MORBUS CŒRULEUS

A STUDY OF 50 CASES AFTER THE BLALOCK–TAUSSIG OPERATION

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The Blalock-Taussig operation for morbus cœruleus was described in February 1945. The position reached in September 1947 was presented to the International Conference of Physicians at that time and has since been published by Taussig (1948) and Blalock (1948*a*). The steps that led up to this operation and the subsequent developments have been described recently (Campbell, 1948), so will not be referred to further. The early results of the first 18 operations at Guy's Hospital, some by Mr. Blalock and some by one of us (R.C.B.), have been described (Campbell, 1948); the earlier cases had then been followed for six months.

In the present paper we wish to describe the results in the first 50 patients operated on at Guy's Hospital with a systemic-pulmonary anastomosis for cyanotic heart disease, all since September, 1947, by R.C.B.

All the patients were thought to have Fallot's tetralogy or some closely related form of congenital heart disease, except three (Cases 33, 42, and 49); these three had a similar clinical picture, but with left ventricular preponderance in the electrocardiogram, which nearly always indicates tricuspid atresia or stenosis with a non-functioning right ventricle (Brown, 1936). The earlier cases have now been followed for from 12 to 15 months and the most recent for 6 months. We are not including in the figures that follow 6 patients who have been submitted to operation for pulmonary valvulotomy during this same period.

Age incidence. Most of the patients were between 3 and 15 and there were rather more boys than girls (28 to 22). One was two and a half years old, 5 were three, 32 were between four and ten, 7 between eleven and sixteen, and 5 (by chance, all men) were between 19 and 27 years of age.

Blalock has suggested 3 as the minimum age for operation except in emergencies, and 10 as the ideal upper limit. The operative risk is much greater in patients under 3 and there seems more risk that cyanosis will return, perhaps because the anastomosis fails to grow as the child develops. Partly because these two risks are not likely to diminish suddenly at 3 years of age, and partly because with the large numbers waiting it seems less harmful to delay operation from 3 to 5 than from say 10 to 12, we have tended, latterly, to make 5 years the earliest age.

We have, however, no evidence from our cases that operation is more dangerous or less successful at 3 years of age. We do, so far, find it more dangerous in those over 20, but it will be many years before one can avoid the dilemma of operating on older patients with an increased risk, or allowing them to deteriorate and die.

Symptoms and Signs

Disability. The disability of these patients was extreme: many selected for operation were severe cases who were deteriorating and could wait no longer, rather than good operative risks. Twenty-six were put in the most severe grade (IV) which means that they were made dyspnœic by a few steps and could rarely walk more than 25 yards (Campbell, 1948). One of these (Case 43) said he had once walked 100 yards as a great occasion; as he was 13 and attended an ordinary school in his wheeled chair it emphasizes the disability. Another 19 were in the next grade (III) which means very severe limitation as they could not play outside without frequent rests and could only walk 50 to 200 yards. Three of these, aged 19, 25, and 27, had at one time walked 2 or 3 miles but had become much worse during the last few years.

There were 5 who could do something more than this and were placed in grade II, but even they were very incapacitated. Two, aged 19 and 16, had been

able to walk one or two miles slowly but were most dissatisfied with being "unable to do anything" and were enthusiastic for operation, whatever the risks. The other three were younger and could walk half to one mile on a good day, but often they could not do as much as this. As further evidence that even these were moderately severe cases, all except one had hæmoglobin percentages between 150 and 126.

The large proportion of older patients among the last eight suggests that they had survived because their condition was less severe but was now deteriorating. All cases of Fallot's tetralogy have not, of course, such severe disability, but the worst have been chosen for early operation.

In addition, some of these patients were becoming rapidly worse so that operation was expedited. For example, Case 46 attended a second time after three months: the distance she could walk had shortened from 150 to 50 yards; her cyanosis and the clubbing of her fingers had become worse. The slightest exertion, even dressing, provoked almost daily attacks of loss of control of her limbs with semi-consciousness.

Our experience suggests that when a patient with Fallot's tetralogy starts deteriorating the prognosis is grave, and several times when this has happened death has not been long delayed.

Cyanosis. All these patients had been cyanosed from early infancy and all had clubbed fingers. The measurement of cyanosis is difficult and is made more so by its quick variation with exertion and temperature. In bed in a warm ward many of the patients look so much less blue than as out-patients that they are hardly recognizable. With Dr. W. D. Brinton we made some attempts at colour matching but they have not so far been successful. The relationship of cyanosis and polycythæmia is reciprocal, the lack of oxygen causing the polycythæmia and this in its turn increasing the appearance of cyanosis.

The estimate of cyanosis was made without knowing the hæmoglobin percentage. In general terms, those in whom it was from 110 to 129 had been placed in grade II or III as regards their cyanosis; those from 130 to 139, in grade III; and those from 140 to 160, in grade III or IV. These and some other details are given in Table I. But there was no very close correspondence and sometimes rather surprising contrasts.

The cyanosis was generally severe (grade III,* 25 cases) or very severe (grade IV, 15 cases). In 9 it was less than this and often less than would have been expected from the disability, though it was always present even with the patient at rest: in these 9 the hæmoglobin averaged 122 per cent.

In one patient in particular (Case 26) where the disability was very great, the cyanosis and clubbing slight, and the hæmoglobin 116 per cent, we were somewhat hesitant about operation but the result was just as successful as in others. The arterial oxygen saturation was 80 per cent falling to 75 per cent with very trivial leg movements (Dr. Zak). If the disability is severe enough, relatively slight cyanosis should not as a rule be a contra-indication to operation.

Onset of cyanosis. The age from which cyanosis was first noticed is of great importance. Naturally, if there is an over-riding aorta and a ventricular septal defect, venous blood will be passing into the aorta from birth or soon after. Cyanosis may be noted at once depending on the amount of shunt, and will become more obvious as greater demands are made on the circulation and as polycythæmia develops. It is most important not to mistake temporary cyanosis *at birth* for cyanosis that has persisted *from birth.*

Cyanosis was noted from an earlier age than 18 months in all except one. The actual figures were from birth or soon after in 30 of the 50, from between 2 and 6 months in 9, from between 7 and 10 months in 6, from between 15 and 18 months in 4, and at 24 months in Case 32.

Of the 5 where cyanosis was not noted till after 15 months, 4 seemed ordinary cases of Fallot's tetralogy though one lived to 27: the fifth (Case 32) had pulmonary stenosis and transposition of the aorta so that cyanosis might have been expected from birth. It is strange, but can hardly be more than a coincidence, that 3 of these 5 patients died after operation.

Polycythæmia. The hæmoglobin percentage varied from 110 to 160 and averaged 137. The highest figures were 160 (Case 35), 158 (Case 29), 157 (Case 6), and 153 (Case 22). All these except the first looked severely polycythæmic.

The red blood cells were generally between 6·0 and 9·0 and averaged 7·8 millions, but in two it was 11·6 and 11·1 millions (Cases 41 and 47), the next highest being 10·1 million and several of 9·0 million or just over. Curiously enough these two had not specially high hæmoglobin percentages, the figures being 140 and 137 so that the colour indices were unusually low—0·58 and 0·60 respectively.

The hæmatocrit reading was generally between 60 and 85 and averaged 74. As might be expected, it generally agreed more closely but not very closely with the red cell count. The highest readings were 94 (Case 41, one of the highest red cell counts), 92 (Case 29, one of the highest hæmoglobins), 88, 87,

* Grade III. Cyanosis moderately severe *at rest* and obvious at a glance. Grade IV. Cyanosis gross, *at rest.*

TABLE I

CASES OF MORBUS CŒRULEUS SUBMITTED TO BLALOCK-TAUSSIG OPERATION AT GUY'S HOSPITAL

Case No.	Initials	Sex and Age	Cyanosis	Disability	Hæmoglobin (percentage)	Red cells (millions)	Hæmotocrit	Hæmoglobin 3 or 4 weeks after operation	Reference No.
1A	A.C.	M 19	4	3	148	10·0	78	Died	P01A
19	S.F.	M 4	2–3	3	112	6·0	58	100	CB04
20	M.E.	F 5	2–3	3	110	5·9	63	100	0018
21	T.G.	M 4	3	3–	138	8·0	(68)	127	C004
22	M.L.	F 13	4	4	153	7·2	87	N.C.*	C005
23	D.S.	M 19	4	4–	130	6·0	77	106	0015
24	C.M.	F 6	4	4	148	8·8	81	128	0019
25	J.R.	F 6	2–3	4	140	7·4	70	101	0005
26	J.L.	M 15	2	4	116	8·4	57	88	P069
27	D.F.	F 10	3+	3	150	8·0	83	N.C.	0027
28	J.L.	F 3	4	4	107	6·6	64	N.C.	H113
29	T.H.	M 10	4	4	158	8·2	92	122	0036
30	G.F.	F 7	3–	3–	133	8·0	72	—	P045
31	V.S.	F 2	2–3	2–3	136	7·4	72	110	C008
32	J.H.	M 11	3–	4	144	8·0	74	Died	P054
33	P.S.†	M 4	3	4	132	9·2	85	109	0022
34	A.W.	F 9	3	2+	118	6·4	63	97	H128
35	M.C.	F 7	3	4	160	7·9	67	Died	P037
36	P.R.	F 16	2+	2+	126	5·3	58	89	C007
37	J.W.	M 5	4	4	136	8·4	81	103	P102
38	C.B.	F 7	3–	4	142	10·1	81	Died	H106
39	P.C.	M 10	3	3	139	8·3	87	114	CB09
40	D.S.	M 5	3+	3	140	7·7	71	105	P089
41	J.H.	F 5	4	4	140	11·6	94	115	CB17
42	S.H.†	M 19	3	2+	144	8·0	—	112	P057
43	T.H.	M 13	3	4	142	7·2	69	—	P056
44	G.S.	F 11	4	4	142	8·6	84	110	H120
45	S.N.	M 3	2	4	109	6·6	—	92	0017
46	K.C.	F 15	3	4	135	8·9	76	—	0105
47	G.K.	M 11	3	3	137	11·1	—	115	0184
48	R.B.	M 27	4	3	144	9·1	85	Died	0115
49	J.C.†	M 8	4	4	125	8·6	73	123	0007
50	M.S.	F 13	3	2+	138	7·2	—	110	0140

* N.C.=No change as no effective anastomosis. † Tricuspid atresia.

Cases 1–17 have been described previously (Campbell, 1948).

and 86; several others were very close to this.

As would be expected, the red cell count increases regularly with the hæmoglobin percentage in the lower ranges, but to our surprise this parallel increase is not continued in the higher ranges. In Table II the cases have been classified according to the hæmoglobin and it will be seen that the red cells and hæmatocrit increase as the hæmoglobin goes up from 112 to 134, but there is no further increase either in the red cells or in the hæmatocrit as the hæmoglobin goes up from 134 to 153. The disadvantages of a red cell count over 8·0 million may produce some mechanism in the body that prevents a further rise, but if so the two cases with counts over 11·0 million are all the more surprising.

Apparently as the hæmoglobin rises above 130 per cent the red cell count and hæmatocrit do not on the average rise further with the result that the colour index rises towards unity.

Clubbing of the fingers and toes. All these cases showed moderate or severe clubbing of the fingers and toes, and Case 26 (whose hæmoglobin was 116 per cent) was the only one who did not have the complete picture as he had curvature of the nails without any noticeable broadening. Fifteen were marked as having moderate rather than severe clubbing and these included 8 with hæmoglobin percentages of from 110 to 126, but the others were about 140. Six were marked as having unusually severe clubbing: their hæmoglobin ranged from

TABLE II

AVERAGE BLOOD COUNTS GROUPED BY THE HÆMOGLOBIN LEVEL

Hæmoglobin		No. of cases	Average red cell count (millions)	Average hæmato-crit	Average colour index
Range	Average				
110–119	112	8	6·5	62	0·86
120–129	124	4	7·2	63	0·86
130–139	134	12	8·3*	77	0·81*
140–149	145	16	8·2	80	0·88
150–160	153	10	7·9	76	0·97
110–129	116	12	6·7	63	0·86
130–160	144	38	8·1	78	0·88
All cases	137	50	7·8	74	0·87

* Without the two cases with counts over 11·0 million these figures would be 7·8 and 0·85.

130 to 160 per cent. This correlation between clubbing and polycythæmia might be expected. Fig. 1 and 2 show severe clubbing of the same fingers a year after operation when the degree of clubbing has become much less.

Squatting. Of these 50 patients, 41 had a history of squatting and 7 gave no such history; there were 2 with no information. Fig. 3 illustrates the typical position. Taussig considers squatting almost constant in Fallot's tetralogy, but our evidence is against this though we find its does occur in about 80 per cent. It seems unlikely that the diagnosis was wrong in 7 who were not squatting. One had tricuspid atresia (Case 42) and the operation was as successful in him as it was in 3 others who did not squat (Cases 5, 10, and 36). It is curious, but again perhaps no more than a coincidence, that 3 of the relatively small number of unsuccessful results (Cases 11, 22, and 27) occurred among this small number of non-squatters.

Mental and physical development. It is surprising that the mental development should be so good, considering the severity of the anoxæmia. All but two of these children were normal or often above the average, although, of course, many were educationally backward. We have used the age of walking and talking as rough measurements.

Generally the child walked at a normal age but a few were late. Most children (33 cases) started walking at or before two years, but it was delayed in 12 till three years, in 2 till four years, in 1 till five years, and in 2 till seven years. There was no mental defect in these children and the last two boys seemed of average or more than average intelligence.

Most children (38 cases) started talking at a

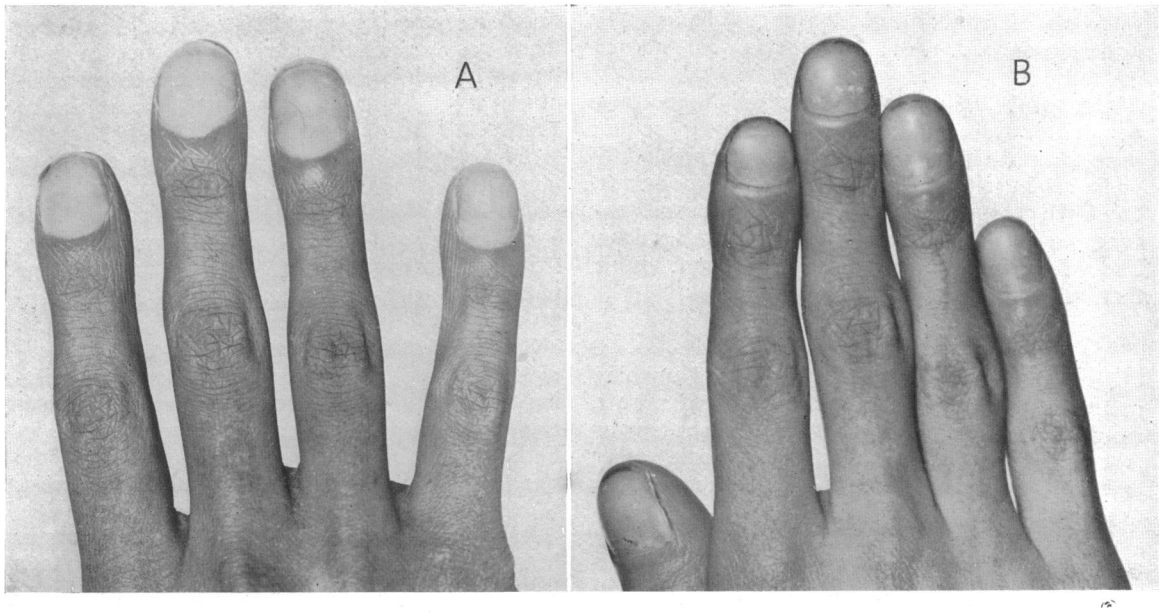


FIG. 1.—Clubbing of fingers and its disappearance after operation. This position shows the disappearance of the broadening from side to side and some diminution of the curvature. With moderate clubbing it may disappear completely in five or six months but with a more severe grade such as this, something is left permanently and the illustration is nearly a year after operation. Case 23.

