Fallot's tetralogy

A 20-year surgical follow-up

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At Guy's Hospital, between 1947 and 1951, 81 patients with Fallot's tetralogy survived operations with a functioning systemic-pulmonary arterial anastomosis; between 1948 and 1951, 46 patients with the same condition survived a Brock operation. As far as possible these patients have now been followed for 20 years.

At the end of this period half of the group of patients with an anastomosis are known to be still alive and 40 per cent are known to have died. About half of these survivors are able to lead virtually normal lives, but only a quarter are doing so with their original operation only. Altogether there are 25 survivors with their original operation only, but only about one-third of these still have good results. The commonest cause of death was hospital mortality related to a subsequent operation. The commonest non-fatal complication was bacterial endocarditis.

A little more than half of the group of patients who had direct operations are known still to be alive and nearly one-quarter are known to have died. Of the 25 survivors, 22 are able to lead virtually normal lives, half of these with only their original operation. In this group also the commonest single cause of death was related to subsequent operation; there were fewer non-fatal complications and in particular only one patient in the whole group had bacterial endocarditis.

The anastomotic and direct operation groups are not strictly comparable, but the difference in the incidence of bacterial endocarditis certainly suggests that this is a particular risk of the anastomotic operation. Severe polycythaemia, developing in deteriorating patients, appeared to be an important hazard. Deterioration in both groups was apparently often related to increasing right ventricular outflow obstruction.

The relevances of the findings in this follow-up study to decisions about subsequent operations and to current decisions about first operations for patients with Fallot's tetralogy are discussed.

In 1947 Professor Blalock visited Guy's Hospital as an exchange visitor from the Johns Hopkins Hospital and during his stay performed 9 subclavian-pulmonary arterial anastomotic operations for the relief of cyanotic congenital heart disease. Maurice Campbell (1948) has given a full account of this event which marked the beginning of the referral to Guy's Hospital of a large number of patients with this condition, nearly all of whom were seen by Maurice Campbell and upon the majority of whom Lord Brock and his team operated during the succeeding years. The initial results of the anastomotic operation were reported by Campbell and

Deuchar in 1953 and of Brock's own operations, of pulmonary valvotomy and infundibular resection, by Campbell, Deuchar, and Brock in 1954. Since then it has been possible to continue the observation of most of these patients in the cardiac clinics at Guy's Hospital. The survivors have now been observed for 20 years and much has been learnt about the course of cyanotic heart disease and the results of the various operations for the relief of Fallot's tetralogy. We are reporting here an analysis of the results of this long-term study.

I Anastomotic operations

Subjects and methods The original report by Campbell and Deuchar (1953) referred to 200 patients treated by anastomotic operations. Of this total, however, 71 had their operations performed at other hospitals, and at that time it was

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recognized that another 35 had some form of cyanotic heart disease other than Fallot's tetralogy. The diagnosis of Fallot's tetralogy was made in the remainder very largely on clinical grounds; many were investigated by cardiac catheterization or angiocardigraphy before operation but the quality of these investigations was then somewhat primitive. Since then nearly all the survivors have been reinvestigated, have had direct operations, or have come to necropsy. For the purpose of this study only those patients whose condition in the light of all the currently available information appears to have been Fallot's tetralogy have been included. This report is therefore concerned with 81 patients who survived operation between 1947 and 1951 with a functioning anastomosis, out of the 96 now thought to have Fallot's tetralogy who were submitted to the operation; there had been 8 hospital deaths (8.3%) and 7 unsuccessful operations (7.3%).

There were 49 male (51%) and 32 female (49%) patients. Their ages at the time of operation ranged from 3 to 29 years, with a mean of 9 years: only 4 patients were over 20 years and 55 were under 10. All the patients were clinically cyanosed at rest and over half had a grade 3 or 4 disability (New York Heart Association). An end-to-side sub-clavian-pulmonary anastomosis was performed in 69 and, because of technical difficulties, an end-to-end anastomosis in 10; 2 patients had an aortic-pulmonary anastomosis of the Potts' type. The immediate results were good, with a conspicuous reduction or disappearance of cyanosis and striking increase in exercise tolerance, in 75; improvement was only marginal in the other 6 though an anastomotic murmur was audible after operation in 5 of them.

