

Adult cyanotic congenital heart disease

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Prusty, S. and Ross, D. N. (1975). *Thorax*, 30, 650–656. **Adult cyanotic congenital heart disease.** The surgical treatment of cyanotic heart disease in the adult poses some technical difficulties in correcting severe anatomical deformities and compromised physiological states over a wide range of conditions.

Various abnormalities and their surgical management have been reviewed. Forty-six patients over the age of 18 years have been operated upon with 10 operative deaths. Of the survivors, 63% have had an excellent clinical result; 69.5% of the total group had an excellent or good result following surgery.

It is concluded that the age of the patient is not a bar to the complete repair of these deformities, and all cases of adult cyanotic heart disease should be investigated with a view to surgical correction.

Since 1945 when Blalock and Taussig introduced the subclavian pulmonary anastomosis to palliate Fallot's tetralogy, many other cyanotic conditions with pulmonary stenosis and complicated malformations have been treated by a shunt procedure. Because of the effectiveness of these earlier procedures, cyanotic heart disease is now encountered in an older age group, and, with improvements in technique, it is possible to offer total correction for most types of cyanotic heart disease at any age. Postoperative mortality depends mainly on the severity of the defect, the secondary changes in the myocardium, and the development of pulmonary vascular changes.

The purpose of this paper is to analyse the surgical experience in 46 patients over the age of 18 years with cyanotic heart disease treated at the National Heart Hospital between 1968 and 1973.

FINDINGS

The lesions present are summarized in Table I and the mortality rates are shown in Table II.

TETRALOGY OF FALLOT In the total group of 46 patients there were 30 patients with tetralogy of Fallot aged 18 to 48 years, all disabled with haemoglobin 19–26 g/dl.

Surgical Considerations Of the 30 cases of Fallot's tetralogy (Fig. 1), 16 had a two-stage correction and 14 had a primary total correction (Table III).

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TABLE I
TOTAL NUMBER AND TYPES OF CASES OF ADULT
CYANOTIC HEART DISEASE

Type of Lesion	No. of Cases	Hospital Mortality
Tetralogy of Fallot	30	3
Pulmonary atresia	4	2
Pulmonary stenosis with PFO	3	—
Transposition of great arteries	2	—
Single ventricle	3	3
Endocardial cushion defect	2	—
Common atrium with AV canal	1	1
Ebstein	1	1
Total	46	10 (21.7%)

PFO = persistent foramen ovale.

The previous palliative procedures, Blalock's anastomosis, Pott's anastomosis, Waterston's shunt and Brock's pulmonary valvotomy (Table IV), were closed at total correction.

Total correction was performed using cardiopulmonary bypass with haemodilution and normothermic perfusion at 2.4 litres per square metre. In five patients a patch of autogenous pericardium or Dacron was used to enlarge the right ventricular outflow tract, and in another seven patients with severe tetralogy, almost amounting to atresia of the outflow tract and pulmonary trunk, a fresh homograft conduit was used to reconstruct the outflow tract (Ross and Somerville, 1966). Where there were unusually large collaterals or a difficult shunt, circulatory arrest with hypothermia was used.

RESULTS

Three of the 16 who had a two-stage correction and two of the 14 who had a total correction died. Of the

TABLE II
RESULTS OF SURGICAL REPAIR

Type of Lesion	Result			
	Excellent	Good	Fair	Hospital Death
Tetralogy of Fallot	21	3	1	3
Pulmonary atresia	1	—	1	2
Pulmonary stenosis with PFO	3	—	—	—
Transposition of great arteries	2	—	—	—
Single ventricle	—	—	—	3
Endocardial cushion defect	2	—	—	—
Common atrium with AV canal	—	—	—	1
Ebstein	—	—	—	1
Total	29 (63%)	3 (6.5%)	2 (4.3%)	10 (21.7%)

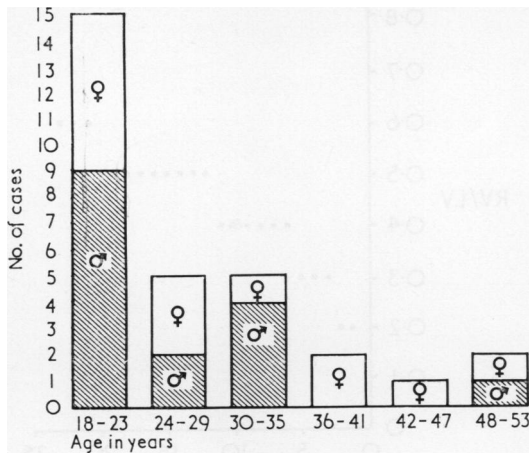


FIG. 1. Age and sex distribution of the 30 cases of Fallot's tetralogy: youngest 18 yr, oldest 48 yr.