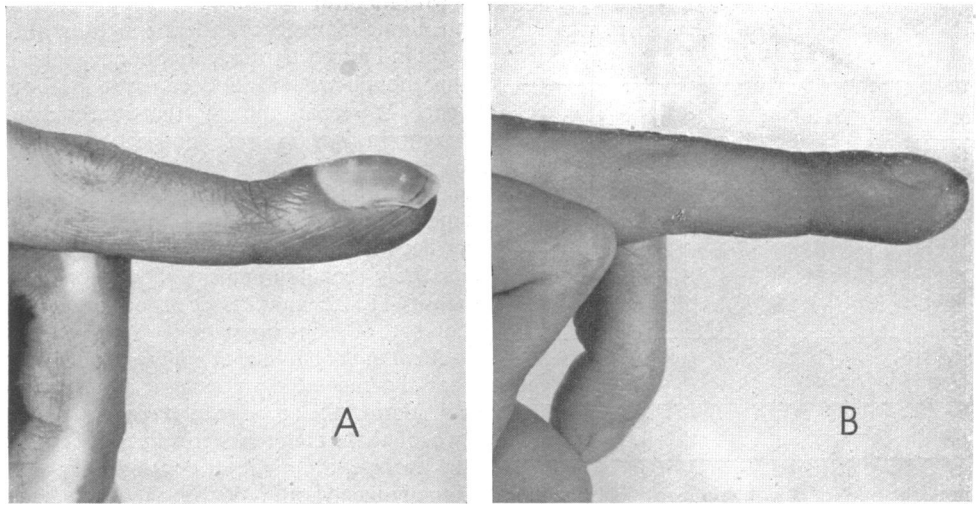


FIG. 2.—Clubbing of the fingers before and a year after operation, showing diminution of the broadening from front to back and of the curvature, and a more normal appearance of the skin. Case 23.

normal age, generally between 12 and 18 months. Seven were said to have been able to talk at two years, 2 at two and a half years, and 3 at three years.

Only 4 (Cases 19, 28, 35, and 49) were slow both in walking and talking. Case 19 was mentally backward and Case 35, who had a cataract also, following maternal rubella during pregnancy, was rather backward. All the others were normal mentally.

Most of the children were under weight and their parents always complained of difficulty in getting them to eat. On the average they were only slightly under height so that their thinness was very obvious. Often the chest was badly developed with some tendency to pigeon chest and to a Harrison's sulcus. The veins over the chest were often more visible than normally.

PHYSICAL SIGNS

The heart is generally of normal size and anything more than very slight enlargement should raise a suspicion that the lesion is not Fallot's tetralogy or is complicated by the addition of some other defect. The size and shape is dealt with more fully in the section on radiology.

Systolic murmur and thrill. There is generally a *systolic murmur* in the second, third, and fourth spaces on the left, becoming fainter towards the apex. The murmur is often loud enough to be conducted widely, sometimes to the right side or to the back. Sometimes the pitch of the murmur differs over the pulmonary area and lower down in the fourth left space and this may be due to there being two different murmurs produced by the pulmonary stenosis and by the ventricular septal defect.

The loudness and harshness of the murmur varies a great deal. There were 4 cases where no systolic murmur was heard (Cases 9, 22, 41, and 48); all were of severe degree and in the last, who died, there was a calcified pulmonary valve. In 2 of these 4 and in 2 others, triple rhythm with an addition of a third heart sound was so obvious as to be almost the main physical sign. It was equally common for the murmur to be described as soft or faint (13 cases); as average (13 cases); or as loud, rough, or harsh (14 cases+6 special cases). The 6 special cases with a harsh or rough murmur were the 3 with tricuspid atresia, Case 28 with a large pulmonary artery, and Cases 32 and 35 with unusual features discovered post-mortem (see page 192).

Such a harsh or rough murmur (or thrill of well-marked intensity) is, therefore, a reason for considering carefully if the diagnosis is correct, though it may be found fairly often in an ordinary case of Fallot's tetralogy, perhaps indicating that the stenosis is fairly severe; but not that there is atresia, when a murmur may be absent.

In none of these cases was any diastolic murmur heard, but some others with the features of Fallot's tetralogy and a diastolic murmur have been deferred for fuller investigation.

A *thrill* was present in more than half, but generally it was faint and might only be felt at times or after exertion. It was usually maximal

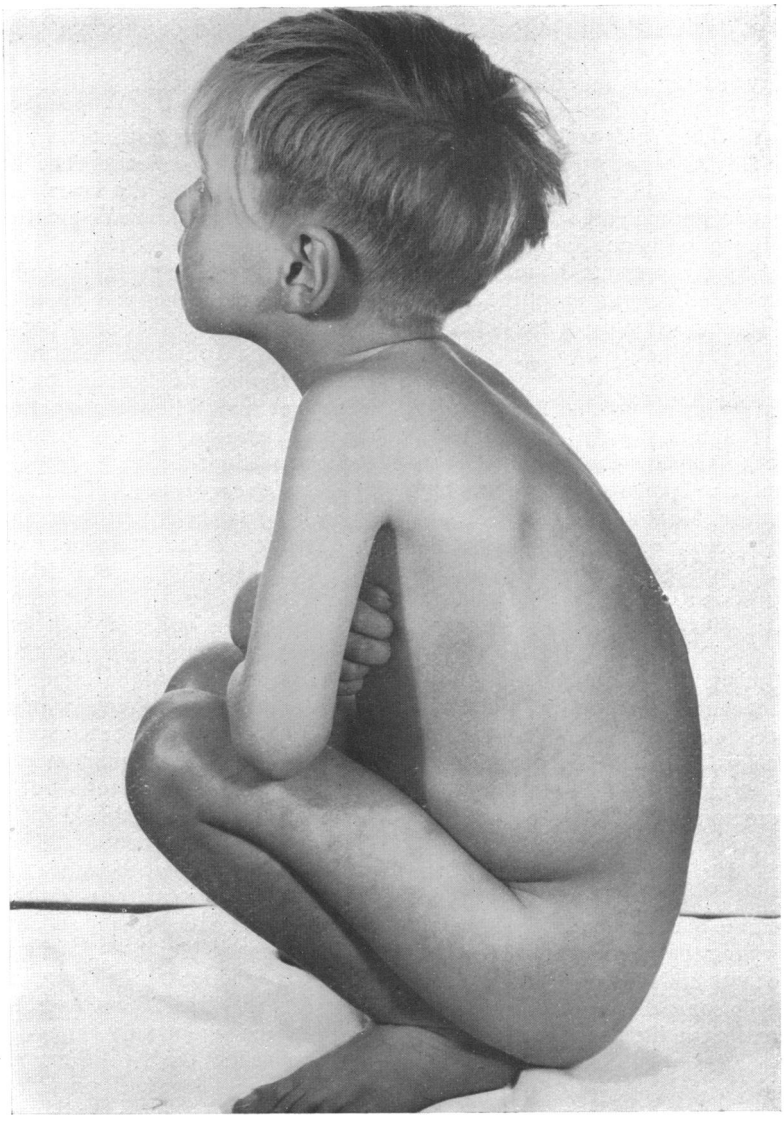


FIG. 3.—Case 33 with tricuspid atresia and non-functioning right ventricle. A typical position adopted in squatting, though in many of the older patients the knees are brought still closer to the chest.

in the pulmonary area but occasionally towards the apex. As would be expected it was closely correlated with the harshness of the systolic murmur. The thrill was never of great intensity; it was of moderate intensity in 13 cases, but this included the 3 with tricuspid atresia and the 2 with unusual findings post-mortem. It was difficult to feel or only felt occasionally or after exercise in 17 cases. No thrill was felt at the many examinations in 20 cases.

Pulmonary second sound. Auscultation should be carried out with the patient sitting up and lying down and during each phase of respiration, before a decision is taken about the intensity of the second sound.

Diminution of the second sound in the pulmonary area has been traditionally regarded as an important sign in the diagnosis of pulmonary stenosis. As far as Fallot's tetralogy is concerned this is not so; the second sound is generally normal and is as likely

to be a little increased as diminished. It was often as loud on the left as on the right side at the base.

Any great increase, however, is likely to indicate that the pulmonary pressure is raised and that on screening a large pulsating pulmonary artery will be seen and that the lung fields will be congested instead of clear. This is specially true if the second sound has a drum-like quality. Visible pulsation in the pulmonary area, and palpable diastolic shock also suggest that the lesion is not Fallot's tetralogy and that the pulmonary pressure is raised. We would emphasize that the significance of the change in the pulmonary second sound depends on a sound that is *much increased* and not to one that is *slightly increased.*

In these 50 cases it was recorded as normal in 17, as diminished in 11, and as slightly increased in 13; in 9 it was more notably increased. These 9 included one where the pulmonary pressure was high (Case 28), one with tricuspid atresia (Case 42), one with severe cyanosis who was hardly helped by operation (Case 22), and one who had infundibular stenosis and a transposed aorta (Case 32); but the other five seemed ordinary cases who were helped by operation.

No case of Fallot's tetralogy has had a drum-like pulmonary second sound, even if the pulmonary artery was more prominent than usual. A diastolic murmur immediately after this sound was never heard in Fallot's tetralogy though both these findings are not uncommon in other types of cyanotic congenital heart disease.

Blood pressure. The blood pressure in these cases averaged 106/73, though sometimes it was hard to get an accurate diastolic reading. In nearly every case it was within the range 115–95/80–65 and in 4 cases where it was about 127/90, all were over 15 years of age. In some of the older patients it seems to be increasing a little, still with a small pulse pressure.

Radiology of the Heart

In the account of the first 18 cases (Campbell, 1948), the size and shape of the heart was discussed at some length and the difficulty of describing any characteristic shape was emphasized. Less than half had hearts that were sabot shaped and the other half had more normal shaped hearts; sometimes with a gross hollow pulmonary bay, but often with an almost straight left border or occasionally even with some slight prominence in the region of the pulmonary conus. The findings are much the same in the present series and we are not discussing the question further.

We would, however, emphasize three negative points as of the greatest importance.

The density of the lung. The most decisive—and perhaps the most decisive point in the diagnosis of a condition that can be helped by systemic-pulmonary anastomosis—is the absence of noticeable pulsation in the lung roots, with an absence of density in the lung fields as a whole.

The size of the heart. The second important point is the size of the heart. In most cases of Fallot's tetralogy, however great the disability and cyanosis, the heart is of normal size or even smaller. Anything more than trivial enlargement of the heart makes one hesitant about operation, partly because it suggests that there are greater complications in the congenital abnormality, and partly because the heart is less able to stand any enlargement that may follow the creation of an artificial ductus arteriosus. This is discussed more fully in the next section.

The pulmonary artery. The third point, of almost equal importance, is that there should be no undue prominence of the pulmonary artery and better still, that there should be a striking hollow in the pulmonary region, though as already stated, some patients have a rather straight left border. There may even be a convex projection in the region of the conus just below the origin of the pulmonary artery due to the prominence of the infundibulum distal to the infundibular stenosis. Or there may be a dilatation of the pulmonary artery beyond a pulmonary valvular stenosis but this is rare with Fallot's tetralogy, and should show no pulsation.

It is desirable that one should be able to see both pulmonary branches, because then a pulmonary vessel is available for the anastomosis and there is no risk of the patient dying suddenly from arrest of the pulmonary blood flow when one pulmonary artery is clamped. This accident occurred in Case 17; in Case 27 no operation was possible because the pulmonary artery was too small. Looking back at the X-ray films we think that in the latter, it should have been possible to tell this before operation, but in the former it would not have been easy, as there was relative density round the lung roots, presumably owing to the collateral circulation.

In many cases, the hollow in the region of the pulmonary artery in the P–A view and the large aortic window in the left oblique, together with the absence of pulsation and the absence of density in the lung fields, make the diagnosis easy after radioscopy, and there seems no need for any further investigation. In others the prominence of the pulmonary artery and the density of the lungs, with or without pulsation far out in the lung fields, at once makes it obvious that the patient is not suitable for operation. But there remain others where the decision is difficult; the density produced by the

collateral circulation causes one of the greatest difficulties but here there is no pulsation and more pin-point scattered shadows. Our first mistake in this direction was in Case 28 where we had been doubtful about the prominence of the lung fields but had decided there was pulmonary stenosis. However, at operation the pulmonary artery was found to be large and pulsating with a pressure well over 80 mm. of mercury. This was in a child who was unable to stand and had been screened lying down.

The right (I) oblique position on radioscopy will help in showing if there is any undue prominence or pulsation of the pulmonary artery or of its left branch. The left (II) oblique will define the relative size of the ventricles and will generally show the left of normal size with the right somewhat, but generally not greatly, enlarged. It also helps to show the size of the pulmonary artery and of the aorta.

The aortic arch. A barium swallow is necessary to determine whether the aortic arch is on the left or right. As the barium is swallowed it is often of help to keep ones eyes fixed on the aortic knuckle, as the barium may sometimes follow a preliminary curve to the right before it reaches the aortic knuckle and if this is small it may be missed. It is hardly necessary to say that the barium should be of thick consistency.

The aortic arch was right-sided in 14 and left sided in 36 of the cases which is nearly the usual proportion of 1 in 4.

The Size of the Heart

The heart size is not easy to estimate with accuracy, and general opinion ranks inspection of the film and still better, of the heart on radioscopy, as a better method than any specific measurement.

The following estimates of the heart sizes have been decided by one of us (M.C.) in retrospect, mainly on the P–A films, because these are easiest for comparison from case to case and were always available. Some attention was paid to the cardio-thoracic ratio (c.t.r.) in each but the decision was made mainly on the general appearance. It is, therefore, interesting to see how these estimates compare with the c.t.r. which are more useful for conveying an idea to others. They are given below against the estimated size of the heart; the figures in brackets refers to the number of cases.

Very small..	37–43 (4)
Small	41–49 (10)
Small normal	45–50 (5)
Normal	45–52 (15)
Large normal	49–54 (8)
Enlarged	52–61 (8)

In 14 cases the hearts were regarded as small and in 8 only as enlarged, though possibly some people might have counted the 8 " large normal " as enlarged: the remaining 28 (or 20) were regarded as of usual size.

The 8 that were enlarged will be discussed in more detail, and taking them from the largest downwards they were as follows:

Case 35 (c.t.r. 62) died after operation and there was a single auricle as well as the other features of Fallot's tetralogy.

Case 33 (c.t.r. 61) had tricuspid atresia with a non-functioning right ventricle and although the operation has been most successful and the patient is able to lead a life quite unlike anything previously, the heart has subsequently enlarged more than we like.

Case 28 (c.t.r. 56) had some doubt about the lung fields before operation, and at operation the pulmonary artery was found to be large with a high pressure and the patient probably had Eisenmenger's complex.

Case 11 (c.t.r. 55) was not helped by operation but we thought this was due to technical difficulties with the anastomosis and did not indicate a wrong diagnosis.

The next four fall into a category of slighter enlargement.

Cases 8 *and* 47 (c.t.r. 52 and 53) seemed to be ordinary cases of Fallot's tetralogy and the operation was satisfactory, Case 8 being 25 years old.

Case 48 (c.t.r. 52) was an ordinary case of Fallot's tetralogy who died after operation; he was 27.

Case 32 (c.t.r. 52) had the aorta arising from the right ventricle and he, too, died after operation. It will be seen that among these 8 cases there were several where the diagnosis was more complicated than straightforward Fallot's tetralogy or where the patients did not do well, and the greatest care should be exercised in chosing any patient with significant cardiac enlargement.