As far as possible all the patients were followed at yearly or two-yearly intervals by the cardiologists or thoracic surgeons at the hospital; in a few cases patients who lived distantly or who had emigrated were followed by reports from their local physicians or surgeons. All the survivors who were available were seen in the cardiac clinic by one of the authors between 1967-1970; reports from other centres were obtained during the same period about a few patients and two overseas patients who had not required medical attention in their own countries of residence wrote personal letters testifying to their good health. Patients who attended the clinic were on each occasion questioned about their general health, exercise tolerance, and symptoms that might be related to polycythaemia or heart failure; they had a routine clinical examination, urine test for albumin, haemoglobin estimation, posteroanterior x-ray of the chest, and a standard 12-lead electrocardiogram.

Follow-up results

Forty-one (50.6%) of the patients were known to be alive at the end of the follow-up, 33 (40.4%) were known to have died, and 7 (9%) were lost to follow-up at some time during the 20 years. The quinquennial attrition from

the series by death or loss of contact is shown in Fig. 1. During the first 5 years 7.5 per cent of the 81 patients deteriorated and approximately half of them (3.5% of the total number) died; 3.5 per cent had a second operation. In the second quinquennium 17 per cent of those surviving the first quinquennium died and 26 per cent deteriorated; 16.5 per cent had a second operation. The percentage of second operations plus deaths exceeds the percentage of those deteriorating in this group, and subsequently, because some of those reoperated upon had not deteriorated, but were regarded as having obtained inadequate benefit from the first operation or were offered another operation in order that they might take advantage of surgical advances which seemed to offer them further improvement. In the third quinquennium 17 per cent of those entering it died and 31 per cent deteriorated: 26 per cent had further operations. In the final quinquennium 14.5 per cent died, 26 per cent deteriorated, and 8.5 per cent had another operation. It will be noted that the mortality in the first 5 years

FIG. 1 Graphs showing the quinquennial reduction from death or loss to follow-up of the group of patients who had anastomotic operations, and the percentage surviving with only their original operation.

was low but subsequently was higher and differed little between each quinquennium, averaging about one-sixth of the survivors entering each period; similarly, deterioration was more common in the last 3 quinquennia, averaging about one-quarter of those entering. The above figures are illustrated graphically in Fig. 1 and 2.

At the end of the follow-up, 20 patients (49% of the 41 known to be alive and 25% of the original 81) were classified as 'good' on the basis of their being able to lead virtually normal lives, but only 9 of these were in this class with their original anastomotic operations alone; the other 11 had had subsequent operations. Fifteen were classed as 'moderate' (minor disability only), and of these only 4 had had further operation. Six were classed as 'poor' (serious disability) and only 1 of these had had another operation. There were therefore 25 patients at the end still surviving with their original anastomotic procedure, but only 9, about one-third, of these (11% of the original survivors) still had a good result. The mean age of the 41 surviving patients at the end of the follow-up was 29 years and the age range was from 23 to 38 years.

The commonest cause of death in the series was operative mortality in relation to a subsequent operation; 14 of the 33 deaths (43%) were due to this, representing a 38 per cent mortality from these operations (Table 1). This high mortality rate was largely due to intractable haemorrhage (8 cases) which was a common problem in patients who had deteriorated badly and had severe polycythaemia. The causes of death are listed in Table 2. Of the 33 patients who died, 25 were male and only 8 were female; age at death ranged from 5 to 34 years, with a mean age of 20 years.

It will be noted that death was not necessarily related to deterioration and of course the converse is also true. The various causes of deterioration are indicated in Table 3 and the quinquennial incidence is shown in Fig. 2.

