.

90–100, and the pulse pressures are rather on the small side. Comparison of the pressure before and after operation is complicated by its variability and by the natural increase that occurs with growth; the former has been allowed for so far as possible by comparing readings taken at different times. From Fleisch's (1927) figures it seems that the pulse pressure increases by about 2–4 mm. Hg a year during most of childhood: in the younger patients, therefore, in four years it may rise about 12 mm. Hg naturally, so that an increase of less than 15 mm. Hg may be without significance. In 18 of the 103 cases there was no change or a small fall in the pulse pressure, and in 60 there was an increase of less than 15 mm.; these changes were not thought to be significant. There were only 25, or about one-quarter of those benefiting from the operation, who showed a larger increase. With one exception (see footnote to Table III) all these showed great improvement. Nine of the 73 showing very good results had an increase of 30 mm. Hg or more, while in the four showing good results with the greatest increase, it did not exceed 25 mm.

There is some correlation between the larger pulse pressures and the larger hearts, but it is not very close. These larger increases must almost certainly be associated with large shunts, and all these patients show at least a moderate increase in the heart size, as well as reduced polycythaemia and lessened cyanosis. Case 15, quoted above, is a good example of this group.

### The Anastomotic Murmur

After a successful systemic-pulmonary anastomosis, whether it is end-to-side or end-to-end, a murmur similar to that of a persistent ductus almost invariably appears. If it is not heard it may be that thrombosis or kinking has obstructed the opening.

The main characters of the murmur are fairly constant. It is essentially continuous throughout systole and diastole, with systolic accentuation. Its systolic element is nearly always distinguishable from the systolic murmur usually present beforehand by its lower pitch and its site of maximum loudness. The diastolic element is, however, the essential identifying feature. It is nearly always best heard in the first to third spaces below the medial half of the clavicle on the side of the operation, and is usually widely conducted so as to be audible through to the back.

The loudness of the murmur varies greatly, but it is easily heard in nearly three-quarters. Even when loud it is rarely accompanied by a thrill—a feature noted in only three patients, all with very good results.

Table IV shows that in most cases the murmur correlates with the result. The one case in which a very good result was obtained but no anastomotic murmur was heard seemed greatly improved at first and was thought to have a continuous murmur, but some days after operation the patient lost ground, the murmur could not be heard, and a thrombus at the site of the anastomosis was suspected. He was recorded as only somewhat improved, but he got better over the next year and eventually achieved a very good result. Another unusual feature is the development of a most striking collateral circulation over the chest, such as one sees in coarctation of the aorta; but it seems impossible that the thrombus should have obstructed his aorta when he has made such steady progress without any major illness, his femoral pulses can be felt normally, and there is no rise of blood pressure in the arms. A systolic murmur that is now heard may be part of the anastomotic murmur or may be due to the collateral circulation.

TABLE IV.—*Relation of Anastomotic Murmur to Result of Operation*

| Anastomotic Murmur | No. of Patients Classified by Result of Operation | | | |
|---|---|---|---|---|
| | Very Good | Good | Some Improvement | No Improvement |
| Loud or good (102) .. | 78 | 20 | 4 | 0 |
| Moderate or faint (28) | 15 | 8 | 4 | 1* |
| None heard (10) .. | 1† | 0 | 7 | 3 |

Patients who died during the follow-up period have been classified according to the result judged before death. 27 cases have not been included in this table either because details about the murmur were not known (22 cases) or because they were obscured by a continuous murmur that had been present before the operation (5 cases).
* Case 9, q.v.
† See text.

The seven cases that showed some improvement though no murmur was heard may have had small shunts through the anastomosis, or possibly may show the slight improvement that sometimes follows a thoracotomy even when nothing else is done.