There were four other patients where we decided to call the heart large normal, though it was difficult to be sure it was not slightly enlarged; it was interesting that the c.t.r. were much the same as the four just described, where we thought there was enlargement.

Cases 45 and 30 (c.t.r. 54 and 53) seemed to be ordinary examples of Fallot's tetralogy and did well after operation. Case 38 (c.t.r. 53) who died after operation had Fallot's tetralogy and Case 49 (c.t.r. 52·5) had tricuspid atresia and did well after operation. As three of these did well and there is no reason to think that the one death was due to the heart size, it is probable that similar cases can reasonably be included as suitable for operation

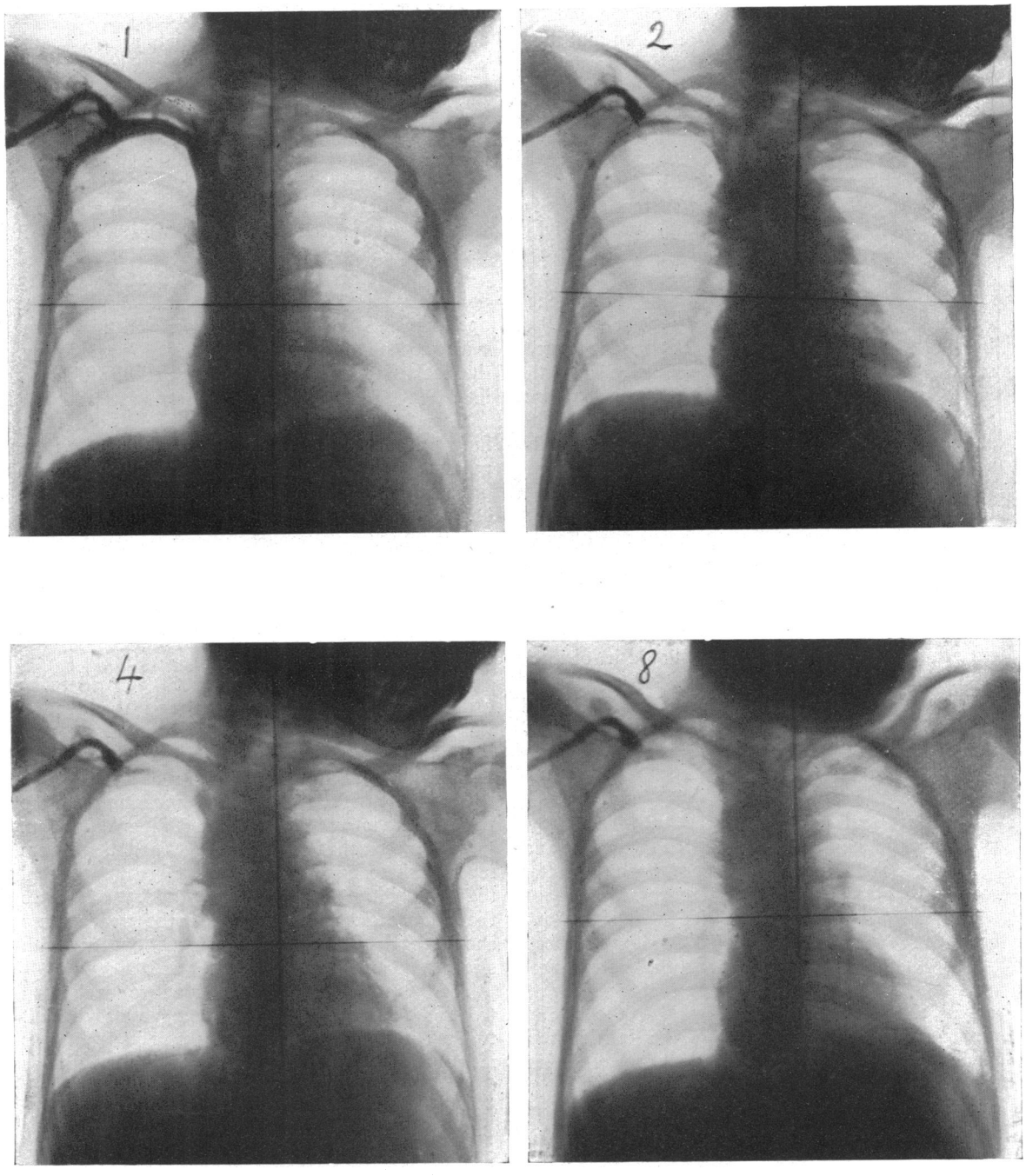


FIG. 4.—Case 39. Fallot's tetralogy. The film at 1 second shows normal filling of the right auricle. The film at 2 seconds shows striking filling of the large aorta arching to the right and some filling of its branches. There is evidence of filling of the left ventricle. There is no significant change in the pulmonary arteries. The film at 4 seconds still shows the aorta but less clearly and the subclavian more clearly. The filling of the pulmonary arteries is trivial but this was the maximum reached. In the film at 8 seconds the shadows are fading though the aorta can still be seen. The pulmonary arteries and the lungs as a whole are lighter than at 4 seconds. The conclusion is a large right to left shunt with a right-sided aortic arch and a fairly severe pulmonary stenosis.

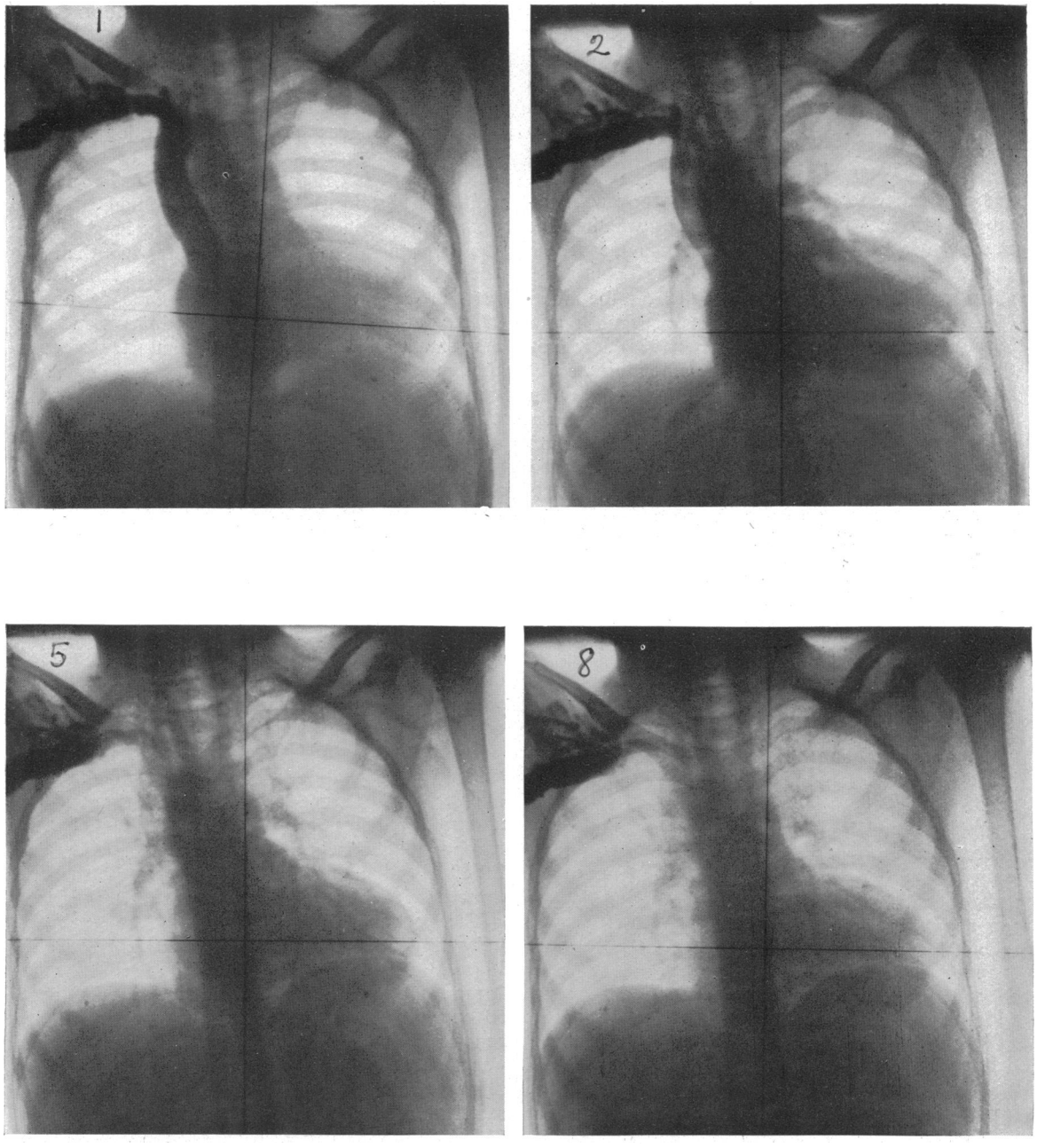


FIG. 5.—Case 47. Fallot's tetralogy. The film at 1 second shows normal filling of the S.V.C. and right auricle. The film at 2 seconds shows dense filling of the right ventricle and of the aorta arching to the right and of its branches as far as the subclavian in the left axilla, indicating an aorta in free communication with the right ventricle; the dye has not passed out to the borders of the left ventricle. At the same time, the pulmonary arteries have begun to fill. The film at 5 seconds has been chosen because it shows as much pulmonary filling as at any time and obviously little blood has entered the lungs. The degree of filling is a far better indication of pulmonary stenosis than the time at which dye can first be seen in the pulmonary arteries. The right ventricle and aorta are clearing. The film at 8 seconds shows that the heart as a whole is becoming clearer. The conclusion is a large right to left shunt with a moderate degree of pulmonary stenosis.

and that if the heart can be passed as not much enlarged a relatively high c.t.r. is not a reason for deciding against operation.

We have no example to report of a successful operation in a patient with Fallot's tetralogy with the c.t.r. over 55. But this may be partly due to selection of patients who had not the larger hearts, and this is a question that still needs more experience. We are glad we did not exclude the boy with tricuspid atresia with c.t.r. of 61 but whether his improvement can last as long as in some of the others remains to be seen.

From our present experience and from the tables of Lincoln and Spillman (1928), of Maresh and Washburn (1938), and of Caffey (1945) it seems that under the age of 2 years the c.t.r. averages 49 and varies between 40 and 65 (60 after one year) in normal children. These figures do not concern our present purpose directly but may help in advising parents whether their children may possibly be helped by operation later.

From 2 to 5 years of age the normal range lies between 43 and 52, and from 6 years onwards the average c.t.r. falls slightly from 47 to 45, with a range from about 40 to 50. The original figures of Danzer as long ago as 1919 gave 39 to 50 as the adult range with an average of 45; he stated that a c.t.r. of 52 might be normal if the heart did not look enlarged, but that one of 53 was pathological. These last figures were of course concerned with adults but seem to agree with our conclusions.

On our present experience, we consider that a heart which seems a little enlarged with the c.t.r. of 52 to 54 should certainly not contraindicate operation, though a larger heart than this often indicates the presence of some complication and is probably a bar to *lasting* improvement. We have not yet sufficient evidence to say how often operation should be advised in these larger hearts for the sake of immediate advantages.

Angiocardiography

We do not propose to discuss in detail the help that can be obtained from angiocardiography which was only available in nine of the later cases of this series. As a rule, there should be no need for this help from the point of view of diagnosis, but in border line cases where a heavy collateral circulation hides the diminished blood supply to the lungs, it may be of the greatest value; and it may help to establish the diagnosis in complicated cases where no diagnosis can be made on clinical grounds alone. Apart from this, it has proved of increasing value from the surgical point of view in delineating the anatomical arrangement of the arterial branches from the aorta and of the size and position of the pulmonary arteries.

As Fig. 4 and 5 show, there is no difficulty in demonstrating the shunt from the over-riding aorta and this is generally well seen in the film taken at the 2nd second in cases of Fallot's tetralogy. Often the pulmonary arteries start filling at the same time, which might suggest there was no great degree of pulmonary stenosis but we have found that the amount of the increased density of the lungs during the subsequent 5 seconds is a better test than the speed with which the opaque substance can first be seen in the pulmonary arteries. Even so, angiocardiography seems to give an added precision to the assessment of the degree of pulmonary stenosis present in different cases of Fallot's tetralogy, and may help in distinguishing between valvular and infundibular stenosis. Both these points may ultimately be useful in deciding the sort of operation that is most likely to be successful.

Electrocardiograms

The two most striking features of the cardiogram are the large pointed P wave especially in lead II and the right ventricular preponderance. This is of such a degree that we think the term ventricular preponderance rather than axis deviation is justified even on the standard leads. We hope to deal later with the value of unipolar chest leads as these were not available for all the early cases.

Wood and Selzer (1939) thought that a tall spiked P wave might be produced by a right auricular hypertrophy. Pardee (1941) accepts the view that abnormally high pointed P waves occur with hypertrophy of the right auricle, while notching and broadening are seen with hypertrophy of the left auricle.

The prominent pointed P wave was generally tallest in lead II and some examples are given in Fig. 6. The width of the P waves was generally 2 mm. and exceptionally up to 3 mm. In these 50 cases P II varied from 2 to 8 mm. in height, generally between 3 to 7 mm. Once (Case 30) it was small and sharply inverted. There was only one other where P II was as small as 2 mm. high, 10 cases where it was 3 mm. high, 16 where it was 4 mm. 11 where it was 5 mm., 6 where it was 6 mm., 4 where it was 7 mm. and 1 where it was 8 mm. high. The average was 4·5 mm. Chamberlain and Hay (1939) give 1·5 mm. as the average size of P II in the first decade with a maximum of 3·0 mm. Pardee (1941) gives 2 to 5 mm. as the usual size for the large P waves of mitral stenosis.

The cause for these large P waves is not certain. It is not due to the tachycardia that is generally

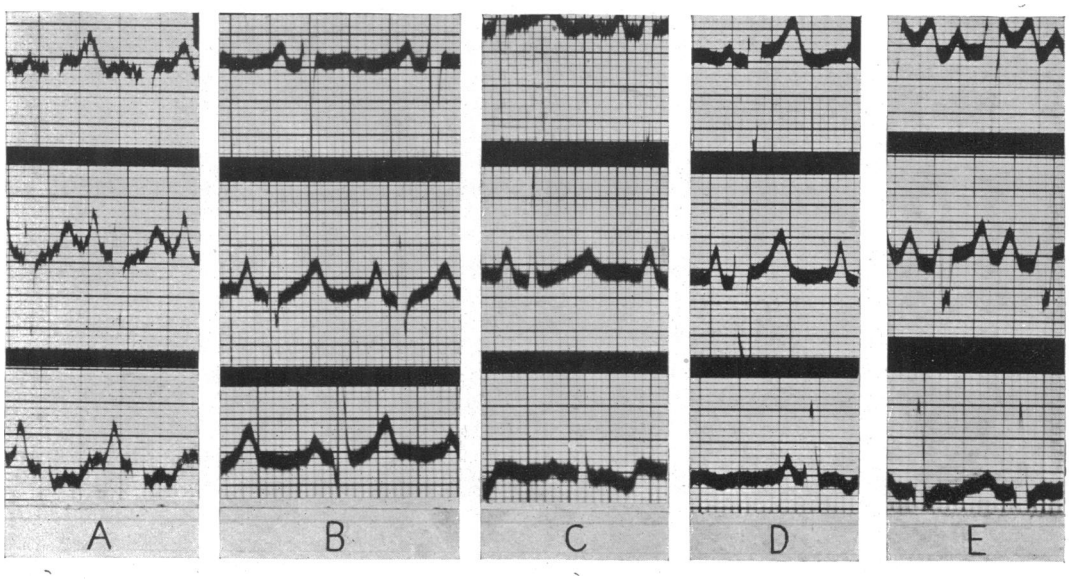


FIG. 6.—Typical electrocardiograms from 5 cases of Fallot's tetralogy showing gross right axis deviation and large pointed P waves. (A) from Case 0208 confirmed post-mortem. (B)–(E) from Cases P067, H120, P022, and P035 confirmed by a successful result of operation for a systemic pulmonary anastomosis. (Reduced to five-sixths.)

present, especially in the younger patients, as equally tall P waves occur in the same subject when the rate is slower. The variability suggests that it is partly due to strain on the right auricle as well as the hypertrophy which should make for greater constancy. Moderately large and pointed P waves may occur with cardiac infarction or in cor pulmonale where there is right heart strain. Large P waves were seen equally in the cases of tricuspid atresia.