TABLE III
TOTAL NUMBER OF CASES OF FALLOT'S TETRALOGY TREATED

Type of Operation	No. of Cases	Mortality	
		Early	Late
Two-stage correction	16	2	1
One-stage correction	14	1	1
Total	30	3 (10%)	2 (6.6%)

TABLE IV
NUMBER AND TYPE OF PALLIATIVE PROCEDURE ON 30 CASES OF FALLOT'S TETRALOGY

Palliative Procedure	Number
Ballock anastomosis	8
Pott's anastomosis	4
Waterston shunt	3
Pulmonary valvotomy	2
Total	17

five, three died immediately postoperatively, and the other two after discharge from hospital (Table III).

Postoperative death was due to raised pulmonary vascular resistance. Late deaths in two were sudden at an interval of two years and four months following surgery. One of these had aortic valve replacement with a Starr ball valve prosthesis and mitral valve replacement with Björk valve for bacterial endocarditis along with total correction of tetralogy of Fallot at the age of 48 years.

Analysing the mortality further, it was found that only 2 of the 18 without an outflow patch died, while three of the 12 with outflow tract reconstruction with patch or homograft conduit died (Table V). This may be due to the severity of the disease in the latter group with increased bypass time.

Of the 25 survivors, 21 (84%) had an excellent haemodynamic result and three (12%) had a good result with some restriction of activities (Table V). Twelve survivors of the one-stage correction had excellent results while only nine of the 13 undergoing the two-stage correction had an excellent result (Table VI).

Analysis of the RV/LV peak systolic pressures throws light on the survival prospects for these patients. Of the 25 in whom this was measured (Figs 2 and 3), the majority had a ratio of 0.5 or less. The RV/LV peak systolic pressure is seen to be a useful guide to patient survival postoperatively.

PULMONARY ATRESIA This difficult group of patients survive to adult life only because of palliative procedures performed earlier or because of extensive collaterals or both. The majority are severely disabled and are treated early on in life.

Four patients had pulmonary atresia with previous palliative shunts and were reconstructed with homografts. One death was due to postoperative lung problems as a consequence of increased pulmonary vascular resistance and the other patient died of uncontrollable bleeding from the Dacron conduit.

TABLE V
ANALYSIS OF RESULTS OF TOTAL CORRECTION OF FALLOT'S TETRALOGY IN 30 CASES AND COMPARISON BETWEEN DIFFERENT PROCEDURES

Type of Procedure	No. of Cases	Result			
		Excellent	Good	Fair	Death
No outflow patch	18	13	3	—	2
With outflow patch	5	3	—	1	1
Homograft conduit for RV outflow reconstruction	7	5	—	—	2
Total	30	21 (70%)	3 (10%)	1 (3.4%)	5 (16.6%)

TABLE VI
COMPARISON OF RESULTS OF TWO-STAGE AND ONE-STAGE CORRECTION OF FALLOT'S TETRALOGY IN 30 CASES

Type of Procedure	Result		
	Excellent	Good	Fair
Two-stage correction	9	3	1
One-stage correction	12	—	—
	21 (70%)	3 (10%)	1 (3%)

TABLE VII
ANALYSIS OF RESULTS OF SURGERY OF DIFFERENT TYPES OF PULMONARY ATRESIA

Type	No.	Result			
		Excellent	Good	Fair	Death
I	2	1	—	—	1
II	1	—	—	1	—
III	1	—	—	—	1
Total	4	1	—	1	2

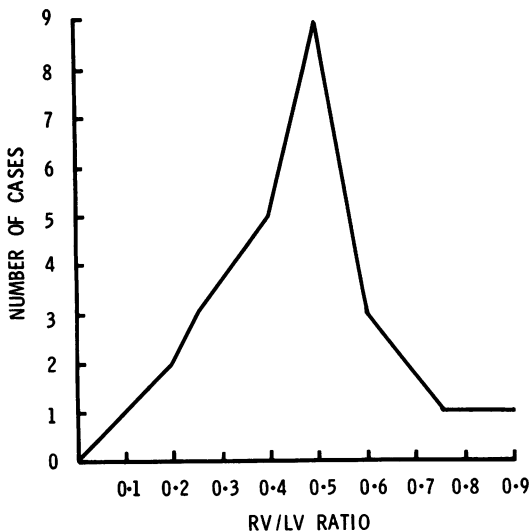


FIG. 2. Curve showing the number of patients in relation to the RV/LV systolic pressure ratio.