### Changes in the Blood Picture

A detailed study of the blood picture in morbus caeruleus before and after operation, including many of the cases reported here, has been published by Brinton (1951). From the experience of this longer series we find no need to amend his conclusions, and therefore do not propose to deal with this matter in detail. He found that in spite of some discrepancies there was, as a rule, good correlation between the clinical assessment after operation and the changes in the blood picture. In the two patients in whom the normal blood picture led to a more favourable view than the clinical assessment, the former has proved right and both have now achieved good results. In the 12 patients in whom the improvement was rated less highly on the blood picture than on clinical assessment the latter has generally been confirmed over the next year or two, and their improvement is certainly due to a successful anastomosis. In about half of these the haemoglobin percentage has fallen to somewhere near normal, but in the others it has still remained high (see Cases 2 and 16).

The haemoglobin has fallen to normal or near to normal in most of our patients who have had a successful operation. The mean fall in those with very good results is from 130 to 102% (Table V), but the range both before and after operation is very wide, and is shown in Fig. 1.

TABLE V.—*Reduction of Haemoglobin After Operation*

| Result of operation: | Mean Haemog'obin Percentages | | | | |
|---|---|---|---|---|---|
| | Very Good | Good | Some Improvement | No Improvement | Death |
| No. of cases .. | 91 | 28 | 18 | 4 | 24 |
| Before operation | 130 | 129 | 137 | 119 | 148 |
| After ,, | 102 | 102 | 122 | 120 | — |

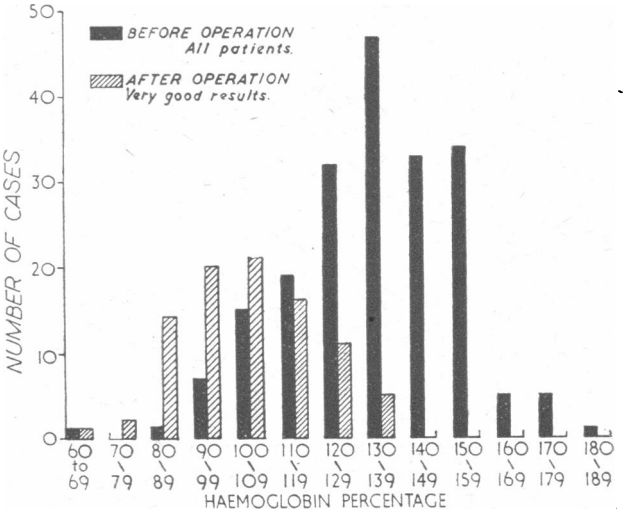


FIG. 1.—Frequency distribution of the haemoglobin percentages in all patients before operation and in those with very good results after operation. Before operation the majority were between 120 and 160%. After operation most were between 90 and 120% (100% = 15.6 g.%).

Before operation the majority were between 120 and 160%: after operation most were between 90 and 120%. Only those with very good results are included in Fig. 1, but the mean change is almost exactly the same in those with good results. It is much less in those who showed some improvement—the mean falling from 137 to 122%—and, as might be expected, does not show any change in the smaller number having no improvement. The high mean value in those who died after operation—148%; range 113–186%—reflects the greater average severity of this group.

Among those patients in whom an anastomosis was completed there were 31 without any significant reduction in haemoglobin. This includes 75% of the small number who were not improved, 39% of those who were somewhat improved, and only 18% of the greatly improved. Why some of the good results should fail to show a fall in haemoglobin is uncertain, but as the overriding aorta remains it is remarkable that so many achieve enough reduction in the anoxaemia to bring the haemoglobin down to normal. The haemoglobin remains high in some in whom the anastomosis is not adequate and probably in a larger number in whom the aorta is so dextroposed that there is still a large amount of mixing. Values less than 100% found in a few patients suggest that they were relatively anaemic—that is, had much less haemoglobin than might be expected for their degree of anoxaemia: correction of this after operation may account for some of those who do not show a fall of haemoglobin in keeping with the good result obtained.