In Case 35 where there was a single auricle the P wave was large and pointed, 5 mm. high. In Case 49 with tricuspid atresia it was tall and bifid as well. In the other two cases with tricuspid atresia the P wave was sometimes large and broad in Case 42, but in Case 33 it was not remarkable. Occasionally the P waves may show a fair amount of variation in height in the same record (e.g. Case 35). Where more than one cardiogram was taken before operation the P waves often differed in amplitude but never to any great extent except in Case 36; although there were large pointed P waves in her first cardiogram with a rate of 115, a subsequent one done six months later, but before operation, showed normal small P waves, with a rate of 75; there were no other significant changes in the two curves.

The most significant finding as one might expect was the high degree of right ventricular preponderance. The main exceptions were the 3 cases with left ventricular preponderance; it is fairly certain that such a case will turn out to be tricuspid atresia or stenosis with a non-functioning right ventricle, especially if on radioscopy the left is the larger of the two ventricles.

In the 47 cases that were thought to have Fallot's tetralogy, the general finding was an absent Q, a diminutive or very small R and a large S in lead I; and an absent or very small Q, a large R and most constantly of all, an absence of S in lead III. Out of these 47 there were only 4 with S III and in these it ranged from 3 to 8 mm. in depth. The average figures for the size showed that R I, Q III, and S III were small, 2·8, 1·7, and 0·4 mm. respectively, while S I and R III were large, 12·9 and 13·3 mm. respectively.

The QRS complexes in lead II may be of the R type (23 cases) or S type (15 cases) or mixed (9 cases) or with very small QRS complexes (4 cases).

Now and again one meets with a case that does not show a high degree of right preponderance. Case 6 is an example where there was no preponderance possibly indicating a single ventricle as well as the usual features of Fallot's tetralogy; the result of operation was very successful but the colour was not improved as much as in most cases. In Cases 1, 30, and 37, all successful operations, there was less right preponderance than usual. Some cases showed a moderate R I in addition, of course, to the very deep S I.

T wave abnormalities were uncommon except for T III inversion: in 11 cases it was sharply inverted and in 7 it was flat or slightly inverted. There were no T II inversion except in Case 30 where it was biphasic.

In two, T I was inverted (Cases 42 and 47) and in one it was flat (Case 17). In Case 42 for some unknown reason it became normal and upright again six months later, before his operation. In Case 21, T I was enormous, 11 mm. in amplitude, and T III was deeply inverted. Fairly large T waves in lead II with an amplitude of over 5 mm. occurred in 8 cases.

As with the P waves, the T waves in some of these cases showed variation in height, unlike most other types of heart disease where the size and shape remain remarkably constant. The S–T interval was often elevated in lead I and sometimes became more normal after operation.

Operative Experiences

In these first 50 cases the original basic technique, as recommended by Blalock, has been closely followed; in 6 other cases valvulotomy was performed or attempted. Pott's modification (anastomosis of aorta to pulmonary artery) was not used in this series, although it has been since.

The Blalock operation of anastomosis of a systemic artery to a pulmonary artery is always difficult and exacting technically, even when the anatomical conditions are favourable and the anastomosis proceeds smoothly. At times, when conditions are not favourable, it becomes an operation of really great difficulty and calls for all the surgeon's technical ability. Some of the earlier more difficult cases were especially exacting, and even with increased experience it is found that a series of straight-forward operations is suddenly interrupted by a complex anatomical situation calling for great patience and not a little endurance.

This is not an operation for the casual operator and indeed, quite apart from the pressure of the waiting list, it is desirable that the surgeon should operate regularly. In Blalock's clinic one operation a day is aimed at; it has not been possible to achieve this here because routine thoracic work must still be done, but a desirable standard should be two or three operations a week. Success is not possible without good team work and one must stress in particular the invaluable part played by the anæsthetists; Rink, Helliwell, and Hutton (1948) have already written a preliminary record of their experiences.

Often, these cyanotic patients are not in the state of general health that one would ordinarily demand before embarking upon a severe operation; they may run a low fever, even with sharp rises, or have recurrent or persistent minor upper respiratory infections, etc. While it is clearly folly to operate in the presence of considerable pyrexia or recent increase of illness, we have learnt that it is often better not to wait for seemingly ideal conditions, which may in fact never materialize. It is sometimes best to seize the opportunity offered, for conditions may deteriorate rather than improve.

Moreover, once the anæsthetic or operation has been begun it is desirable, however unfavourable the outlook may appear, to press steadily on until it becomes quite clear that success is impossible. This applies not only to anatomical difficulties but to physiological ones as well. On several occasions the anæsthetist has reported the patient's condition as very grave and the temptation to abandon the operation has been great. In almost all of these cases continuance with the operation after a short wait has been rewarded by final success. The most striking example of the soundness of this policy came in the case of a small child (Case 41) aged 5 years, who was extremely ill; she was deeply cyanosed, with severe recurrent pain in the chest even when resting in bed, and was incapable of any activity at all. During induction of anæsthesia a bronchial spasm developed and she stopped breathing, and it was only with the greatest difficulty that the anæsthetist could inflate her lungs with an intratracheal tube in place; her heart then also stopped. Whether she was then dead is a nice point. Intracardiac injection of adrenalin was given and artificial respiration continued. The heart started again but spontaneous respiration did not begin for another 40 minutes; her condition was, of course, still desperate. At the end of an hour her condition had begun to improve and after much deliberation the operation was started, for it was certain that no second attempt could be made and the outlook was otherwise hopeless. The operation was completed and the child made an excellent recovery; ten days later she was learning to walk about the ward, a thing she had not been able to do before. She has continued to do well.

All our greatest hazards have appeared in the operating theatre; in this series the deaths occurred either in the theatre or within a few hours of return to the ward. If the patient left the theatre in even fair condition, recovery always followed. There was only one instance of bleeding from the anastomosis, that of a man aged 27, very disabled, very blue, who had already had a hemiplegia; he died several hours later from hæmorrhage from the anastomosis which had been quite dry when the chest was closed (Case 48). Bleeding may, of course, occur temporarily when the clamps are

first undone but it either stops spontaneously or has been controlled by insertion of fresh sutures. In one or two of the earlier cases this re-suturing led to narrowing and impairment of the efficiency of the anastomosis and was responsible for some of the poor results.

It has been our experience that patients over 20 years of age carry a far greater operative risk; not only are they commonly severely disabled, but their heart muscle seems to have suffered from the long-continued strain, in contrast to the younger children in whom the myocardium seems surprisingly good. In addition the anatomical hazards may be greater, and the fatal hæmorrhage in this last case was certainly due to the very thin-walled pulmonary artery, aided by a certain degree of extra strain on the anastomosis during systole as the subclavian artery curved down over the prominent aortic arch.

Surgical Problems

Certain general technical features of the operation need discussion in the light of our experiences, for most of these features introduce important practical problems that concern, or should concern, the surgeon undertaking this work.

(*a*) *Blalock's operation or the Potts' modification.* Although the Potts-Smith modification of aortic-pulmonary anastomosis was not used in this series, it has been employed since and has attracted sufficient attention to make it desirable to discuss its advantages and disadvantages. It should be realized that it does not introduce a new principle, but is a technical modification based upon the original principles laid down by Blalock and Taussig. They postulated that certain cases of cyanotic congenital heart disease in whom pulmonary stenosis existed could be improved by anastomosing a systemic artery to a pulmonary artery in order to increase the flow of blood to the lungs, and in their preliminary discussions mentioned the use of the aorta as a possibility in place of one of its branches. It remained for Potts and his colleagues to introduce the ingenious clamp that made this possible.

The advantages of using the aorta would appear to be as follows.

(1) The operation may be easier and quicker.

(2) It is especially useful in small children in whom the subclavian may be too small to furnish an adequate additional blood-flow to the lungs.

(3) It allows the size of the stoma to be varied at will and to be measured exactly.

(4) It may provide a ready solution to the problem of the case with a difficult, deep, short, and narrow subclavian artery.

(5) It avoids the dangers of cerebral damage associated with the use of the carotid or innominate arteries.

(6) It is the simplest, and sometimes the only, way of overcoming the problem of a very high aortic arch with its branches arising at the very root of the neck (Fig. 7).

The disadvantages of using the aorta would seem to be as follows.

(1) It demands a postero-lateral approach (the operation is probably possible, but certainly very awkward through an anterior incision).

(2) It may be very difficult if the aorta is right-sided, as occurs in about one quarter of the cases. This is because the right pulmonary artery is often very short and deeply placed in the mediastinum.

(3) The pulmonary artery may be so small and narrow that it cannot be used, whereas a small thin artery can still sometimes be employed efficiently for end-to-end anastomosis with the subclavian.

(4) In older patients there may be too much disproportion between the size and thickness of the walls of the pulmonary artery and that of the aorta to make a safe junction.

(5) Direct aortic-pulmonary anastomosis may cause a greater strain on the heart and a greater risk of pulmonary œdema.

It is still too early to make a final assessment of the Potts' modification, but the most certain advantages it offers would appear to be a more rapid and less elaborate dissection when the aortic branches are small and deeply placed in the mediastinum, or arise very high in the chest; the possibility of adjusting the size of the stoma to meet the needs of the individual case, especially in very small children; and freedom from the dangers of cerebral damage following use of the carotid or innominate arteries.

In cases where it would be about as easy and satisfactory to use either the subclavian artery or the aorta, it would seem to be surgically sounder and wiser to use the subclavian and to avoid exposing the patient to the greater perils that must attend the deliberate manipulation of a structure of such importance as the aorta. After all, if some mishap befalls the subclavian artery the situation can readily be remedied by ligating it and the aorta could then be used. If some mishap occurs when the aorta is being used it would certainly be more difficult, and perhaps impossible, to retrieve the situation.

(*b*) *Antero-lateral or postero-lateral approach.* Blalock's practice of using the antero-lateral intercostal incision was followed almost exclusively in the first 50 cases, but has often been departed from since then. The antero-lateral thoracotomy incision

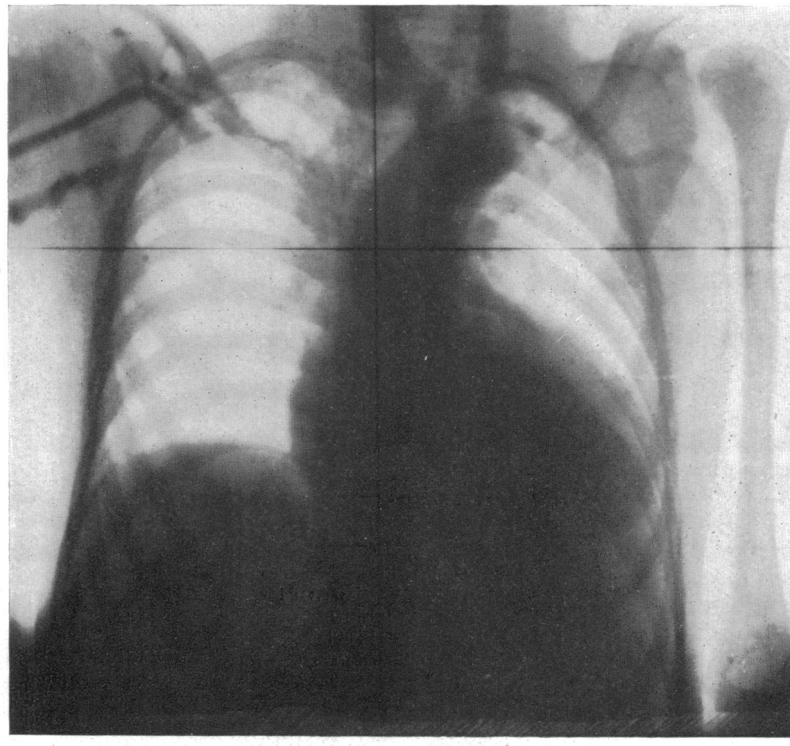


FIG. 7.—Case PO35. Angiocardiogram at 3 seconds. This shows a left-sided aorta which is dilated and very high, with the innominate rising in the ridge of the neck to the right where it had been easily felt in the neck. The illustration also demonstrates the large right to left shunt with pulmonary stenosis.

has always been more popular in American surgery than in that of other countries, as witness the popularity enjoyed by the anterior approach for pneumonectomy and for ligation of the patent ductus arteriosus. In a very ill patient the antero-lateral approach may throw a smaller strain on the lungs and circulation than a postero-lateral one. In general, however, the exposure provided by the antero-lateral intercostal incision may be very cramped, and if the ribs are awkwardly shaped so as to make a high narrow chest, the difficulties of dissection and ligation of the highest branches of the subclavian artery may be extreme. In such cases the faulty exposure commits one to a set of circumstances that really constitute faulty surgery.

One important reason that determined the use of the antero-lateral approach in many cases in this series was our policy of opening the pericardium and examining the heart condition carefully to make as complete a diagnosis as possible and to be prepared to perform a valvulotomy if indicated. The pericardium can, of course, be opened and the heart inspected through a postero-lateral incision, but it would be difficult to utilize the right ventricular approach to the pulmonary valve without considerable and most undesirable dislocation of the heart.

Our present practice is to use the antero-lateral approach only in those cases in which it may be necessary to operate directly upon the right ventricle itself. For all other cases a postero-lateral incision is used with resection of the whole length of the fourth rib from transverse process to costal cartilage. The resection of the rib is much more satisfactory than an intercostal incision which causes more bleeding at the time and may cause dangerous or even fatal oozing afterwards; the risk of bleeding is increased by the use of pericostal sutures, which are also a cause of unnecessarily severe post-operative pain. The postero-lateral thoracotomy with resection of a rib is followed by far less pain than either a postero-lateral or antero-lateral intercostal thoracotomy. The exposure afforded by the long postero-lateral approach is a great advantage and allows a much more easy, rapid, and safer exposure of the vessels and inspires the surgeon with much greater confidence that he has

more complete control of the situation. There is no difficulty in application of the pulmonary artery clamp provided the incision is carried well forwards; the performance of the actual anastomosis is easier than from the front. Moreover, the postero-lateral incision allows use of either the subclavian artery (Blalock's operation) or the aorta (Potts' modification), according to the conditions found.

With but few exceptions, in which the third intercostal space was used, all the operations by the antero-lateral route have been done through the second interspace which is definitely preferable, except in very small children.

(*c*) *Right or left side.* Blalock's earlier recommendation was to use the side opposite to the aortic arch in all but adults and patients in the later teens. His reasons for this were that if the subclavian is used as it arises from the innominate it forms a more satisfactory angle with the parent vessel when it is turned down for the anastomosis; whereas if the subclavian is used as it comes off the aorta it may be sharply kinked at its origin, or flattened as it passes over the prominence of the aortic arch. Also, if the subclavian artery is found to be unsuitable the innominate or the carotid artery can be used instead.