A substantial number of non-fatal complications occurred during the period of study, involving 26 of the 33 patients who eventually died and 34 of the 41 patients who survived. Neither the nature nor the incidence of these complications differed significantly between those dying and those surviving (Table 4). These complications occurred during a total of 1142 patient-years of follow-up. Seven patients in both groups were free of non-fatal complications throughout their period of observation. A more detailed analysis of the complications observed, according to the organ systems involved, is given in Table 5.

FIG. 2 A graphical representation of the quinquennial deterioration, mortality, and reoperation rate in the group of patients who had anastomotic operations. The shaded area represents the patients known to be alive at each 5-year period. The blocks representing the deterioration, mortality, and reoperation rates show the number of patients affected, expressed as a percentage of the total number of patients at the start of each quinquennium.

Some patients were observed to lose their anastomotic murmurs in association with deterioration; in some of these occurring in the first few years, examination at necropsy or at subsequent operation showed that the anastomosis was no longer patent. Two patients whose murmur disappeared more than 10 years after the operation were interesting. They had both developed severe poly-

cythaemia and then suffered a major spontaneous haemorrhage (one gastrointestinal and the other pulmonary); coincident with the post-haemorrhagic haemodilution the anastomotic murmurs again became audible, presumably as the blood became less viscous, permitting better flow through the anastomosis and allowing turbulence to occur more readily.

II Direct operations

Subjects and methods The original report by Campbell *et al.* (1954) concerned 100 patients with Fallot's tetralogy who had had pulmonary valvotomy, infundibular resection, or both. Twenty-seven had been patients of the late Paul Wood and were operated upon at the Brompton Hospital. The whole series included patients who had been operated upon between 1948 and 1953. For this study, we have confined our attention to the 46 survivors of operations performed at Guy's Hospital up to the end of 1951.

There were 25 (54%) male and 21 (46%) female patients. Their ages at the time of operation ranged from 2 to 31 years, with a mean age of 13 years; 3 patients were over 20 years (one was 31), 18 were 10 years or under. Two patients had had a poor result from anastomotic procedures 2 years previously and therefore appear in both series now being reported. Approximately half the patients had a grade 3 or 4 disability and all were clinically cyanosed at rest. A closed pulmonary valvotomy was performed in 23 patients, an infundibular resection in 13, and both procedures in 10. The operation procedures have been described by Brock and Campbell (1950a, b). A substantial improvement was obtained in 42 (91%) and the result was considered very good in 26; 4 patients (9%) were considered not to have improved.

The follow-up procedure for these patients was the same as for the patients who had anastomotic operations.

Follow-up results

Twenty-five (54%) of the patients were known to be alive at the end of the follow-up, 10 (22%) were known to have died, and 11 (24%) were lost to follow-up during the 20-year period. The quinquennial loss from the series by death or loss of contact is shown in Fig. 3.

During the first year after this operation, 26 patients were regarded as having obtained a good result; this generally meant that they had only minimal disability, if any, and were acyanotic at rest. Of the other 20 survivors, 16 were regarded as improved but having some significant residual disability and cyanosis at rest, and the remaining 4 were not thought to have gained significant benefit. Of these 4, 2 had a successful anastomotic operation within

TABLE 1 *Subsequent cardiac operations performed after anastomotic procedures in 32 patients (4 patients had more than one additional operation)*

	No.	Operative mortality
Pulmonary valvotomy	12	5 (41%)
Infundibular resection	7	2 (28%)
Pulmonary valvotomy + infundibular resection	7	0 —
Total correction	11	7 (63%)
Total: all operations	37	14 (38%)

TABLE 2 *Immediate causes of death during follow-up period of 33 patients treated by anastomotic operations*

Further cardiac surgery	14
'Inadequate anastomotic flow' (including 4 sudden deaths in severely cyanotic patients)	6
Congestive cardiac failure	1
{ Cerebral abscess	4
{ Meningitis	1
Renal failure	2
Coagulation disturbances (1 mesenteric thrombosis, 1 puerperal haemorrhage)	2
Abdominal operation	1
Cause unknown	2