## Size of Heart

The reduced anoxia that follows from the increased blood flow to the lungs benefits the heart directly, but this is likely to be offset by the extra mechanical work that an effective anastomosis imposes. No reduction of heart size is therefore to be expected from the operation, and, indeed, an increase would be more likely, and was soon recorded (Blalock and Taussig, 1945).

It is almost a principle of cardiology that, other things being equal, the larger the heart the worse the prognosis. Naturally, therefore, the enlargement following these operations has been viewed with apprehension lest it should be progressive and foreshadow cardiac failure. It has, indeed, been used as the main argument against the operation. We have therefore paid particular attention to the heart size, assessing it for simplicity in terms of the cardiothoracic ratio—the maximum transverse diameter of the heart in the teleradiograph as a percentage of the transverse diameter of the inside of the thoracic cage, just above the diaphragm.

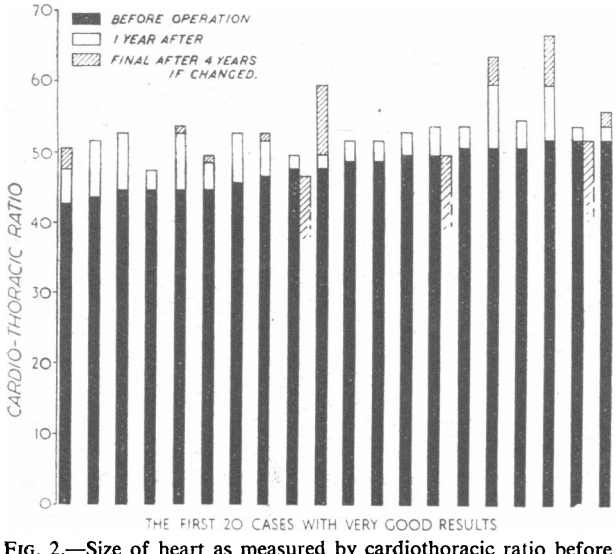


FIG. 2.—Size of heart as measured by cardiothoracic ratio before and after operation. The first 20 patients with very good results have been chosen for illustration and have been arranged according to the initial heart size. The unshaded area above each black column shows the increase in size within the first year. The lightly shaded area above this shows the increase during the subsequent three years, but, as will be seen, there was often no further increase; sometimes there was a decrease, shown by the shaded column on the right. Only three of these patients showed a significant increase after the first year.

The changes in the heart size in the first 20 cases with very good results are shown in Fig. 2. The mean values in the various groups are given in Table VI, where they have been grouped according to the results, and in Table VII, where they have been grouped according to the time that has elapsed since operation. The mean cardiothoracic ratio before operation is essentially the same in all groups, except that it is a little less in those who have shown very good results and much more in those who gained no benefit, probably because several of these had some more complex defect than Fallot's tetralogy, in which the heart is usually little, if at all, enlarged. After operation the heart size increases rapidly in the first month or so (figures not included in the table), and this increase on the *average* persists unchanged, at least for the four-year period of our observation. The average increase in the cardiothoracic ratio was 3 points—that is, from 49% to 52% or from 51% to 54%. This picture, gained from the means of the whole series, obscures some of the individual variations. The majority do behave in the manner described, but there are three smaller groups that behave differently: (1) those in whom the heart size shows no change; (2) those in whom it increases at first but subsequently gets less; and (3) those in whom it increases progressively.