In his latest paper Blalock (1948*b*) states that he uses the right side (when the aorta is left-sided) in all patients between the ages of 2 and 12, but prefers the left approach in children below 2 and in patients over 12 who have attained most of their growth or who are more than 5 feet in height. He mentions that some other surgeons have preferred to use the left-sided approach for all cases.

Blalock's earlier recommendation was followed in most cases in this series, but, recently, departures have been made. In spite of increasing familiarity and experience with the operation the dissection of the systemic arteries on the right side may be extremely difficult, not a little dangerous, and certainly very exacting; especially so when the superior vena cava is large and dilated. It was therefore decided to extend the use of the left-sided approach to children under 12 years as well as to older patients and this was done successfully in a number of cases and with much greater ease; even though the subclavian may appear unduly kinked and flattened at the time the result has been just as good, and it seems probable that the artery elongates and adapts itself. The right-sided approach was then used again on a small child aged 5 years (Case P043) and after a long, tedious, and exacting dissection a very deeply placed and long innominate artery was found which divided high up near the superior thoracic inlet and gave rise to a subclavian artery too short to bring down to meet the right pulmonary artery. This could have been done if the carotid artery had been ligated and divided, but in addition to the carotid another artery almost as large passed into the neck and it seemed that the two vessels must carry a large supply of blood to the head and brain. Alternatively the innominate could have been used, but in addition to the dangers of cerebral ischæmia this artery was so large that there seemed considerable danger of causing heart failure and acute pulmonary œdema if it were used. Accordingly the operation was abandoned with the idea of using a left-sided approach on a later occasion. This experience has finally decided us in favour of using the left-sided approach in all cases unless angiocardiograms suggest the right pulmonary artery is small or absent, indicating it would be dangerous to occlude the left branch while the anastomosis is made. The only other indication for a right-sided approach is the presence of a right aortic arch in a case on which one wishes to do the Potts' operation.

(*d*) *The use of the carotid or innominate arteries.* The danger of cerebral ischæmia is very real if the innominate or carotid arteries are used; in Blalock's series the mortality was 30 per cent. The carotid was divided to allow the innominate to be turned down in two patients in this series (Cases 24 and 35) and in one since (Case P034). The first did extremely well; the second developed an acute pulmonary œdema as soon as clamps were removed and the anastomosis allowed to function, and died after a few hours; the third became comatose and hemiplegic soon after operation and died the next day. Blalock (1948*b*) in his last paper emphasizes that the carotid or innominate should not be used if it can be avoided and states that in many of his earlier cases in which one of them was used, a little longer careful and patient dissection of the subclavian might have spared the carotid or innominate. It would appear to be purely a matter of chance whether interruption of the carotid circulation is followed by paralysis or death or by a good result. It is indeed a gamble, and a poor gamble as well, and therefore is neither surgically nor morally sound. In our opinion, the use of the carotid or innominate arteries is unjustifiable and should be abandoned.

If the surgeon is contemplating using the right-sided approach he should first of all study good angiocardiograms which display the disposition of the aortic arch and its branches. In this way he should be able to assess whether or not it is likely to be possible to use the right subclavian artery. This is a far better way in which to obtain the information than a thoracotomy. An unfavourable

arrangement of the great vessels, such as would make sacrifice of the carotid blood-supply inevitable, is seen in Fig. 7, 8, and 9. In Fig. 7 the aortic arch is unusually high and the innominate artery actually lies above the clavicle and could be seen and felt in the neck; clearly Blalock's operation would be impossible on either side without using a long length of carotid artery from the neck. Such an arrangement seems to demand Potts' operation, which was successfully used in this case. In Fig. 8 and 9 the position of the innominate artery, deep in the mediastinum, is clearly seen and also its very high division with a resultant very short subclavian artery. In these cases the left subclavian artery was successfully used for the anastomosis.

(*e*) *Absent or small pulmonary arteries.* The value of angiocardiography to display the disposition of the aorta and its branches is clearly proven in these cases and it has been of equal value in others. It has been less satisfactory in displaying the main pulmonary artery and its right and left branches, often because of the slow and feeble concentration of the opaque solution in them due to pulmonary stenosis, and especially when a large and rapid shunt has caused the contrast medium to pass rapidly into the systemic circulation. In the normal patient it may be easier to display the chief pulmonary arteries more clearly, especially when oblique or lateral views are used in addition to the postero-anterior ones. One must be prepared to be disappointed with the delineation provided when pulmonary stenosis exists with a large shunt.

The angiocardiograms may, however, indicate that one pulmonary artery is either entirely absent or very small. Fig. 10 shows an example in which the left pulmonary supply seems much smaller than the right. This suggests that it would be unsafe to use the right pulmonary artery as death would probably soon follow its necessary occlusion while the anastomosis is being made. The demonstration or suspicion of a small right pulmonary artery would provide an indication for using a right-sided

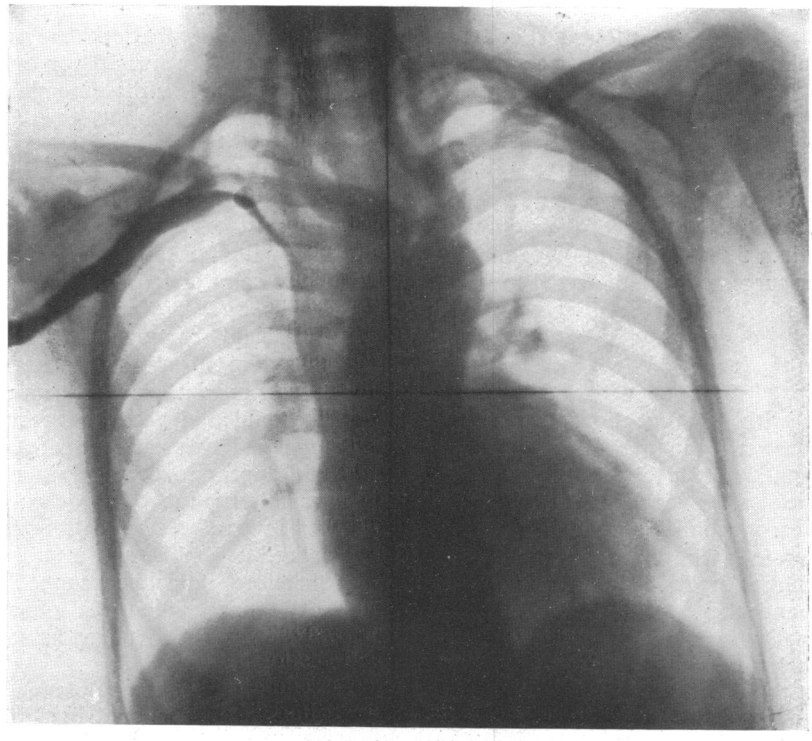


FIG. 8.—Case C017. Angiocardiogram at 3 seconds, showing the origin of the innominate artery fairly deep in the mediastinum with a high division of the subclavian from the innominate and a large gap that would have had to be bridged between this and the pulmonary artery. It also shows an over-riding aorta, a high grade of pulmonary stenosis, and a dilated superior vena cava.

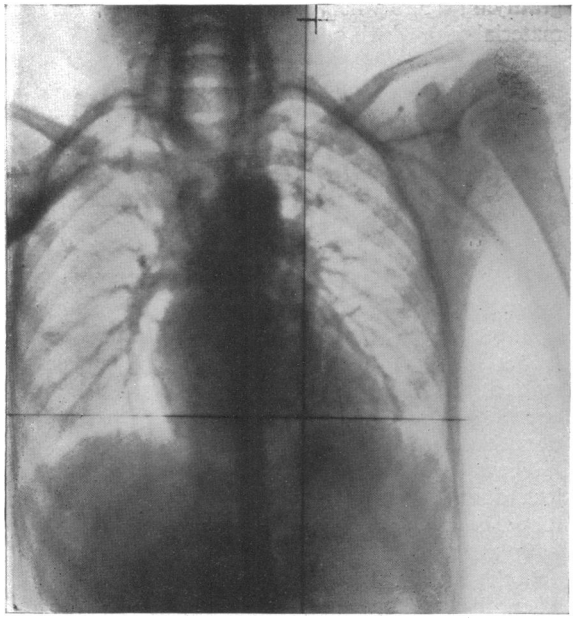


FIG. 9.—Case 0053. Angiocardiogram at 3 seconds. It shows the origin of the innominate artery fairly deep in the mediastinum with a high division and a resultant short subclavian artery that would have been difficult for a pulmonary anastomosis. It also shows an over-riding aorta with moderate pulmonary stenosis.

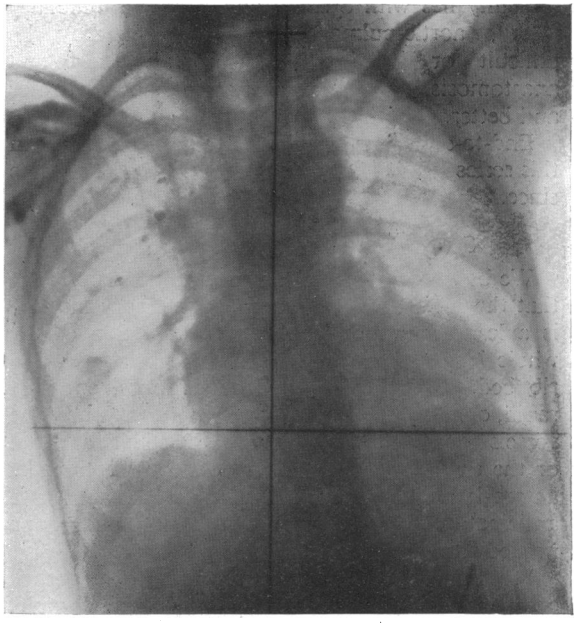


FIG. 10.—Case P048. Angiocardiogram at 4 seconds which suggests that the left pulmonary artery provides a much smaller blood supply than the right and that it would probably be unsafe to occlude the right branch. Operation will, therefore, be performed on the left side.

approach in preference to a left-sided one. Careful screening and plain radiography may also enable one to identify both pulmonary arteries or to suspect that one is small or absent.

In one case, after a long dissection, the right pulmonary artery was found to be not much larger than an intercostal artery and so could not be used; no ill-effects followed the exploration (Case 27). In another (Case 17) the right pulmonary artery was prepared and had been clamped for 12 minutes when the heart stopped; it was assumed that this was due to absence or obliteration of the left pulmonary artery. The heart was started again after massage and injection of adrenalin, and end-to-end anastomosis was performed between the subclavian artery and the first branch to the right upper lobe. Unfortunately the heart stopped several times and finally could not be started again; autopsy confirmed that the left pulmonary artery was completely obliterated. Such cases sometimes are unavoidable and part of the hazards of the procedure, but may be avoided by more careful radioscopy of the pulmonary arteries.

(*f*) *End-to-end anastomosis.* In order to secure extra length of vessels Blalock not infrequently ligates the pulmonary artery medially, divides it, and performs end-to-end anastomosis to the subclavian. This can be a most valuable step in some of the more difficult cases and may indeed be the only possible way to bridge a gap when the subclavian is short. It is especially useful when, on the left side, the prominence of the aortic arch threatens to kink and flatten the down-turned subclavian artery and to cause tension on the anastomosis. It should certainly be used in preference to end-to-side anastomosis to a narrow pulmonary artery in which most of the width of the pulmonary artery would be encroached upon by the anastomosis. One is naturally reluctant to take the step of ligating and dividing the pulmonary artery but, as Blalock says, a good end-to-end anastomosis is always preferable to an uncertain end-to-side one. The only difficulty may arise from considerable disproportion between the two vessels; Blalock states that he does not mind this provided the pulmonary artery is no more than two to three times the size of the subclavian.

Potts' modification may provide an easy alternative if the subclavian artery is too short to use without end-to-end anastomosis, but if the main

difficulty lies with a small and narrow pulmonary artery, aortic-pulmonary anastomosis may be difficult or impossible and direct end-to-end anastomosis with the subclavian is much the safer and better.

End-to-end anastomosis has been used twice in this series (Cases 8 and 17) and has been used twice since.

Summary of Surgical Procedure

We believe, in the present state of our experience, that the most useful and most satisfactory incision is a left postero-lateral one with resection of the whole length of the fourth rib and incision of the rib bed. This gives a perfect exposure, enabling rapid, comfortable, and much safer dissection of the vessels and also permits use of either the subclavian or the aorta for the anastomosis. We reserve the antero-lateral approach for those cases in which we anticipate that right ventricular cardiotomy may be needed for valvulotomy.

We have abandoned the right-sided approach except for cases in which radioscopy and radiography (including angiocardiography) suggest the right pulmonary artery is unduly small or absent. A right-sided postero-lateral incision is needed if Potts' operation is contemplated in the presence of a right aortic arch.

In general we use Blalock's operation in preference to Potts' modification when it is feasible. The greatest value of Potts' operation is in small children in whom the subclavian artery is too small to furnish an adequate extra flow of blood to the lungs, and in older patients when the subclavian artery is too small or too short and it would otherwise be necessary to use the carotid or innominate arteries. We feel the use of either of these vessels is unjustified, owing to the much higher mortality from cerebral complications or cardiac failure and to the risk of permanent residual paralysis. End-to-end anastomosis of the pulmonary artery and the subclavian artery has a useful place when an end-to-side anastomosis would be difficult, impossible, or under undue tension.

Angiocardiography is an invaluable method to allow pre-operative study and assessment of the pulmonary and systemic arterial pattern.

Much emphasis has been laid on the difficulties and anxieties of these operations, but this gloomy side is relieved by the more satisfactory side of success. In spite of the hazards and long hours of work one derives great satisfaction from contemplation of the successfully completed anastomosis and the rapid, indeed at times dramatic, improvement that follows the operation. As stated elsewhere an excellent result was obtained in 66 per cent; a most gratifying figure when one considers the very poor material with which one is working.

Post-operative Treatment

In our experience the post-operative period is much less stormy and anxious than might be expected. Children particularly tolerate the extensive and prolonged thoracic exploration remarkably well, and the difficulties encountered have been mainly in the older subjects. Penicillin therapy is started 24 hours before operation, and pre-operative instruction in breathing exercises is a routine. Cyanosed and polycythæmic patients should never be left for long periods without fluids and particular care should be given to the fluid intake before operation as a simple precaution against the additional danger of thrombosis at this time.

These patients are well fortified against blood loss during the operation so that transfusion of blood is not needed during or after the operation, but plasma or gum saline are given to combat shock, and the drip is continued on return to the ward. They are encouraged to drink as soon as they recover consciousness. The need for intravenous fluids seldom continues longer than 24 hours, unless the blood pressure fails to rise—it is usually back to the pre-operative level or above in 12 hours—or unless cerebral thrombosis is a complication. An adequate fluid intake of at least 1000 ml. in a small child to 2000 ml. in an adult is needed to prevent thrombosis in the days after operation, and is best given by mouth. With increasing experience we have found that the amount that has had to be given intravenously has decreased; in the first 15 cases it averaged 37 ounces while in subsequent cases it has dropped to an average of 25 ounces. Blood transfusion was needed only in two cases, in one (Case 5) who bled into the pleura and in one (Case 8) who had profound postoperative shock.

The patient is nursed in an oxygen tent on return to the ward and the time that this is needed is judged by tentative periods of removal without development of cyanosis or distress. The time varied from six hours to four days, with an average of 36 hours. Any delay in regaining consciousness or inability to move a limb must be noted as an indication of cerebral thrombosis. Breathing exercises and postural coughing are started as soon as the patient is conscious and it must be emphasized that though this may seem unkind, almost brutal, after a serious operation, the children are not unduly disturbed, and insistence on this early stimulation is amply repaid by the rarity of serious chest complications. A portable radiograph is taken within the first 24 hours and at frequent intervals subsequently, for evidence of collapse and particularly for pleural

effusion. Our experience is that morphine should be used with great care; it is not always needed in the first day, and seldom afterwards. In those with distress from coughing, and this is not common, codeine is useful.