TABLE 3 *Causes of clinical deterioration in 74 patients with complete follow-up information after anastomotic operations*

	No.	Reoperated	Living at end of follow-up
Inadequate anastomotic flow	34	25	17
Congestive cardiac failure	4	1	1
Renal failure	4	0	1
Neurological disorder	2	1	0
Respiratory disorder	1	0	1

Note: The cause of the initial clinical deterioration was not necessarily the same as the cause of death in those patients who subsequently died.

5 years and one had a total correction procedure with a good result during the second 5 years; the last of the 4 improved without further operation during the succeeding 5 years, but the mechanism of this was obscure. One of the 'improved' patients had a second infundibular resection within 5 years. At the end of the first 5 years, 34 (74%) had maintained their condition, 6 (13%) had deteriorated, 2 (4%) had died, but both from non-cardiac causes, and 4 (9%) were lost to follow-up. One of these last 4 was the only patient who sustained a disabling complication from

an operation which was otherwise successful; this patient had a hemiplegia following the operation but her heart condition was good; she was lost when she emigrated with her family to Canada. Forty patients entered the second quinquennium and none had died by the tenth year; 5 (12.5%) deteriorated and 2 (5%) were lost. Seven (15%) patients had further operations of which 3 were Brock operations and 4 were 'total correction' procedures; there was no operative mortality. During the third quinquennium of the 38 patients, 5 (13%) died, 5 (13%) deteriorated, and 5 (13%) were lost; this unfortunately large number of patients lost from the series at this time was the result of some dislocation of the follow-up programme occasioned by Maurice Campbell's retirement. Four patients (8%) had further operations of which all were now 'total correction' procedures; 3 of these died in hospital including one in whom closure of the ventricular septal defect was performed on account of congestive cardiac failure following her original pulmonary valvotomy which led to pulmonary incompetence and considerable pulmonary plethora. Another patient (M.G.) who died after her second operation was also of interest; at her original operation an unusually effective infundibular resection led to flooding of the lungs and collapse of the systemic circulation, and for a time this had to be controlled by compression of the pulmonary artery. Subsequently the position stabilized and she recovered to achieve an excellent clinical result with complete relief of all disability; this was however associated with considerable cardiac enlargement and obvious pulmonary plethora. There was little change for 5 years but subsequently it was noted that the heart and pulmonary plethora were diminishing; this process continued and eventually she again became cyanosed and disabled. It was for relief of this situation that the second operation was undertaken and at that operation severe fibromuscular infundibular obstruction was found. During the final quinquennium there were 3 more deaths (10%), of which one was a fatal outcome of a Waterston-Cooley anastomosis intended to palliate before a more definitive procedure in a patient who had deteriorated very rapidly during the preceding 2 years. Three other patients (making a total of 20% of this group having further operations) had total correction procedures and all survived. Three patients (10%) deteriorated but none was lost to follow-up. The quinquennial statistics are summarized in Fig. 4.

At the end of the 20-year period there were

TABLE 4 *Non-fatal complications recorded in patients who had anastomotic operations (several patients had more than one complication)*

	In 34 of 41 survivors	In 26 of 33 patients dying	Total
Infective (SBE)	24 (15)	12 (6)	36 (21)
Haemoptysis	7	3	10
Cerebral embolism	4	1	5
Atrial arrhythmias	4	1	5
Epilepsy or Parkinson's syndrome	8	—	8
Renal and metabolic disturbances	5	4	9
Miscellaneous	8	10	18
Total	60	31	91

SBE = subacute bacterial endocarditis

TABLE 5 *Analysis of non-fatal complications occurring in 70 of total 81 patients involving a total of 1142 patient-years of observation*