TABLE VI.—*Cardiothoracic Ratio and Result of Operation*

| | Cardiothoracic Ratios: Mean and Ranges | | | | | | |
|---|---|---|---|---|---|---|---|
| | Very Good Results | Good Results | Some Improvement | No Improvement | No Anastomosis | Operative Deaths | All Cases |
| Before operation | % 48·8 | % 51·0 | % 50·1 | % 57·8 | % 52·5 | % 50·8 | % 50·4 |
| | 39–63 | 44–61 | 43–57 | 51–64 | 43–57 | 45–61 | |
| After operation | 52·0 | 53·7 | 51·9 | 59·5 | — | — | — |
| | 39–67 | 44–69 | 41–58 | 52–67 | — | — | — |

TABLE VII.—*Average Cardiothoracic Ratio at each Year After Operation (Very Good and Good Results Combined)*

| No. of Cases | Cardiothoracic Ratio | | | | |
|---|---|---|---|---|---|
| | Before Operation | 1 Year After | 2 Years After | 3 Years After | 4 Years After |
| | % | % | % | % | % |
| 19 | 48·0 | 53·4 | 53·8 | 53·5 | 53·5 |
| 28 | 48·5 | 51·6 | 5·.9 | 51·8 | — |
| 35 | 49·7 | 52·7 | 52·1 | — | — |
| 32 | 50·0 | 52·7 | — | — | — |

In most instances any decrease during the period of observation probably results from a relatively greater increase in the chest than in the heart. Nearly all the patients in whom the heart later became smaller were about the age of puberty, a period when rapid growth of the chest may be expected. Only three of those who showed little if any improvement had any increase in heart size—presumably a reflection of the inadequate size of the shunt. There are, however, many patients who show very good results but no increase in the size of the heart. These patients and the others in whom the increase is only moderate and non-progressive must represent the ideal, where the general benefit to the patient is correctly balanced against the increased load on the heart. A smaller shunt would not give the benefit sought and a larger one might overburden the heart. It is unfortunate that so far we lack the knowledge and the means to tailor the anastomosis so as to achieve this optimum in everyone, but it is encouraging that, despite this, so many do achieve it.

There are also the patients in whom the shunt is too large as judged by the increase in the size of the heart and by

other changes. Symptomatically, these patients do very well unless the shunt is so much too large that congestive failure develops.

Twenty patients are doing well but have shown an increase in heart size that is progressive or at least greater than one would wish. In ten of these, all but one of whom showed very good results, it has shown no tendency to be progressive. In seven of the ten the increase was 7–9 points, but the heart was small, so the cardiothoracic ratio is still between 53 and 55%, and there seems no reason why the very good result should not last. In the other three the increase was only 6–7 points; but the heart was larger before operation, so that the ratio now is between 58 and 60% : in none does it exceed 60%, and it is hoped this will continue, though the long-term prospects of these patients can hardly be as good as in those with smaller hearts.

Of the ten patients in whom the heart has so far enlarged progressively three were not improved (Cases 9 and 10) or died after some initial improvement (Case 6); none of these had Fallot's tetralogy, two having tricuspid atresia and one some more complex lesion. The remaining seven all continue to show very good results, most of them for three years or more. Three had small hearts before operation, so that an increase of about 10 points still leaves them with cardiothoracic ratios between 52% and 55%, and, provided the increase now stops, they should continue to do well. The last four have ratios between 60% and 67%, either because the heart was larger at the start (one case) or because the increase was from 12 to 15 points (three cases) instead of the average of about 3 points. Case 15, the patient with the largest increase, has already been described. These four are the only patients who make us anxious about the possible onset of congestive failure in the near future, though in all other ways they are extremely well. The increase may be reversed, as has happened at least once (Case 18), but we can have no assurance of this.

The outlook is certainly better if the heart is smaller. The proportion of very good results to poor results combined with those in which the patients showed no improvement or died is 3.5 : 1 where the c.t.r. was 49% or under and 1 : 1 where it was above this.

We suggested earlier (Baker *et al.*, 1949) that the heart size helped in choosing patients suitable for operation, and that if the c.t.r. was above 52 or 54% less certainty could be felt about a good result. This is borne out by longer experience, but it is clear that there are several (nearly 10%) with a c.t.r. of 55% or above who have shown a very good result, even if it has less chance of lasting. So far these patients do not include an undue proportion of those with hearts getting progressively larger. One of the largest hearts was in this group, but the largest of all (c.t.r. 62%) was in Case 4; this patient was doing very well and showing some decrease in the heart size when he died with a cerebral abscess.