There is an immediate rise of temperature after operation but very seldom above 101°, and excluding four cases with complications where it was prolonged over ten days, the average duration was five days. The pulse rate was the better indication of the general condition: it was frequently at its highest in the second 24 hours, and had usually settled to a steady level of 10 points above the pre-operative level by the end of the first week. The close correlation between temperature and pulse rate, particularly the latter, and the presence of a pleural effusion is mentioned later.

Two indications of the degree of success of the anastomosis in the period immediately after operation are the colour, and the presence of a murmur. There is an immediate improvement in colour at operation as soon as the anastomotic circulation starts, but at this time controlled respiration is occurring. In a successful case this improvement in colour is maintained in the oxygen tent, and after a short period of up to 48 hours will continue outside it. Some cyanosis must be expected from the right to left shunt which remains, and this will be increased if breathing is embarrassed by obstructive secretions in the respiratory tract or by pleural effusions. If these factors are taken into account, a comparison of the colour after operation, particularly of the extremities, with the depth of cyanosis before operation is a reliable guide to the degree of functional improvement that may be expected.

The thrill over the anastomosis, which can be felt by the surgeon, is followed after operation by a murmur heard with the stethoscope; this is usually continuous as heard over a patent ductus arteriosus but may be systolic and diastolic or merely systolic. Though chest complications may make recognition more difficult in the first few days, its presence is an encouraging sign of success and its absence suggests that thrombosis has occurred at the suture line and that the anastomosis is not patent. We have become increasingly impressed with the murmur as a sign that a good result may be expected when the patient is able to start walking, which is usually after the first week.

There are three important post-operative complications: cardiac failure, which is rare; pleural effusion, which is common but generally harmless; and cerebral thrombosis, which is the gravest.

One of the criteria for operation is a heart that can adjust itself to the altered circulation which the anastomosis causes, so that much enlargement of the heart is a contraindication. In this respect the selection of cases would appear to have been satisfactory for there have been few difficulties or anxieties on account of the heart. The blood pressure rises to the pre-operative level, or above, in the first 12 to 24 hours, and any delay in this rise favours the development of thrombosis and calls for the exhibition of a "pressor" drug, such as methedrine, and plasma transfusion. This happened in one of our early cases (Case 8), an adult, where the pressure fell to 50/40 and the systolic was below 100 for the first 36 hours; he developed a cerebellar thrombosis in the first 24 hours. A rise well above the pre-operative level is mentioned by Taussig but we have not experienced it; should it occur, a venesection would be indicated.

It might be expected that the sudden increase in the pulmonary circulation, with the additional work demanded of the left ventricle by the anastomotic shunt induced by the operation, might cause pulmonary œdema in the early post-operative period. We have not experienced this in any case surviving operation though it occurred in one death after operation (Case 35) in one lung on the side where the innominate had been joined to the pulmonary artery. Case 21, a poor result as judged by colour and the absence of a murmur over the site of anastomosis, was slow to recover from this operation, and, despite the absence of a pleural effusion, his temperature failed to settle till the 15th day. On the 27th day he complained of pain over the heart and in the left arm, and was collapsed with ashen cyanosis and dyspnœa; the pulse rose to 140 and the blood pressure dropped to 65 systolic. He recovered with morphia but the attack was repeated next day, and again responded to the same drug. There was no change in the electrocardiograph to suggest a coronary thrombosis. There seems little doubt that these were attacks of left ventricular failure and they were treated as such; he subsequently developed a cerebral abscess which was successfully treated by operation. There were no cardiac incidents in the other patients. Congestive failure was never seen, nor was digitalis used, despite the increased work demanded by the anastomosis, as shown by the increase of heart size after operation in most cases.

The second complication of pleural effusion on the side of the operation is common and occurred in 32 of 43 cases after operation; in 11 it was of moderate size but in 21 it was large enough to demand aspiration, and in 10 of these, more than once. The fluid commonly developed immediately after operation, or in the first few days and naturally it is usually blood-stained. An average example

is shown in Fig. 11. But effusion may suddenly increase, recur, or even first develop later in convalescence and this happened in 5 cases. An early effusion successfully aspirated is on the whole less of a handicap in convalescence than the slowly developing effusion where aspiration is hardly necessary in the first few days and the decision is delayed. It is in these cases that later and repeated tappings are necessary, often difficult, and incomplete in their results. It is these patients particularly whose stay in hospital is prolonged, whose temperature and pulse fail to settle, and 5 such cases were discharged with "pleural thickening" on radiography or showed a very small residual effusion: all cleared up subsequently.

The temperature and pulse chart reflect remarkably clearly the presence of fluid in the chest; an early effusion successfully aspirated is associated with a quick return to normal temperature and pulse rate: a rise on the second and third day, which is not uncommon, almost denotes a slowly developing effusion: a sustained temperature and pulse indicates the persistent effusion, so hampering in convalescence: while with one exception—the case with left ventricular failure—a rise in pulse later in convalescence always pointed to the pleura as the cause. This last point was well shown in Case 39 whose convalescence was extremely satisfactory until the 19th day when he complained of abdominal pain and felt unwell; a rise in pulse rate was the only significant sign but it was sufficient to predict that an effusion was developing on the operative side. It was apparent to clinical signs and radiography the following day and aspiration allowed his convalescence to continue uninterrupted. The late effusion was best illustrated by Case 2 who had a small effusion late in convalescence which resolved, but after returning home a massive effusion developed and necessitated readmission to hospital (Fig. 15, Campbell, 1948). We have regarded these as mechanical setbacks and have not allowed them to retard progress by prolonging bed rest unduly, and this, with the exception of Case 2, has been justified for subsequent examinations have shown a clear and moving diaphragm. There has been no evidence that these effusions are associated with pulmonary emboli and, except once where a very small amount of fluid was noted on the opposite side, the effusions have always been on the side of operation. Persistent hæmorrhagic effusion occurred in Case 5 only, necessitating transfusion; repeated aspiration of 11 pints in all was needed over 4 weeks, the fluid gradually decreasing in colour and gradually in amount until it quite suddenly stopped and did not recur. The correlation between temperature and effusion is roughly shown by an average length of 3 days in those with no fluid, of 5 days in those with moderate effusions, and of 8 days in those where aspiration was needed.

The third and most serious complication of cerebral thrombosis occurred in 3 of the 43 post-operative cases. Case 1, a boy aged 7, with a hæmoglobin of 126 per cent, had a history of a brain abscess on the left side when 2 years old. A right hemiplegia of moderate severity which was noted immediately after operation began to improve on the seventh day and when he was discharged on the sixteenth day there was only a slight limp. He has made a complete recovery. Case 8, an adult, was a severe case with hæmoglobin of 141 per cent and had the severe drop in blood pressure in the first 36 hours already noted. The two factors of marked polycythæmia and shock were therefore present to encourage thrombosis, and this was noticed in the first 24 hours, the left anterior cerebellar artery being involved. Recovery from this was complete but on the eighth and twenth-fourth days he had thrombosis of systemic arteries and on the twentieth day a thrombosis of the anterior cerebral artery. From this severe complication he has made a gradual but not a complete recovery, a disappointment in view of the excellent physical result. The third, Case 29, was a boy of ten, with a hæmoglobin of 158 per cent; he was drowsy after operation and was found to have a thrombosis of the cortical ascending parietal branch of the left middle cerebral affecting the arm area. The blood pressure was not unduly low after operation but during the operation there was marked collapse when the right pulmonary artery was occluded, and it was probably then that the thrombosis occurred. He is left with impairment of movements of the right arm and hand, a considerable disadvantage though compensated by an otherwise excellent result.

A further severe complication which we regard as due to the operation was in Case 21 who was admitted to Oldchurch County Hospital one month after discharge from Guy's after a prolonged post-operative stay of 56 days due to persistent temperature and left ventricular failure as already mentioned. The cause of this was a cerebral abscess which was successfully dealt with by aspiration one month later. It is not impossible that a post-operative thrombosis was the basis of this complication, though it was some time after.

It will be seen that cerebral thrombosis is the most important complication and one which to a lesser or greater degree nullifies the physical benefits that an otherwise successful operation gives. It should be most feared in those with gross polycythæmia.

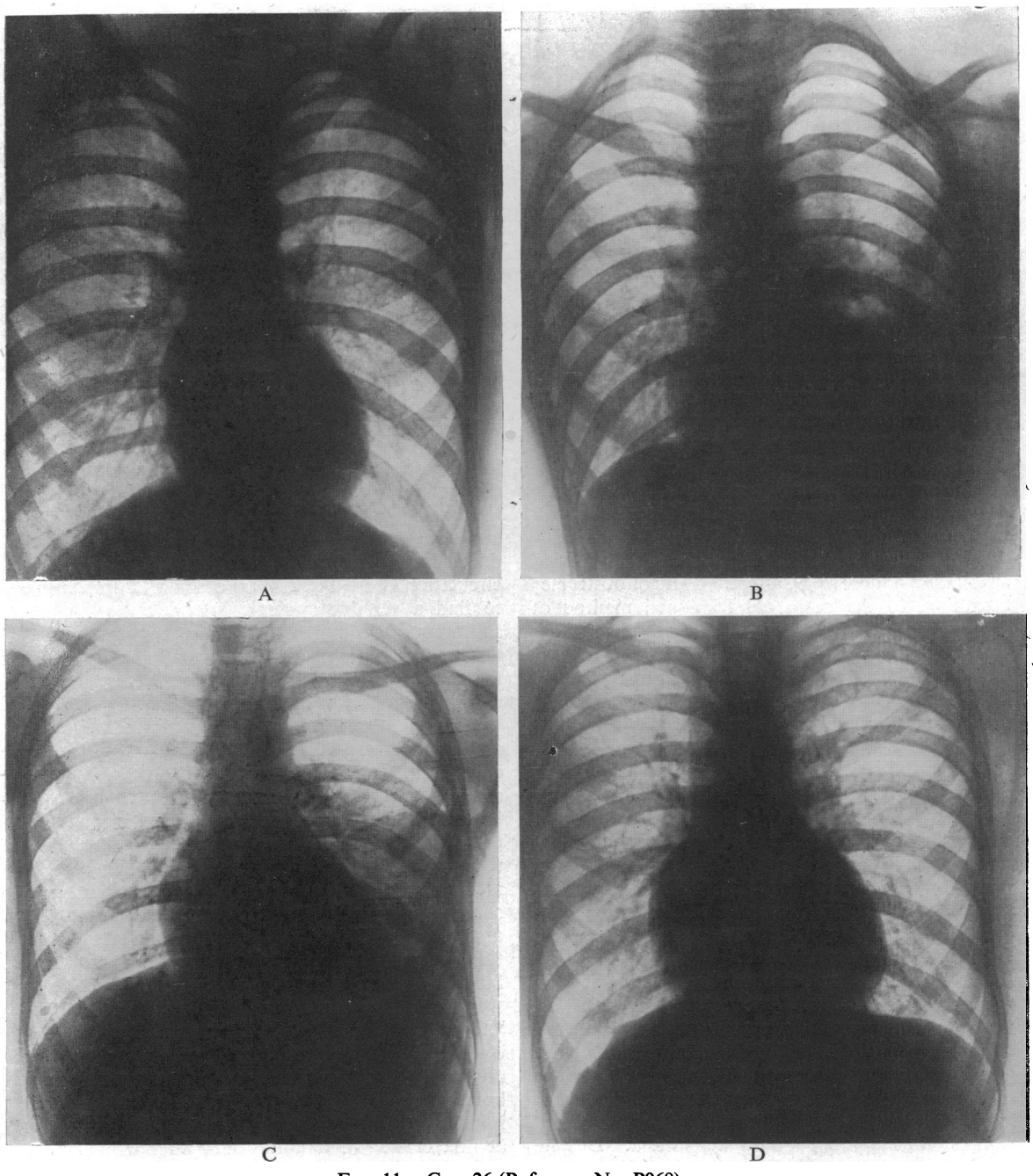


FIG. 11.—Case 26 (Reference No. P069).

(A) Teleradiogram in P.A. position before operation showing the clear lungs and some pulmonary bay with a raised apex. Both pulmonary branches can be seen (9/1/48).

(B) Portable radiogram, one day after operation, showing a moderate sized effusion at the left base with some collapse (11/2/48).

(C) Portable radiogram, showing gradual clearing of the effusion, the heart size becoming more visible but not yet easy to measure accurately.

(D) Teleradiogram, 7 months after operation, showing some increase in heart size from before operation, c.t.r. 54 instead of 46, with less apparent change in the density of the lung than in many cases, although the result was most successful.

with severe operative and post-operative shock, and in adults more than children. Prophylaxis with anti-coagulants introduces the danger of hæmorrhage and for this reason we do not use it. Nor have we felt sufficiently confident to start treatment by heparin within three days of operation, though there is a strong case for giving it promptly if thrombosis is diagnosed. If blood loss at operation has not been great, and polycythæmia is still present, blood letting is indicated and also intravenous fluids, by which method the heparin can be given.

In comparison with these three, other post-operative complications are slight. Some collapse of the lung on the side of operation, independent of effusion, occurred and required no treatment except continuation and insistence on breathing exercises. A small pneumothorax was present in one case and required no treatment and in only one was surgical emphysema of any degree present. Obstruction to breathing by secretions in the upper respiratory tract is largely obviated by insistence on breathing exercises and postural coughing, but was present in 17, though in only 4 did it cause any anxiety. Case 3 responded to the old fashioned steam tent, and Case 49, after 36 hours of obstructed breathing, to the more old fashioned method of turning him upside down, both exhibited by a watchful and wise ward-sister. Cases 40 and 41 both had severe laryngitis, and in the first, where a heavy growth of yeasts was cultivated from a throat swab, tracheotomy was necessary but led to little delay in his complete recovery.

The disability in the arm on the operated side is remarkably slight, and though the vascular change is apparent in signs, symptoms are slight and the disinclination to move it is soon overcome. A diminution in the size of the pupil on the side of operation, occasionally with ptosis, is almost a constant finding but had generally disappeared by the time the patient was discharged. No disturbance in kidney function was noted.

On the whole the postoperative period is surprisingly tranquil and uneventful for such an extensive and eventful operation. Children stand it particularly well and it is the adolescent or adult who is more likely to present difficulty. No better example of this can be shown than Case 41, a gravely ill girl of 5, whose heart, as well as breathing, stopped during induction of anæsthesia. Despite this and an operation lasting over three hours, and later a severe tracheitis, she was out of bed on the fourth day. Excluding 12 cases where the stay was prolonged on account of cerebral thrombosis, wound sepsis, or pleural effusion, the average stay in hospital was 23 days, and the average time in bed was only 7 days.

Results of Operation

In these first 50 patients there have been 7 deaths, a mortality of 14 per cent. Two of these deaths were among the relatively small number of patients of 19 and over (Cases 1A and 48). One had a single pulmonary artery and died when it was clamped (Case 17). In the others, there seemed no special reason except their serious condition and the severity of the operation (Cases 12, 32, 35, and 38).

Fallot's tetralogy was present in five of the seven. The remaining two had somewhat more complicated forms of morbus cœruleus, one having a single auricle in addition (Case 35) and the other having an infundibular stenosis, an aorta arising from the right ventricle without transposition of the pulmonary artery, and a small left ventricle with its only exit a ventricular septal defect (Case 32).