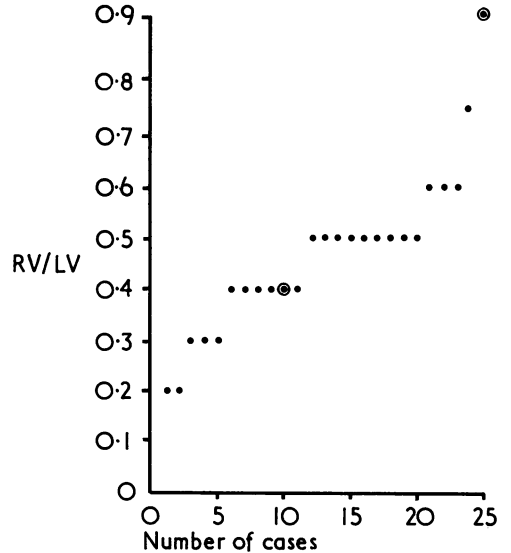


FIG. 3. Actual number of cases in different RV/LV ratios and their fate. Each dot represents one patient.

PULMONARY STENOSIS WITH PERSISTENT FORAMEN OVALE All three patients who had relief of pulmonary stenosis and closure of the persistent foramen ovale survived with an excellent result.

TRANSPOSITION OF THE GREAT ARTERIES Two patients with this anomaly survived and achieved an excellent result after surgical correction by a right ventricular conduit reconstruction (Rastelli, Wallace and Ongley, 1969).

SINGLE VENTRICLE In the two patients with single ventricle and a common atrioventricular valve, an attempt was made to close the atrial septum and redirect the right atrial blood to the pulmonary artery by a Fontan-type of procedure (Fontan and Baudet, 1971). Both died in a low output state.

EBSTEIN'S ANOMALY One adult patient was treated by means of a frame-mounted homograft replacement of the tricuspid valve but died in a low output state and with uncontrollable arrhythmias.

DISCUSSION

In general, adults with cyanotic heart disease present with similar conditions to children, allowing for the fact that some children with severe cyanotic disease will not live to adult life. Those with common conditions such as Fallot's tetralogy have died (if untreated) at adolescence often as a result of infective intracerebral lesions. In fact cerebral abscess was until recently one of the common causes of death in Fallot in children and young adults.

As a result of the thousands of palliative operations for Fallot (usually Blalock shunts, but also the Brock operation) we now have a number of patients presenting in adult life who have survived to this age as a result of palliation in childhood.

We can expect to have more congenital cyanotic heart disease presenting in adult life as a consequence of the palliative and so-called curative operations now being performed. These patients, if they live into adult life, may present with complications resulting from their corrective procedure (the Mustard operation) rather than with cyanosis.

An important group of patients with cyanotic heart disease in adults are those with a reversed or reversing shunt as a result of increasing pulmonary vascular resistance (Eisenmenger syndrome), or with conditions such as single ventricle or the complex forms of transposition. These patients, for whom no established surgical treatment is at present available, often suffer from severe headaches and haemoptysis as a result of their polycythaemia. It has been said that patients with a reversed ductus are less disabled than those with a reversed ventricular septal defect or atrial septal defect since the reversing desaturated blood enters the aorta distal to the cerebral vessels, and this fact can be offered as some comfort to the patient. Otherwise we are dependent on the development of lung transplantation to achieve correction in the Eisenmenger group by replacing one or both lungs with lungs of normal vascular resistance.

From a surgeon's point of view, patients with cyanotic heart disease presenting in adult life are treated along the same general lines as a child but there are practical differences and problems which combine to increase the operative risk.

The heart muscle which has been supplied with partially desaturated viscous blood has often been contracting against an obstructive lesion for years and no longer resembles healthy myocardium. This

is particularly true of the right ventricle. Surgical manoeuvres are consequently technically more difficult and hazardous. Also postoperative low-output states are likely to be more intractable, and residual pulmonary regurgitation after Fallot correction is less well tolerated.

Since the blood supply to the lungs is often through large bronchial and systemic collateral vessels, haemorrhage from these will be considerable throughout any operative procedure. Also the systemic collaterals may be a cause of increased pulmonary vascular resistance and persistently raised right-sided pressures.

Polycythaemia in adult cyanotic heart disease is generally severe and, apart from the symptoms already described, there is an increased tendency to bleeding after surgery and the blood loss can be massive. The exact cause of the bleeding is unknown, but is possibly related in some way to impaired thromboplastin generation (Somerville, McDonald, and Edgill, 1965). The degree of impaired thromboplastin generation is related to the degree of polycythaemia and increase in haemoglobin.