## Progress after Operation

We have written so far, except in the discussion on the heart size, as if these changes occurred at once after operation and remained unaltered thereafter. A survey of the 107 patients followed for two years or more shows that this is generally but not always true. The first few months usually see all the main changes established.

The increased capacity is obvious almost immediately after operation, and the only patients not easy to assess early are the very young and those with such poor muscular development that they need some time to show their improvement (see Case 17 below). Apart from these, only eight patients were noted as improving further a year or two after the operation; in some this was probably no more than increased confidence, but in others, as judged by the objective signs, it seemed real organic improvement. These are balanced by six in whom there has been some suggestion of deterioration. In two this was due to the development of pulmonary tuberculosis and in two others to chronic infections of the upper respiratory tract. There are, gratifyingly, only two in whom slight objective deterioration and a lessening murmur suggest that the anastomosis may be getting smaller. Even if this is so, and it is not certain, they form such a small proportion of the whole that they need not be regarded as early examples of what is likely to befall the majority at a later date.

*Post-operative Complications.*—Immediate complications have mostly not affected the results. There were three patients with Horner's syndrome on the side of the operation. Five patients had some intracranial thrombosis after operation, but the paresis has made such an excellent recovery that none find it a source of major disability, except one in whom a cerebral thrombosis has left him less intellectually able to do his work. Two patients showed signs of failure soon after the operation : this lasted a few weeks and cleared up spontaneously or with digitalis, but both have done well since.

*Bacterial Endocarditis and Cerebral Abscess.*—Later complications of the operation have, apart from the cardiac failure and deaths discussed elsewhere, been few. Rather surprisingly, bacterial endocarditis has not been troublesome, though Taussig *et al.* (1951) report 18 cases in their series of 1,000, two-thirds of them occurring soon after operation. Only one of our patients was treated (elsewhere) for subacute bacterial endocarditis, and the diagnosis was not established; subsequently he had a very good result (Case 17). In these patients a cerebral abscess has been found much more often than bacterial endocarditis. This is not the result of the operation but is connected with the right-to-left shunt which remains; it has also been more common than bacterial endocarditis among other patients.

Five patients developed signs of a cerebral abscess, 3, 4, 18, 26, and 34 months after their anastomosis. All except Case 4 (q.v.), who died, have made a good recovery after a further operation, but with some residual symptoms—for example, one has a visual defect of one quadrant and would otherwise be passed as normal except by those who know him well, to whom he seems to have lost something of his exceptional energy and enthusiasm. Two others have slight residual weakness but are able to lead reasonably normal lives. One other was admitted as a suspected case of cerebral abscess, but was thought to have a cortical thrombophlebitis; she recovered with conservative treatment but did not quite regain her previous activity.

## Results in Special Groups

So far we have discussed the 200 cases as a whole, but some special groups are worth separate consideration. Two can be distinguished easily—tricuspid atresia and various forms of dextrocardia and isolated laevocardia—and a third, pulmonary atresia, with less certainty. It is not possible to separate the last completely, because the diagnosis can be certain only in fatal cases, and some successful ones may be listed as Fallot's tetralogy.

### Tricuspid Atresia

There were seven cases of tricuspid atresia with a non-functioning right ventricle; this diagnosis was made when there was left ventricular preponderance on the electrocardiogram and left ventricular hypertrophy on radioscopy. All these seven survived the operation; but this must be regarded as good fortune, as it cannot be safer than in Fallot's tetralogy. It seems, however, that it is at least no more dangerous.

Two of the seven died within a year (Cases 6 and 10). The other five have all done well, three of them for two or three years. One has been described (Case 3). Another boy, aged 19, has been regularly at light manual work for two years and can walk four miles with ease, instead of half a mile with difficulty. His heart was very small, so that an increase in the c.t.r. from 39% to 46% still leaves it small.