Cardiovascular system	31
Subacute bacterial endocarditis	21
Atrial arrhythmias	5
Deep vein thrombosis	3
Congestive cardiac failure	2
Respiratory system	22
Haemoptysis	10
Recurrent respiratory infection	8
Asthma	2
Tuberculosis	1
Aspergillosis of lung	1
Central nervous system	18
Epilepsy	7
Cerebral embolism	5
Cerebral abscess	4
Meningitis	1
Basal ganglia lesion (? thrombotic)	1
Renal	9
Renal failure	4
Glomerulonephritis	2
Gout	3
Miscellaneous	11
Total	91

25 survivors (54% of the original 46 patients) and 22 of these (88% of the survivors or 48% of the original 46) were regarded as 'good'. Eleven of the patients (24% of the original 46) with good results had maintained their improvement without a second operation. The percentage of the patients surviving with their original operation at the end of each quinquennium is shown in Fig. 3. The mean age of the surviving patients at the end of the follow-up period was 31 years and the age range was from 22 to 42 years.

As in the anastomotic series, the commonest

cause of death was subsequent reoperation, accounting for 4 of the 11 deaths (36%), which represents a 19 per cent mortality from these operations (Table 6). In contrast to the anastomotic series the sex incidence of the 10 patients dying during the follow-up was virtually equal in this small number (4 male and 6 female); age at death ranged from 12 to 38 years, with a mean age of 28 years. The sole cause of deterioration, excluding non-cardiac complications which were either fatal or temporary, appeared to be declining pulmonary blood flow leading to a return of cyanosis and exercise intolerance. This seemed to be related to increasing right ventricular outflow obstruction as exemplified most strikingly by patient M.G. quoted above.

Non-fatal complications occurred in a number of patients, but the incidence of these was much less than in the anastomotic group. These are listed in Table 8.

Discussion

The effect of a successful anastomotic operation for Fallot's tetralogy on the patient's functional capacity, cyanosis, polycythaemia, and heart size has been reported previously, for patients from Guy's Hospital by Campbell and Deuchar (1953) and Campbell (1958), and for other series by, for example, Fabricius *et al.* (1963), Harris, Segel, and Bishop (1964), Shumacker and Mandelbaum (1960), and White *et al.* (1956). After a good operation the patient's disability is reduced or relieved, the colour is improved, the haemoglobin and packed cell volume are brought closer to normal levels, and the heart size is often somewhat increased. The last feature is clearly the consequence of a bigger cardiac output after the creation of a significant aorto-pulmonary shunt and the others are the indirect consequence of an increase in pulmonary blood flow. These features are so well documented and generally known that further data on these points have not been presented in this paper. An attempt has been made here to record the natural history (if that term can be used in this connexion) of the patients with Fallot's tetralogy after a successful anastomotic operation.

The progress of the patients during the first five years was relatively good and this is similar to the experience reported in the papers quoted above. After that time the mortality and deterioration rate increased, and then remained more or less constant till the end of the study. Similar findings were reported by Taussig *et al.* (1962) who found that 42 per cent of a group of patients who were living five years after the operation had

FIG. 3 Graph showing the quinquennial reduction from death or loss to follow-up of the group of patients who had direct operations, and the percentage surviving with only their original operation (the straightness of the last line from the start to the 15th year is coincidental as it is drawn through the correct number for each quinquennium).

TABLE 6 Additional cardiac operations performed upon 18 patients in direct operation series including 2 patients whose first operation was an anastomotic procedure (3 patients had a total of 3 operations each)

	No.	Operative mortality
Infundibular resection	2	0
Pulmonary valvotomy + infundibular resection	5	0
Total correction	10	2 (20%)
Ventricular septal defect closure	1	1 (100%)
Anastomotic procedure	3	1 (33%)
Total: all operations	21	4 (19%)

TABLE 7 *Immediate cause of death during follow-up period of patients treated by direct operation*

Further cardiac operation	4
Cerebral abscess (with SBE)	1
Meningitis	1
Ascending paralysis	1
Paradoxical cerebral embolism	1
Congestive cardiac failure	1
Haemoptysis	1

SBE=subacute bacterial endocarditis.

deteriorated by 10–13 years after and 12 per cent had died.