There were three patients where no anastomosis could be performed. One, an error in diagnosis already described, because the pressure was high in the pulmonary artery (Case 28), one because the right pulmonary artery was too small (Case 27), and one because of the technical difficulties introduced by the enormous dilatation of the collateral circulation (Case 14). In no case was it impossible to find a suitable systemic artery, but often the shortness of the subclavian led to practical difficulties and was, we think, sometimes the reason for the lack of success. This leaves 40 cases to be considered.

There were three where, in our opinion, there was no improvement (Cases 11, 19, and 22), though even then the parents of two thought there had been some. In one, the anastomosis was thought to be too small at the time (Case 11) and in a second there was bleeding from the anastomosis when it was nearly completed, and to save the patient's life many stitches had to be put in that probably led to some intra-arterial thrombosis and occlusion of the anastomosis (Case 22). It is possible that these three were also errors in diagnosis and not technical failures of the operation, but we do not think so.

In four the result has only been included as "fair." In one of these a similar difficulty with bleeding from the anastomosis occurred (Case 43); after operation he was able to walk upstairs which he had never done before but was not greatly improved. In the second (Case 21) the result was at first classed as a failure and after a long convalescence a cerebral abscess followed: when seen later after this had been cured, he was certainly improved but not as much as most of the others. In the other two the result was as good as possible as far as the heart was concerned, but the patients were handicapped by thrombosis resulting in partial hemiplegia; in one of these severely (Case 8), and

in the other, causing much limitation of his arm and hand movements but not preventing a great increase in his physical activity, so that he could walk about the greater part of the day (Case 29).

This leaves 33 of the 50 cases (66 per cent) where the result was almost perfect. The patients' capacity was enormously increased up to walking several miles instead of a few hundred yards, or getting about all day instead of being an invalid mainly in the house. To the parents the improvement seemed almost miraculous. One boy (Case 1) who had been tied to his mother's apron strings was at school within two months and after a year was running about all day, playing cricket, and could easily walk 4 miles. After running fast for 25 yards there was slight cyanosis in his lips and nails and some dyspnœa, but this cleared up very rapidly.

Another (Case 2) who had rarely been out of his parents' sight had within a year walked 6 miles and climbed hills in Switzerland as easily as his parents: he was anxious to become a medical student and was doing well at school.

Another (Case 4), who had been carried into hospital by his father, was leading a normal life at school and was able to go roller skating within a few months. Case 23, aged 19, who had been a complete invalid, had been on a camping holiday and had walked 6 miles. Such accounts could be repeated for nearly all the patients and all who have done less well have been mentioned individually.

The cyanosis was also much improved. In most of them it was absent on casual inspection though it could generally be seen in the nails at rest (but often not in the lips) and tended to be noticeable only on a cold day or after vigorous exertion. In one of the severe cases (Case 6) the cyanosis was still moderate, but it had been extreme and the child was able to do so much more that we feel it right to include her in this group. Case 49, with tricuspid atresia, who was very severely limited has not yet made as much progress as the others, but after leaving hospital fit and apyrexial, he was admitted elsewhere a month later with suspected bacterial endocarditis. He made a good recovery and can get about all day and his limitations seem more his unusually poor muscles than his heart. His colour has improved, but cyanosis can still be seen in his lips. We hope on the analogy of other cases that his improvement will go still further as his muscles improve.

The clubbing of the fingers also disappeared in some patients in the course of four or five months; some improvement was generally noticed very early. Where the clubbing was gross it seems more doubtful if it will disappear entirely, though it has changed greatly and become much less noticeable in the course of six months. No trouble has been experienced from the arm where the subclavian was divided, though the brachial pulse has not returned with sufficient strength to be able to measure the diastolic blood pressure in this arm.

In nearly all the successful cases a continuous murmur, such as is heard with a patent ductus, has persisted though often the thrill seems less than might be expected with the murmur. This is a good guide to the success of the operation and unless such a murmur can be heard early and easily, it is unlikely that the patient is going to be one of the most successful results. Occasionally, when there was less improvement than usual, there was a systolic murmur only.

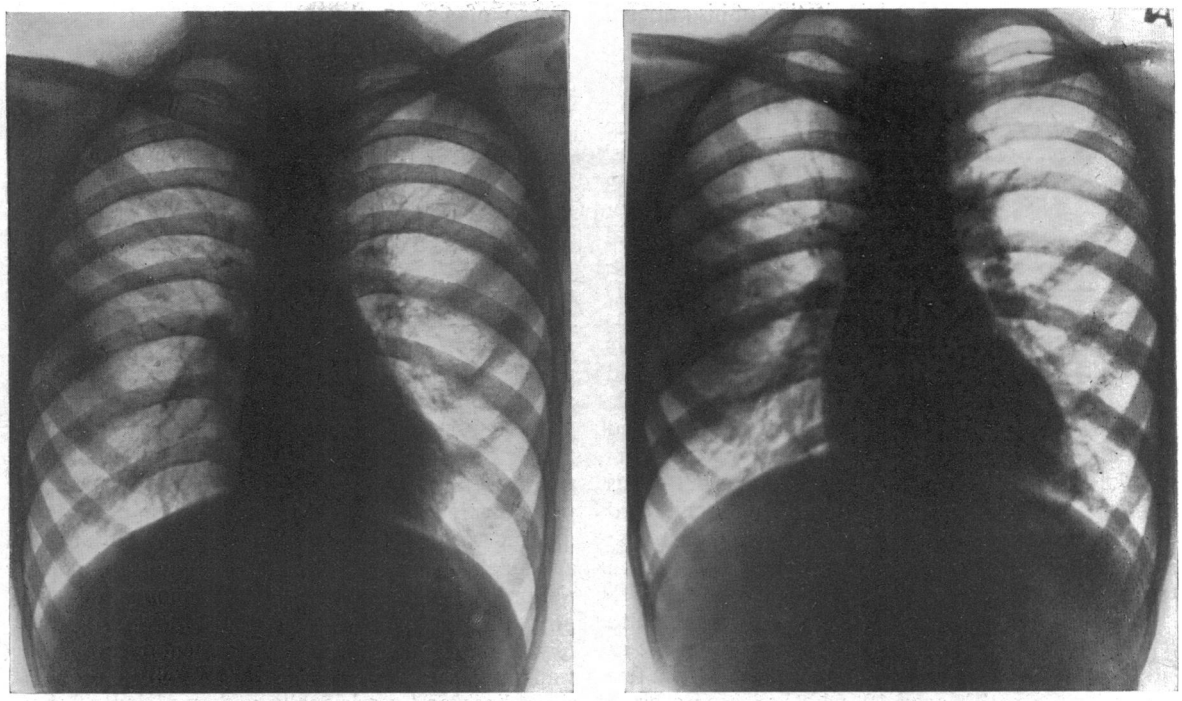
The enormously increased ability of these patients to get about often reveals orthopædic disabilities that have not mattered previously. Owing to the poor development of the muscles through lack of use and the small blood supply, and sometimes to an added deformity of the limbs from their continuous squatting, their new activity reveals many postural defects. We have made it almost a routine for the patient to have exercises and to be trained in walking correctly, and with simple supervision on these lines and occasionally with wedging of the shoes they have made rapid progress and had no serious difficulties.

We were at first a little surprised that there was not a quicker increase in the weight. As already stated, parents were nearly always anxious about the difficulty of making their children eat, and generally the appetite improves at once and they eat well. Probably the greatly increased activity prevents them having anything to spare for putting on weight for a time, but they can be expected to gain after a few months rather than a few weeks, and most of them have increased and gone some way to catch up with the weight that is normal for their age.

Increase in Size of the Heart

One of the points that has been emphasized as a drawback to the Blalock-Taussig operation is the increase in the size of the heart that may be expected from the work added by the left to right shunt. We have, therefore, paid special attention to the size of the heart before and after operation.

Of the 50 cases, 7 died, 3 had no anastomosis, and in 3 the operation was thought to have been unsuccessful. This leaves 37 where the heart size can be compared before and after a successful anastomosis. Owing to many of these patients living so far from London it has not been easy to get a regular follow up. We have, however, seen and obtained reasonably comparable radiograms in all



A B

FIG. 12.—Case 23, a boy aged 19, with Fallot's tetralogy.

(A) Before operation, showing a slightly raised apex and a straight left border.

(B) Eight months after operation, showing some increase in the size of the heart, the c.t.r. being 48 instead of 42, but still within normal limits. There is also some increase in the shadow of the pulmonary artery, especially on the left, the side of the operation as the aortic arch was right-sided.

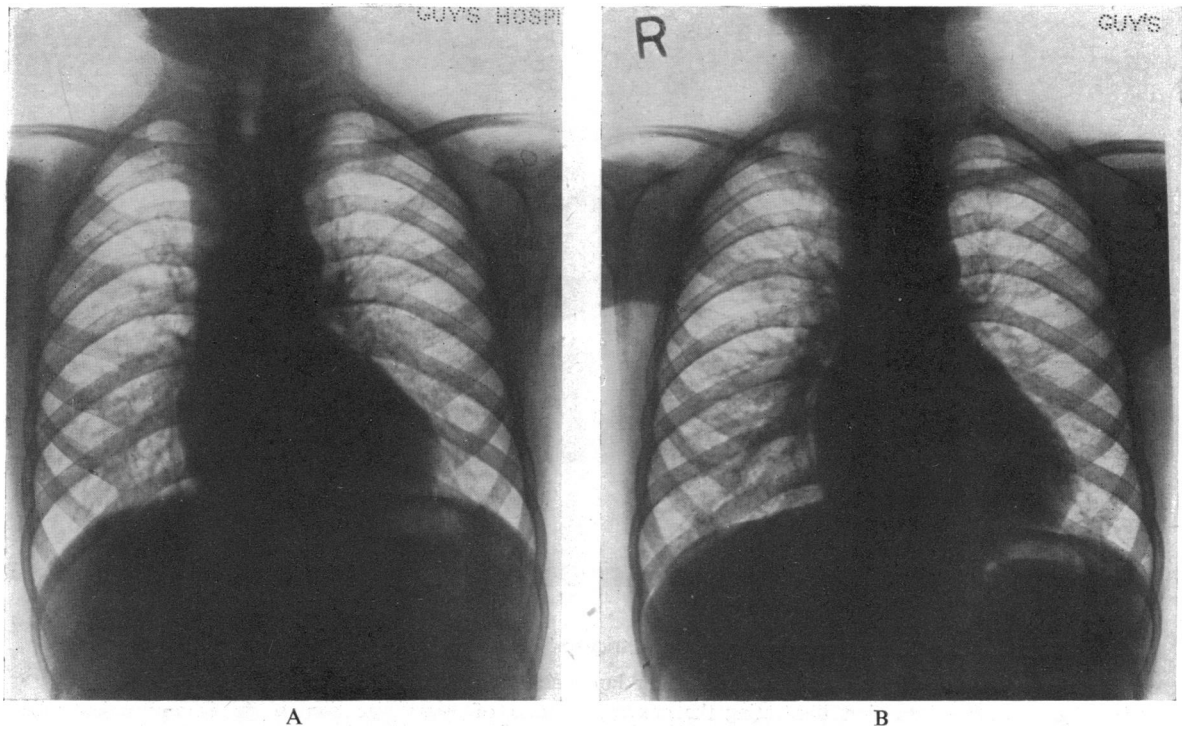
but two where we are relying on reports from their doctors: in neither of these had the heart increased in size a few weeks after operation. In one patient who lived in Cyprus we were fortunate in getting a report and teleradiogram from Dr. Hills who had taken the films at Guy's Hospital before operation. Examples of an increase of average amount, of very little change, and of the greatest increase we have seen are given in Fig. 12, 13, and 14 respectively.

This leaves 35 cases to be considered, and we have taken separately those where the follow up was more than a year after the operation.

Of the cases operated on more than a year ago all 13 have been followed up and teleradiograms have been obtained. Two of these showed no significant change in the size of the heart, but there was some increase in the other 11. In 6 cases there was no further increase after the first month and in another 3 there was little or no increase after about four months, but 2 who had not shown much increase in the first few months showed some increase between four months and a year. The average increases, shown in Table III might look like a slow but progressive rise; however, consideration of the individual cases shows only 3 out of 13 with any increase after four months (from 51 to 55). Naturally these three will be followed up with special interest to see if they are in fact exceptions.

Of the remaining 22 cases, some had been followed up for 11 months and all for more than 7 months, except three so far only followed for 5 months. Three of these showed no significant change in the size of the heart, but there was some increase in the other 19. In 10 cases there was no further increase after the first 4 to 6 weeks; in 9 there was some increase after this, but in 4 of the 9 it was only by 2 or 3 points per cent and in another 3 by only 4 points per cent. We had not, as a rule, intermediate records at about 4 months in these patients, but in the three where we had, the increase had all taken place by this time. The average increases are shown in Table III.

Taking the two groups of cases together the average increase in cardio-thoracic ratio was from 48·0 to 52·7 per cent. It had not increased in 5 of



A B

FIG. 13.—Case 29, a boy aged 10, with Fallot's tetralogy.

(A) Before operation showing a blunt apex and a fairly large pulmonary bay.

(B) Four months after operation showing hardly any increase in the size of the heart (c.t.r. inchanged at 52) although the anastomosis was most successful, as shown by his improved ability to get about and the improved colour. The pulmonary artery is more prominent on the right which was the side of the anastomosis as the aortic arch was left-sided.

TABLE III

CARDIO-THORACIC RATIO BEFORE AND AFTER BLALOCK-TAUSSIG OPERATION

Number of cases	Before operation	After operation			Time after operation
		About one month	About four months	At last visit	
13	47·2	51·0	51·8	52·5	12 mo.
22	48·6	50·9	—	52·8	6–11 mo.

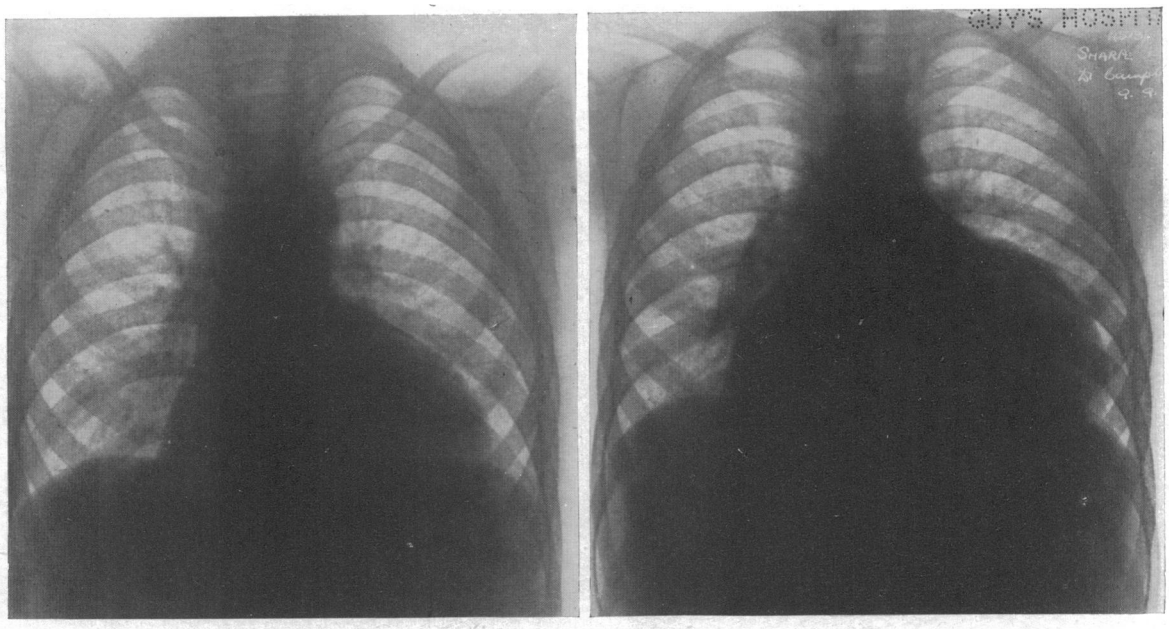
the 35 cases. It had increased by 2 to 3 points per cent (from 48 to 50 or 51) in 8, by 4 or 5 points per cent in 10, by 6 to 8 points per cent in 10, by 10 points per cent in 1, and by 12 points per cent in 1 case. In the last two the hearts had been small before operation (c.t.r. 41 and 44).