A third boy, aged 8, was extremely blue, and walking 20 yards left him exhausted. At first it was difficult to say

how much his operation had helped, as he had such poor muscular development that he could not do much. Each year he has improved, so that now, after three years, he can run 50 yards or walk five miles and is regular at school. He has grown 6 in. (15 cm.) and put on 21 lb. (9.5 kg.) in weight (Case 17).

These three compare well with the best results in Fallot's tetralogy and have maintained their improvement for two or three years without much increase in heart size. The other two have done well but the heart has increased rather more in the first year, so longer time is needed to know if they will continue as good.

The difference between the successes and the two deaths may be related to the hearts being larger in the latter, although in one who did well it was nearly as large. The heart size seems the best guide to whether a patient with tricuspid atresia is likely to maintain the improvement after operation: if the c.t.r. is under 54% the good result may last at any rate for several years, but if it is 60% or more it is less certain, though there is not yet enough evidence to decide that operation is not worth doing.

The results of Taussig *et al.* (1951) show a mortality of 26% within six months of the operation. Our results are similar and not as good as in Fallot's tetralogy. The natural prognosis of tricuspid atresia is, however, much worse than that of Fallot's tetralogy. There have been six deaths in the 31 cases of tricuspid atresia among 1,124 cases of congenital heart disease seen in 1947–51. This is a higher mortality than in any other group, except perhaps in pulmonary atresia and transposition of the aorta and pulmonary trunk.

### Dextrocardia and Isolated Laevocardia

There were six cases in this group—two with dextrocardia and complete situs inversus, one with isolated dextrocardia, and three with laevocardia but with the abdominal viscera transposed. In cases such as the last four the congenital defects are generally very complex. The mortality in this group was very heavy and half the patients died. These three have been described in the section on deaths after operation (Cases 12, 13, and 14).

Three patients survived, two doing extremely well. One (Case 18) had dextrocardia and situs inversus, probably with Fallot's tetralogy: after operation she could walk three miles instead of a few hundred yards. Her heart increased in size soon after operation, but became smaller during the next two years (c.t.r. 58 to 67% to 63%). Another, who had a continuous murmur that was thought to indicate bronchial artery blood supply to his lungs, probably with complete situs inversus, was able to lead a normal life without much increase in the size of the heart (Case 19). The third, with laevocardia and transposed viscera, was only slightly improved.

Very good results may therefore be obtained but cannot be expected with confidence in dextrocardia and isolated laevocardia. The results of Taussig *et al.* (1951) are better than these, but agree in showing a rather heavy mortality of 17% directly, and another 12% during the ensuing six months.

### Pulmonary Atresia

Cases of pulmonary atresia are more difficult. The difference in the surgical treatment of this group and of Fallot's tetralogy is one reason for their separation (Allanby *et al.*, 1950), but the distinction is often difficult, and some cases of atresia are probably left undiagnosed among the cases of Fallot's tetralogy. This will give an unduly pessimistic view of the results of operation.

In a patient with the general picture of Fallot's tetralogy but with a loud and single second sound in the pulmonary area and without a systolic murmur the diagnosis of pulmonary atresia is likely. If the cardiac outline shows a very sharply defined pulmonary bay with an absence of a pulmonary trunk the diagnosis becomes more certain, and is further supported if there are also the special features that indicate a bronchial artery blood supply to the lungs—isolated nodular shadows in the hila and heavy vascular markings in the right upper quadrant caused by bronchial arteries passing from the arch of the aorta (Campbell and Gardner, 1950). There may be a continuous murmur that is indicative of pulmonary atresia (or sometimes truncus arteriosus) with a collateral blood supply through a persistent ductus or bronchial arteries.