It is not possible to draw detailed comparisons between the Guy's Hospital series and the others reported, as there are obvious differences of age distribution, preoperative severity, criteria for selection for operation, etc. One striking postoperative difference, however, deserves comment. There have been reports from several centres (Folger and Otken, 1969; Hancock, Hultgren, and March, 1964; Leeds, 1958; Ross, Taussig, and Evans, 1958; Wagenvoort, DuShane, and Edwards, 1960) of complications apparently related to excessively large anastomoses (Ferencz, 1960). The commoner of these complications is the development of pulmonary hypertension with gradual diminution of the anastomotic shunt and clinical deterioration; the other is of excessive cardiac enlargement and congestive cardiac failure associated with radiological evidence of pulmonary plethora. No case exactly comparable to the latter occurred in our anastomotic series, though there was a hospital death (therefore not included here) from postoperative pulmonary oedema, apparently the result of an excessively large anastomosis. Among the patients in our series with end-to-side anastomoses there were no apparent examples of pulmonary hypertension; one patient in this series with an end-to-end anastomosis, however, showed obvious pathological evidence of pulmonary hypertension on the side of the anastomosis when examined after death from a second operation (Lopez Bescos, Stevens, and Kirk, 1970). The freedom of our patients from this complication is almost certainly related to the smaller anastomosis fashioned by Lord Brock as compared to that of some other surgeons; the initial few patients in this series who had their operations performed by Blalock nearly all had greater cardiac enlargement and wider arterial pulse pressures after operation than the later cases, and it is clear that he usually fashioned a larger anastomosis.

FIG. 4 *A graphical representation of the quinquennial deterioration, mortality, and reoperation rate in the group of patients who had direct operations. The shaded area represents the patients known to be alive at each 5-year period. The blocks representing the deterioration, mortality, and reoperation rate show the number of patients affected expressed as a percentage of the total number of patients at the start of each quinquennium.*

Deterioration was generally associated with increasing cyanosis and polycythaemia, presumably the result of diminishing pulmonary blood flow; it was not possible in many cases to say whether this was due to increasing obstruction to right ventricular outflow or to narrowing of the anastomosis. In some cases the anastomotic murmur disappeared, though in at least the two instances mentioned above this was related to high blood viscosity from polycythaemia; in others the pulmonary systolic murmur was noted to go. Sabiston *et al.* (1964) have reported the development of

pulmonary atresia in 11 patients after anastomotic operation for Fallot's tetralogy; it seems, therefore, that diminution of pulmonary blood flow can occur from either mechanism. The evidence that increasing right ventricular obstruction may develop in patients with Fallot's tetralogy after an anastomotic operation, or after a Brock operation, as occurred in our patient, M.G., is of considerable interest. A similar process is almost certainly involved in the deterioration which is the rule in the natural history of the condition without operation. Apart from those who had symptomatic deterioration it was notable that many patients, especially during adolescence, developed increased polycythaemia. This in turn was frequently associated with proteinuria; studies of these and other cyanosed patients have shown a significant incidence of impaired renal function apparently related to the polycythaemia (Rennie, 1969). Polycythaemia played a part also in relation to haemorrhage, both spontaneous and at subsequent operations, vascular thrombosis, and symptoms of headache and vertigo. The gross polycythaemia that occurs in some patients with cyanotic heart disease is in itself a major factor in bringing about symptoms, complications, and death.

Bacterial endocarditis occurred in more than a third of the patients surviving 20 years who had had shunt operations as compared with only 1 in 25 who survived 20 years after direct operations; this suggests that the infection in these patients was more often in relation to the anastomosis rather than to the primary anatomical lesions. There was no necropsy proof of this in these patients, but 2 who had bacterial endocarditis developed aneurysms, presumably mycotic, at the site of the anastomosis.