Only one patient (Case 33, Fig. 14) has made us anxious about his future by the degree of increase in the size of his heart. His clinical improvement was as much as in the others and there was difficulty in restraining him from doing everything. He was treated as a normal child at school except for games and had nothing except occasional attacks of tachycardia.*

PULMONARY VALVULOTOMY

During the same period 6 patients have been submitted to the operation of pulmonary valvulotomy. Four of these were thought to have Fallot's tetralogy with pulmonary stenosis as an important feature and the other two were thought to have pure pulmonary stenosis with some degree of patent

* We have since heard that eight months after operation he developed bronchitis with a temperature of 103 degrees; œdema of the legs soon followed, the temperature persisted, and he died in a few days.



A B

FIG. 14.—Case 33. From a boy aged 4, with tricuspid atresia and non-functioning right ventricle.

(A) Before operation showing an enlarged and rather horizontal heart.

(B) Five months after operation illustrating the greatest increase of heart size seen so far in any patient after operation, the c.t.r. having increased from 61 (11·0/18) to 69 (13·0/18·75). The pulmonary artery had become more prominent on the right, the side of the anastomosis, as the aortic arch was left-sided.

foramen ovale or auricular septal defect. The operation appears to be of much greater danger and three (Cases P031, H117, and H121) of these six have died, but they were all older patients, 22, 24, and 29, and this and the relatively small numbers make any exact comparison impossible.

In the remaining three, the operation was successful and the patients were considerably improved in colour and in their ability to move about without acute dyspnœa. Unfortunately, two of these were handicapped, one by hemiplegia (Case H109) and one by some disability of the leg from arterial embolism (Case P079). Whether this is a greater risk with valvulotomy remains to be seen because here again, they were older cases, aged 18 and 23, and it appears that age (though it may be the associated degree of polycythæmia) makes thrombosis and embolism a greater risk.

The remaining patient (Case H107, Case 2, Brock, 1948), a girl aged 11, was as brilliantly successful as any with an anastomosis. She was active all day and was able to walk several miles and looked a normal colour with only trivial or even doubtful cyanosis on careful examination. Her arterial O_2 saturation had increased from 81 to 91 per cent. Twelve months after operation, she has developed no signs suggestive of pulmonary regurgitation.

SUMMARY AND CONCLUSIONS

The method of chosing the first 50 patients for the Blalock-Taussig operation at Guy's Hospital and the results obtained have been described. Three cases were thought to have tricuspid atresia with a non-functioning right ventricle, and all the others Fallot's tetralogy, though sometimes with a known or suspected complication. All had great disability and severe or moderate cyanosis dating from birth or early infancy, with polycythæmia and clubbing of the fingers.

Most of these patients had a systolic murmur—often not very loud—in the pulmonary area, and in about half of them a thrill could be felt at this site. None had a diastolic murmur or a greatly accentuated second sound. Four-fifths of the patients squatted habitually and panting on exertion was nearly as characteristic.

The heart was generally within normal limits though some right ventricular hypertrophy could be seen on cardioscopy and shown electrocardiographically; sometimes the heart was small, and

occasionally a little, but never much, enlarged. If the cardio-thoracic ratio is under 45, the heart will probably stand a fairly large anastomosis with still greater improvement for the patient. More experience will be needed to decide how much enlargement of the heart may be allowed: slight enlargement with the c.t.r. 52 to 54 should certainly not contra-indicate operation but larger hearts with the c.t.r. 55 and above require special consideration and the improvement may not be so lasting.

The heart was sometimes sabot-shaped and sometimes of more normal shape with a straighter left border or even with a slight prominence of the pulmonary region. The pulmonary vascular shadows were generally much diminished though sometimes more mottled shadows, probably produced by the collateral circulation, made this decision difficult.

The pre-requisite of a successful operation is that the disability and cyanosis should be mainly due to an inadequate blood flow to the lungs and, in general, this is indicated by the clinical and radiological findings that have been given.

The other prerequisites are that there should be a suitable systemic artery and a pulmonary artery large enough for an adequate anastomosis. Angiocardiography helps with both these points by showing the anatomy of the aortic branches and of the pulmonary artery and so in helping one to plan the details of the operation that is most likely to be feasible and successful. Careful radioscopy should generally be able to decide about a suitable pulmonary artery.

The operation was usually an end-to-side subclavian-pulmonary anastomosis on the side opposite to the aortic arch. The aortic arch was right-sided in a quarter. Reasons have been given for thinking it is better to operate on the left side regardless of the side of the aortic arch and this has become the usual routine recently. Our conclusions about the surgical procedure that should be adopted have been summarized (page 188).

The immediate upset caused by the operation is less than might be expected and children stand it well and recover quickly. Morphia should be used in small quantities only, and intravenous fluids are not needed in large amounts, though fluids should be taken freely by mouth before and after operation. Breathing exercises should be started at once and carried out vigorously. The only complications that are at all common are pulmonary—some collapse of the lung and pleural effusion. These generally clear up quickly though they often need aspiration once and may sometimes be slower and cause trouble. Arterial thrombosis at the time of operation or soon after has been the second main complication and as already stated two of these patients have been left with some residual disability. This risk seems greater when the polycythæmia is severe.

On the average the temperature had settled to normal by the third day, and excluding 12 cases where there were complications from cerebral thrombosis, wound sepsis, or pleural effusion, the patient was up and getting about the ward on the eighth day and was able to leave hospital after 23 days.

There were 7 deaths in these 50 cases—a mortality of 14 per cent. There were 3 others where no anastomosis could be performed.

Of the remaining 40, there were 3 where we thought the improvement of little or no significance, and 4 where we have only classed it as fair, though in 2 of these the result was excellent as far as the heart was concerned but was marred by some residual disability from thrombotic hemiplegia.

In 33 of the 50 cases (66 per cent) the results were almost perfect and the patient was able to get about all day and walk up to 5 or 6 miles instead of a few hundred yards or less. The cyanosis almost disappeared except slightly in the nails and on a cold day. Many children quickly started at ordinary schools, and cricket, camping, and roller skating became the occupations of some who had before been invalids doing hardly anything.

A murmur similar to that of a patent ductus arteriosus was heard on the side of the operation in the successful cases. After operation with the added work of the heart from the new left to right shunt the heart generally increased in size a little but not greatly. In 5 of 35 cases there was no significant increase; in the remainder the cardio-thoracic ratio rose from 48·0 per cent to 50·9 when the patient left hospital and to 52·7 per cent when they were last seen (generally after 7 to 14 months). Although these average figures might look like a progressive, if slow, increase, individual cases show that the greater part of the increase had generally taken place in the first 4 to 6 weeks. Only one patient so far—and he had tricuspid atresia—has caused us any anxiety for the future by the size of the increase in his heart, and symptomatically he has improved as much as the others.

It was particularly encouraging that none of the patients seen a year after operation had failed to maintain or increase all the improvement they had made at the earlier follow-up after a few months.

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Appendix on Cardiac Catheterization

By G. A. Zak

Nine of these cases underwent cardiac catheterization. Most were investigated when the procedure was still at an early stage of development at this hospital; with increasing skill and improved technique the results in later cases have been more complete.

In view of the envisaged operation the main object of venous catheterization was to get information regarding the pressure in the right ventricle and pulmonary artery, the presence and localization of shunts, and the volume of the pulmonary blood flow in relation to the height and weight of the patient.

It is not intended to discuss the problems and findings of cardiac catheterization in detail. From the rapidly growing data on this subject several widely accepted facts have emerged. For example the presence of blood of significantly higher oxygen saturation in the right auricle than in the venæ cavæ indicates arterial blood passing from the left auricle through an auricular septal defect; and the sudden fall in pressure from a high to a low level on passing from the right ventricle to the pulmonary artery is the best evidence of pulmonary stenosis. To calculate the pulmonary arterial flow with the aid of the Fick principle mixed venous blood must be obtained, and the mixing of the venous blood from the various sources may be incomplete in the right auricle. If the catheter can not be got into the pulmonary artery a blood sample obtained from the outflow tract of the right ventricle can be substituted. The pulmonary arterial flow is not necessarily equal to the total pulmonary flow (pulmonary capillary flow) as an appreciable amount of blood may reach the alveolar capillaries by way of a collateral circulation. No attempt has been made to assess this.

The sudden fall from high to low pressure on passing from the right ventricle to the pulmonary artery and sometimes the feel of release from a grip on the catheter is the best evidence of pulmonary stenosis, and a decreased pulmonary arterial flow is confirmatory evidence. If it is not possible to reach the pulmonary artery with the catheter, a high pressure in the right ventricle and a decreased pulmonary flow may be taken to lend good support to an assumption of pulmonary stenosis in the absence of clinical signs of mitral stenosis or heart failure.

The magnitude and direction of an overall intracardiac shunt is calculated by deducting the pulmonary arterial from the systemic blood flow or vice versa. The adjective "overall" is used to denote that in many instances there is actually a to-and-fro movement of blood from the two sides across a single septal defect but as a rule one component is larger to such an extent as to dominate the clinical picture.

An evaluation of the output of each of the two ventricles of the heart, and the consequent calculation of a possible shunt is an approximation only. The calculation of the pulmonary arterial flow is often the more reliable. Of the three components necessary for its computation, the O_2 uptake through the lungs and the O_2 content of the mixed venous blood are known. If the degree of the arterial oxygen saturation is normal the pulmonary venous blood is taken to be of equal saturation. Even if it is lowered one is justified in assuming the pulmonary venous blood as still being 95 to 98 per cent saturated with O_2. This was shown to be true by other investigators in the absence of pulmonary conditions likely to interfere with the gaseous exchange in the alveoli.

Case No., Age, and Date of catheterization	Pressure (mean) in mm. Hg. from the skin of the back			Systolic B.P. (clinically)	Degree of O_2 saturation percentages				Output in % of the average output of 3·0 l. per min. per sq. m. of body surface		Hb. % 100% = 15·6 g. Hb.	Surface area in sq. m.	O_2 consumption in ccs. per min. at S.T.P. (B.M.R.)
	R.A.	R.V.	P.A.		R.A.	R.V.	P.A.	Syst. artery	System	Pulm.			
Case 23, 19 yr., 13/1/48.	15	43	—	115	29·0	33·1	—	46·5	100	30	120	1·24	170 (−5·0)
Case 49*, 8 yr., 6/2/48.	14	(27)	—	100	SVC 46·8	23·2	—	58·8	180	—	125	0·83	140 (−3·0)
Case 35, 7 yr., 23/4/48.	7	45	15	110	—	—	—	—	—	—	—	—	—
Case 36, 17 yr., 12/5/48.	12	—	—	122	60·0	—	—	83·0	85	—	118	1·42	200 (0·0)
Case 42*, 19 yr., 20/5/48.	11	—	—	95	55·7	—	—	73·0	110	—	140	1·32	220 (+19·0)
Case 39, 11 yr., 25/5/48.	16	44	—	100	47·7	—	—	55·6	250	—	143	1·00	185 (+4·5)
Case 48, 27 yr., 4/6/48.	15	50	—	115	38·2	46·6	—	62·7	90	35	153	1·77	250 (+4·0)
Case 44, 11 yr., 15/6/48.	10	40	10	110	67·6	60·7	60·1	71·8	110	45	160	1·01	165 (−5·5)
Case 46, 15 yr., 18/6/48.	10	62	10	125	53·3	55·4	46·6	61·8	100	50	142	1·59	210 (−5·5)

* In these two cases the diagnosis was tricuspid atresia; in all the others it was Fallot's tetralogy.

The systemic blood flow in cases of septal defects cannot, however, always be calculated with the same degree of reliability as the right ventricular output. Here, the mixed venous blood component is the weak link in the chain. Often there is evidence of imperfect mixing in the right auricle, the coronary vein as a rule adding blood very much lower in oxygen content. The inferior vena cava in turn produces, generally speaking, more saturated blood than the superior vena cava. A blood specimen obtained from the inferior vena cava, however, may not be of uniform composition on account of the nearness of the openings of the hepatic veins. This fact makes the recognition of small atrial septal defects producing admixture of arterialized blood uncertain if one is to rely on the taking of single blood samples only. Because of the errors involved in calculating the pulmonary and the systemic flow one must hesitate to diagnose a small overall right to left shunt on small differences; larger differences can however be taken as reliable.

In these nine cases the clinical diagnosis was supported. Such support was felt to be forthcoming in cases of Fallot's tetralogy if there was evidence of pulmonary stenosis and an overall right to left intracardiac shunt.

Of the two cases diagnosed on clinical grounds as having tricuspid atresia only a single right chamber with normal right auricular pressure could be demonstrated and no evidence of an atrial left to right shunt were found. In neither of these cases could the catheter be made to cross over to the left auricle.

There was evidence of an overall right to left shunt in every case.

The A–P diameters of the chest ranged from 18 to 24 cm. with an average of 19 cm. To obtain the pressure as measured from the sternal level 15 mm. of Hg. has to be deducted from the values given in this table.

The catheterizations were carried out by Dr. G. A. Zak and Dr. H. E. Holling.

Case 23. Pulmonary artery not entered as tip of catheter got caught in papillary muscles towards the base of the heart, without getting near the pulmonary valves.

Case 49. Results vitiated by crying. The superior vena cava value is substituted for the right auricular value, as the former followed a period of calmness. It was thought at first that the right ventricle had been entered, but probably the catheter was in the coronary sinus, with the pressure high from partial occlusion of the lumen of the catheter.

Case 35. The use of an inhalation anæsthetic made gas analyses of the blood specimens impossible. The raised right ventricular pressure in conjunction with the lowered pulmonary arterial pressure indicates stenosis of the pulmonary ostium.

Case 36. Venospasm led to abandonment of procedure.

Case 42. Only the right auricle could be entered. The tip did not pass into the left auricle through a possible atrial septal defect. If such a defect was present, a left to right shunt through it would appear unlikely on account of the S.V.C. and I.V.C. saturations, which agreed with the value found in the R.A.

Case 39. The right ventricle was only entered with difficulty and attempts at entering the pulmonary artery failed. Tip was at the base of the heart. The pressure in the right ventricle was recorded but no blood specimen was obtained, and when on withdrawing the catheter too far this chamber had been left the permissible screening time did not allow further search for the right ventricle.

Case 48. The pulmonary artery could not be entered though tip of catheter was brought to base of heart. Good agreement between S.V.C. and right auricular saturation. The increased right ventricular saturation over the right auricular sample favours presence of a ventricular septal defect, giving rise to shunting of the blood in both directions though differing in quantity. The smaller opposing shunt can be detected if the tip of the catheter happens to be near such a defect, which seems to have been the case here.

Case 44. Superior vena cava, 54 per cent, and inferior vena cava, 60 per cent saturated, favour a small left to right shunt through an atrial septal defect, which is, however, far outweighed by the large right to left shunt through a ventricular septal defect or overriding of the aorta.

Case 46. Good agreement between the saturation values in both venæ cavæ, right auricle, and right ventricle. The low pulmonary artery saturation is thought to be due to obstruction by the catheter of an already narrowed ostium for about two minutes, prior to withdrawing the blood sample. The pulmonary flow has been calculated from the right ventricular saturation.