Generally, angiocardiography should be used to confirm the presence of adequate pulmonary arteries beyond the atresia, but in patients with recurrent attacks of unconsciousness we have sometimes thought that thoracotomy is the lesser risk. It should never be dispensed with in this group on other grounds, even though there seem to be pulmonary arteries on radioscopy.

(*a*) *With a Continuous Murmur.*—There were nine of these cases (including the two considered in the dextrocardia group). Two died soon after operation (Case 14 and another); in a third no artery adequate for an anastomosis could be found. In the other six the surgeon succeeded in carrying out a subclavian-pulmonary anastomosis, though in three this had to be end-to-end: all these patients were improved. The first, instead of being an invalid who could walk 200 yards, could soon lead a normal life and walk four miles, including many Devonshire hills; her cyanosis was much less and her haemoglobin percentage returned to normal. Her heart, which had been small, increased soon after operation (c.t.r. 43 to 53%) but no further in the next three years, and her improvement is maintained. Case 19, with an equally good result after two years, has been described. The other four have not been followed for so long; all are improved, including two who had end-to-end anastomoses (one very good, two good, and one somewhat improved). All were thought to have patent pulmonary arteries beyond the atresia, one with a persistent ductus and the other three with a bronchial artery blood supply to the lungs.

(*b*) *Without a Continuous Murmur.*—There were 12 cases: five of these died, two before the special problems of this group had been fully appreciated, and three later, when we were well aware of these. In four more no operation was possible as no artery suitable for an anastomosis was found: two of these were early cases, but the two recent ones, in which the mistake should have been avoided, were accepted as having adequate pulmonary arteries—one after radioscopy and one even after angiocardiography.

These results are so bad that pulmonary atresia might seem unsuitable for operation, but two of the three good results were outstanding, and it seems unlikely that those without a continuous murmur are worse subjects for operation than those with such a murmur. As an example, one boy was deeply cyanosed and could hardly walk at all; now, two years later, he goes to an ordinary school and is ready to spend his time fighting with the other boys, and hardly shows any change in colour. There is no undue increase in the size of his heart. The clinical diagnosis of pulmonary atresia with a bronchial artery blood supply seems almost certain. In the third the improvement was only moderate.

## Comparison of Fallot's Tetralogy with other Groups

Discarding the 32 cases of the special groups described and three cases with transposition (one with the addition of pulmonary stenosis) wrongly put forward for operation, there remain 165 cases in which the diagnosis was simple Fallot's tetralogy. We know that, in fact, two of these had a single ventricle and another an aorta that arose entirely from the right ventricle, but these are points that cannot be diagnosed with certainty on clinical grounds, so we have left them with the group of Fallot's tetralogy. Even so, the results for Fallot's tetralogy are very much better than for the other groups, and the operative mortality is 8% and the expectation of a good result 75%. In the more complicated groups, on the other hand, the mortality so far has been nearly 30%, and the good results only about 35%. Unless this result can be improved with further experience, operation should be advised in these cases only when they seem favourable and only in special centres where every facility for investigation and surgical treatment is available.

**Summary and Conclusions**

The results of 200 anastomotic operations in patients with morbus caeruleus are described, nearly all of them having been followed for more than a year and several for four years. Most had Fallot's tetralogy, and here the results were excellent. Many were seriously ill, but only 8% died and 75% benefited greatly, so that they can walk a mile or more and lead lives that are almost normal without too much physical activity. In the smaller number of more complex lesions, such as tricuspid atresia, dextrocardia, and pulmonary atresia, the results were much less good. The total mortality was nearly 30%, and only 35% obtained the good results. More experience is needed to decide which of these patients can hope to benefit from operation, but some tentative suggestions are made.

Most patients have an increased capacity and lose much of their cyanosis, clubbing, and polycythaemia. Some cyanosis, at least on exercise, and some clubbing generally remain unless they were very slight. The haemoglobin usually falls from between 120 and 160% to between 90 and 120%. In nearly all the successful cases a continuous murmur can be heard at the site of the anastomosis, and the presence of this soon after the operation is often one of the best indications of success. The pulse pressure may be a little wider as the result of the anastomosis, but not often much wider.