Apart from the lower incidence of bacterial endocarditis it is clear that the patients who had direct operations did better than the anastomotic series in other respects, and the incidence of other non-fatal complications and the mortality rate were less. Unfortunately a valid comparison of the two series cannot be made, as the patients were not randomly assigned to have one operation or the other but were partly selected on the basis of their suitability for each operation; this and the fact that the anastomotic series started first meant that the anastomotic series contained a higher proportion of more severely affected patients. On the other hand, it is unlikely that this factor alone is responsible for the differences: patients who have had a good direct operation are often completely acyanotic (i.e. have normal systemic arterial oxygen

TABLE 8 *Non-fatal complications recorded in patients who had direct operations*

	25 surviving patients	10 patients who died and in 11 surviving until lost to follow-up	Total
Infective	2 (1 SBE) (1 PTB)	3 (2 SBE) (1 PTB)	5
Epilepsy	1		1
Hypertension	1		1
Cardiac arrhythmia	1		1
Complete heart block (post-op. after total correction)	1		1
Totals	6	3	9

SBE = subacute bacterial endocarditis. PTB = pulmonary tuberculosis.

saturation) at rest, at least, and sometimes on exercise as well, whereas the best anastomotic procedure can never attain this. The best direct operation results are better than the best anastomotic procedure results, and are achieved without the production of an anatomical situation which almost certainly exposes the patient to a significant risk of bacterial endocarditis. The better systemic arterial oxygen saturations which can be achieved by the direct operations result in a lesser incidence of the serious polycythaemia which seems to be an important factor in the development of late complications.

There are apparently no reports of strictly comparable series of patients with Fallot's tetralogy treated by the Brock procedures. Flege and Ehrenhaft (1967) reported 16 patients treated in this way, with an additional 9 mentioned in an addendum; of the 16 patients fully reported, 8 had a good result and 6 were improved; later 9 of these had a total correction procedure. Turski and Michalowski (1968) also reported 19 patients who had Brock's operation but gave no follow-up information. Taylor *et al.* (1963) reported the use of the Brock operation in 16 children between 3 months and 3 years of age; there were 3 deaths, and 3 with long narrow infundibular channels obtained only partial relief, but the other 10 had good results. The only large collection of similar patients is contained in a report of the section of cardiovascular surgery of the American College of Chest Physicians compiled and prepared by Rumel (1958); this report summarizes 39 replies to a questionnaire, concerning surgical treatment of Fallot's tetralogy which had been sent to members of the College. Information concerning a total of 4062 patients was collected

but only crude figures for operative results are analysed. No long-term follow-up information is given, but the early results of the anastomotic and the Brock procedures were similar, with an overall 80 per cent of good results in the survivors. Besterman, O'Donnell, and Somerville (1959) reported cardiac failure associated with cardiac enlargement and pulmonary plethora in 7 patients who had had infundibular resection and/or pulmonary valvotomy. These patients, however, differed from those of our series in that the operations were open procedures conducted under hypothermia, and deliberate attempts were made to achieve complete relief of the obstruction to the right ventricular outflow. The closed Brock procedure involves only a stab incision in the right ventricle and inherently was rarely capable of achieving anything approaching complete relief of the outflow obstruction; the only 2 patients in this series in whom this did occur have been briefly described above. The complication described by Besterman *et al.* clearly has much in common with the consequences of a too large anastomosis.