The most useful measures to control this bleeding are to ensure that these patients have additional vitamin K preoperatively and to have adequate amounts of absolutely fresh blood and frozen plasma available for the postoperative period. Some advocate bleeding patients preoperatively to reduce the polycythaemia but, in our experience, this has always made the sick patient worse and has increased the preoperative risk.

Adult cyanotic patients usually have some degree of impaired renal function with albuminuria, reduced creatine clearance, and raised blood uric acid. Since the blood entering the renal capillaries may be made up of 70% cells and then loses a great deal of its liquid content through the glomeruli, it is difficult to understand how the kidneys function at all. However, in spite of our initial misgivings renal failure has not been a major postoperative problem.

These patients often have distressing and intractable infected acne, which constitutes a definite surgical risk factor which may be reduced with an antiseptic skin preparation and culture of any pustules for bacterial sensitivities.

As a consequence of these added difficulties, in adult cyanotic disease surgery can be technically demanding.

TETRALOGY OF FALLOT In the reported series of congenital heart disease in the adult, tetralogy of Fallot has been the most frequent cyanotic lesion (Fisher, Wilson, and Theilen, 1962; Cooley, Hallman

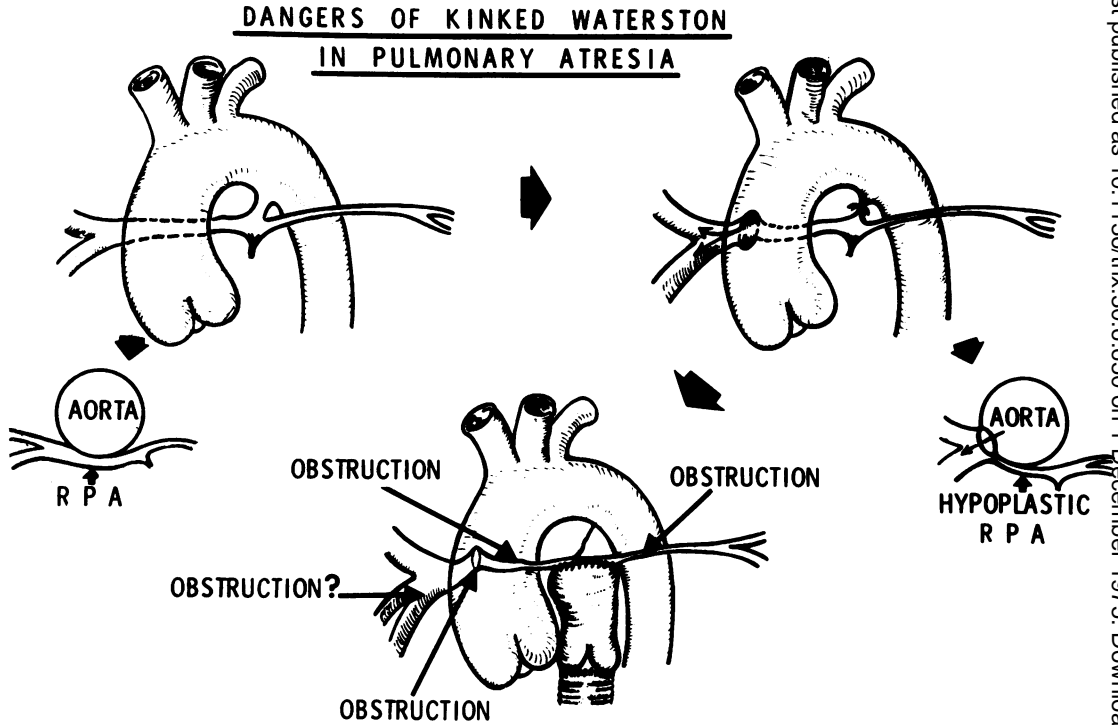


FIG. 4. *The complications of Waterston's shunt.*

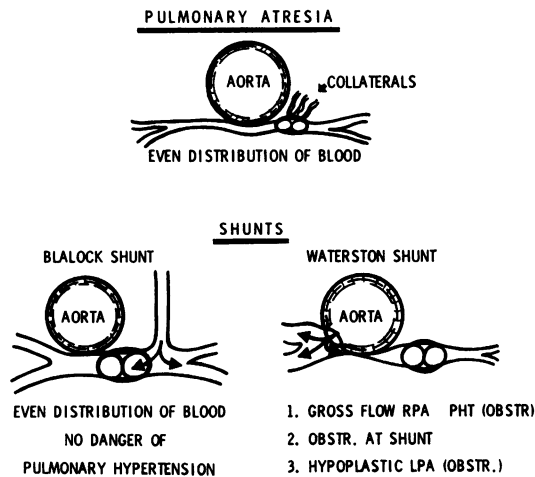


FIG. 5. *Comparison of blood distribution to both lungs between Blalock shunts and Waterston shunts.*