Generally the improvement gained in the first few months after operation is a good guide to the future and is maintained. The annual mortality rate is not much above 1%, and this includes the risks to which these patients are inevitably exposed whether they have an operation or not.

Special attention has been paid to the increase in the size of the heart because this has been thought to be the great drawback to the operation. Generally the increase is not great, and is not progressive after the first few months. The cardiothoracic ratio may increase quickly from 48 to 52% and then remain constant during the period of observation. A few patients in whom the increase is great or even progressive make us anxious lest they may develop congestive heart failure, but except for three with complex lesions in whom this happened within the first six to nine months no others have yet done so.

It is a pleasure to express our thanks to the surgeons who have made this work possible: Mr. R. C. Brock, who has operated on the largest number, Mr. T. Holmes Sellors, and Mr. Ian Hill. Mr. Holmes Sellors has added to our indebtedness by the great trouble he and his surgical staff have taken in keeping us so fully informed about patients operated on in another hospital. We should also like to thank the staff of the cardiac department, who during the last four years have helped with the follow-up at different stages, the doctors who have sent us reports on their patients, and Miss M. Waldron, of the department of illustration, Guy's Hospital, for the charts.

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# RADIOACTIVE PHOSPHORUS ($P^{32}$) IN TREATMENT OF MENORRHAGIA*

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If menorrhagia persists after curettage in the over-40 age group a decision on radical treatment either by induction of the menopause or by surgery may be necessary to avoid anaemia and increasing general debility. Both methods have disadvantages, however; radium or deep *x* rays practically castrate the patient, so that flushings and premature age-effects such as senile "spotting" of the vagina or introital fissuring may develop (McLaren, 1950). Surgery with ovarian conservation may avoid these sequelae, but it carries a small but definite mortality and morbidity.

The idea of avoiding radical treatment by destroying the endometrium is not new; ten Berge (1936) in Holland, for instance, has described a method of uterine thermocautery which had some success. Our present experiments of irradiating the uterus with beta particles were conducted on the suggestion of Dr. A. G. Glucksmann. As beta particles from a source such as radioactive phosphorus are stopped by the surface layers of tissue it seemed that surface endometrial cautery and uterine haemostasis might be achieved without destroying ovarian function.

The difficulty in constructing an applicator which would pass the human cervix yet allow irradiation of the whole of the cavity of the uterus led us to use a balloon on the lines described by Simon (1949) and by Douglas and his colleagues (1950, 1951). The human uterus, however, is not distended without a high pressure of air or fluid, and this led to difficulty in containing the balloon *in utero*. In the end a simple rod-shaped applicator designed by one of us (J. C. H.) was used, and proved to be easy and safe to handle. Despite the shape of the normal uterine cavity a wide area of endometrium was burned by $P^{32}$.

## The Applicator

In the preliminary experiments three layers of filter paper were wound tightly around an acrylic resin rod 4 cm. long and 1 cm. diameter (Fig. 1). An aqueous solution of sodium phosphate containing the radioactive phosphorus isotope $P^{32}$ was distributed evenly over the filter paper and evaporated. The completely dry filter paper was coated with a solution of acrylic resin.

In later experiments moist filter paper was moulded over the whole surface of a cylindrical "perspex" former with a hemispherical upper end. An aqueous solution of sodium hydrogen phosphate in which the phosphorus is radioactive was allowed to evaporate to produce a dry surface evenly covered with $P^{32}$. A thin sheath of perspex (0.25 mm. thick) was slipped over the filter paper and a seal made with perspex solution in chloroform at the lower end. This applicator has two advantages: (1) it is easier to assemble, more robust, and less likely to contaminate the patient; and (2)

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