With the current development of the techniques of total correction using heart-lung bypass so as to achieve a satisfactory high proportion of good results with a sufficiently low mortality rate (Gotsman *et al.*, 1969; Kirklin *et al.*, 1965; Zerbini, 1969), palliative procedures, whether anastomotic or direct, now have a smaller part to play in the management of Fallot's tetralogy. To some extent therefore the long-term results of these palliative procedures is less relevant to current practice. The results reported here of these pioneer operations are, however, not only of historical interest; the patients illustrate much of the natural course of cyanotic congenital heart disease, they provide a standard by which other operations can be judged, and there are many such patients still being cared for, as the result of the earlier surgical activities in various centres. Only a relatively modest proportion of our patients have so far been subjected to subsequent total correction procedures. In many patients, especially among those who have had direct operations, the excellence of the result of the original operation has made them disinclined to accept further major heart operations. In the earlier years of open corrective surgery, the high hospital mortality rate in our patients and as reported elsewhere (Kirklin *et al.*, 1965) did not encourage advising patients with good results to have further operations. With more recent experience giving better results (Azar *et al.*, 1969; Goldman, Mustard,

and Trusler, 1968; Gotsman *et al.*, 1969; Zenker and Klinner, 1967), this cause for inhibition has been removed. It is relevant, however, to compare the late results of these early palliative procedures with those so far reported for the so-called 'total correction' procedures. Kirklin *et al.* (1965) report a number of unexplained late deaths and the existence of a number of residual problems after correction. Wolf *et al.* (1965) also report good clinical results but the occurrence of a high percentage of residual abnormalities. Lillehei *et al.* (1964) in a haemodynamic study of 69 patients out of a series of 305 who had correction operations found 15 with residual interventricular shunts, 12 with residual pulmonary stenosis, 16 with pulmonary incompetence, 2 with pulmonary hypertension, and 2 with heart block requiring pacing. Even allowing for the fact that patients with postoperative problems were more likely to be restudied than those who were well, these figures indicate that correction operations are not always 'total'. On the other hand, it must be appreciated that these reports relate to early series and since then techniques have improved, with a consequent reduction in these residual lesions. The possibility of residual lesions as well as the risk of the operation have to be taken into account when considering further operation for a patient with a maintained good result from an initial palliative procedure. We do not yet know whether closure of the ventricular septal defect will protect these patients from the tendency to increasing right ventricular outflow obstruction seen to occur after anastomotic and direct operations in some cases. Psychological and social problems also arise in connexion with advising further operation, when at the time of the original operation it was regarded by the patients or parents as a definitive procedure and where the patients have done well enough to have become parents themselves. Corrective surgery as a second procedure is possibly more hazardous than when done as the primary operation. Bahnson *et al.* (1962) have pointed to the problem of aberrant coronary arteries crossing the right ventricle which may be obscured by adhesions after previous direct operations, and found a high mortality among patients who had had previous end-to-end or bilateral subclavian-pulmonary anastomoses. Kirklin *et al.* (1965) observed a higher mortality among patients who had had prior shunt operations. On the other hand, Zerbini (1969) did not find this in a detailed statistical analysis of the factors affecting the results and mortality of corrective operations; Azar *et al.*

(1969) also observed no significant difference between those with and those without previous shunts. Hypoplastic right ventricular outflow tracts with small pulmonary valve rings and severe polycythaemia are generally recognized to be adverse features; these conditions are likely to develop in the deteriorating patient who has had a shunt operation. There should be no delay in advising further operation in these patients; the high mortality in such patients in our series was probably in large measure due to the fact that a number of them deteriorated too much before operation was offered.

It is not yet possible to compare the 20-year results of corrective surgery with those of the earlier operations. It will be interesting to do so, though there can be no doubt now that despite the risks and the incidence of incomplete repair the proper surgical aim should be to achieve closure of the ventricular septal defect and the greatest possible relief of the right ventricular outflow obstruction. It can only be expected that surgical techniques will continue the evolution, towards obtaining better immediate and long-term results, which has occurred during the 25 years that have passed since the work forming the basis of this study was started.

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Erratum

In 'Correlation of Mortality Rate and Serum Enzymes in Myocardial Infarction. Test of the Efficiency of Coronary Care', by B. L. Chapman (*British Heart Journal*, 1971, **33**, 643), the reference (p. 645) to Chapman, B. L. (1971) should read *Australian and New Zealand Journal of Medicine*, **1**, 53.