and Hammam, 1966; Ochsner, Jordan, and Moore, 1967; Leeds, 1958). There have been reports of the successful total correction of this lesion in adolescents and adults (Bender *et al.*, 1971; Higgins and Mulder, 1972). The basic factor in the surgical management of this lesion depends upon the anatomy of the right ventricular outflow tract and the pulmonary artery and the condition of the myocardium of the right ventricle. The effects of the obstruction may be more troublesome than in a child with a compliant right ventricle.

The RV/LV systolic ratio is a good practical guide to the adequacy of the relief of right ventricular obstruction (Kirklin and Karp, 1970). Where possible a patch in the outflow tract is avoided but a certain degree of pulmonary regurgitation is better tolerated than a significant residual outflow tract obstruction (Kirklin *et al.*, 1965; Ken-ichi and Eguchi, 1970). However, in order to reduce the pulmonary insufficiency a valve-retaining patch graft reconstruction has been used with encouraging results (Eguchi and Asano, 1968; Kaplan *et al.*, 1973).

There remains another group of patients with tetralogy of Fallot in whom the outflow tract is severely hypoplastic and can only be adequately dealt with by means of a valved conduit (Ross and Somerville, 1966). Since 1966 there have been several reports on the successful use of such homograft conduits (Weldon, Rowe and Gott, 1968; Bender *et al.*, 1971). Seven patients in our series of 30 with tetralogy of Fallot required outflow tract reconstruction with the use of aortic homograft conduits.

The three early deaths in this series of 30 patients is acceptable when compared with other reports in the literature (Bender *et al.*, 1971). The mortality in this small series does not seem to have been influenced by the two-stage or one-stage correction, and our inclination is to recommend one-stage correction where possible. Of the survivors, 96% have had an excellent or good clinical result.

PULMONARY ATRESIA This is a serious type of cyanotic anomaly, and surgical correction entails closure of the ventricular septal defect and reconstruction of the pulmonary outflow tract, and sometimes the management of collaterals. Such a lesion was first corrected with a homograft conduit in 1966 (Ross and Somerville). In this series there were four patients with type I, type II, and type III pulmonary atresia (Somerville, 1970). One patient died, probably as a result of changes in the pulmonary vasculature causing persistent right ventricular hypertension, and another from uncontrollable bleeding from the Dacron conduit. The management of this disease

is difficult. It usually presents in early childhood with severe cyanotic disease, and some form of aortopulmonary anastomosis is needed—a Waterston anastomosis in our series. This has the disadvantage, however, of causing kinking and obstruction of the right pulmonary artery and subsequent difficulties when the reconstructive procedure (Fig. 4) is done. It is consequently our current belief that a left Blalock or similar shunt procedure is preferable as it is more likely to perfuse both pulmonary arteries (Fig. 5).

At the corrective procedure it is an advantage to use deep hypothermia and, if necessary, circulatory arrest to deal with the heavy collateral blood flow. It is also necessary to divide the large collaterals usually arising from the descending aorta and this makes surgical access difficult. One technique is to control the collaterals through a left thoracotomy and then to perform a transsternal correction (Kirklin *et al.*, 1973).

TRANSPOSITION OF THE GREAT ARTERIES Rastelli *et al.* (1969) reported the first successful repair of transposition of the great arteries with ventricular septal defect and pulmonary stenosis in a 14½-year-old boy. Since then this procedure has been well standardized (McGoon, Wallace, and Danielson, 1973).

SINGLE VENTRICLE This still remains a complex embryological and anatomical defect. The recent literature does show promising results of successful repair of such lesions where there are two AV valves (Sakakibura *et al.*, 1972; Edie *et al.*, 1973). In our two unsuccessful cases with a common AV valve, we have tried to isolate the right atrium and turn it into a pumping chamber as in the Fontan repair of tricuspid atresia (Fontan and Baudet, 1971).

EBSTEIN'S ANOMALY This has been successfully treated in recent years with replacement prosthetic valves (Kitamura *et al.*, 1971). We feel that tricuspid valve replacement with a frame-mounted aortic homograft would work efficiently in this low pressure chamber without any danger of valve dysfunction or embolization.

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