Surgery for Cyanotic Heart Disease in the First Year of Life

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Data are reviewed on 248 patients less than 1 year old who presented with a diagnosis of cyanotic heart disease between January 1976 and January 1982. No infant had had prior surgical treatment. The patients were classified according to diagnosis: tetralogy of Fallot, transposition of the great arteries, pulmonary atresia and anomalies of the tricuspid atresia or single ventricle type. Other remote forms of cyanotic heart disease were excluded from the analysis. Management of these patient groups is discussed in relation to their potential for corrective surgery early in infancy or later. The proper selection of palliative procedures that will permit bilateral growth and development of pulmonary arteries and equal distribution of pulmonary blood flow is emphasized. Morbidity and mortality in each patient group are discussed.

The report of Blalock and Taussig (1) in 1944 of the creation of a shunt between the subclavian and the pulmonary artery changed the clinical approach to children with cyanotic heart disease. The ability to decrease cyanosis by increasing pulmonary flow has extended life and improved its quality immensely. Since that report, other types of shunts have been reported, and their successes and complications documented (2–4).

Unfortunately, the complications of the shunts, such as pulmonary hypertension from excess flow, kinking or obstruction of the pulmonary artery or failure of the shunt, may adversely affect future corrective surgery. Thus, in some instances it may be preferable to perform the corrective procedure as early as possible to reduce or eliminate the complications of a palliative procedure. The question of early corrective surgery is still being discussed and no unanimity of opinion has been reached (5,6). It is recognized that for early corrective surgery to be accepted, the operative mortality in infancy must not exceed the combined mortality of the palliative and subsequent corrective operations.

The following report relates to the experience of patients in the first year of life presenting for the initial treatment of cyanotic heart disease.

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Study Group

Two hundred forty-eight infants less than 1 year old were admitted to the University of California, San Francisco Medical Center between January 1976 and January 1982 for consideration of some form of surgical therapy for cyanotic heart disease. At the time of admission, no infant had received prior surgical treatment. Table 1 shows the age range of the patients. History, physical examination, routine laboratory blood analyses and an electrocardiogram were performed in all patients; chest X-ray films and echocardiograms were also obtained. The diagnosis of cyanotic heart disease was confirmed at cardiac catherterization in 244 of the 248 patients. In four patients in extremis taken directly to surgery, the diagnosis was based on the combination of the other procedures with the exception of cardiac catheterization.

Table 2 classifies the general condition of the patients at the time of admission. The majority of the patients had clinical evidence of cyanosis. Only 22 were considered to be in extremis, requiring intubation, administration of prostaglandins or other cardiotonic agents. One hundred ninety-two were considered to be in stable condition in that they were resting easily in oxygen without requiring assisted ventilation, cardiotonic agents or other support measures even though they may have been intensely cyanotic. The surgical approach to the patients will be discussed according to their general anatomic classification.

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Table 1. Age Distribution of the 248 Patients With Cyanotic Heart Disease Presenting in the First Year of Life

Age (days)	Number of Patients
0 to 30	68
30 to 90	79
90 to 180	42
180 to 360	59

Surgical Procedures and Results

Tetralogy of Fallot

Sixty-two infants were considered to have a form of tetralogy of Fallot. In these patients, the pulmonary artery was in continuity with the right ventricle and, during angiography, some amount of contrast material could be seen moving from the right ventricle into the pulmonary artery even though the degree of pulmonary stenosis may have been severe. The type of surgical approach to each patient was determined primarily by the size of the pulmonary arteries. If the right pulmonary artery in its crossing beneath the aorta was considered to be one-third or greater than the diameter of the aorta, a form of corrective surgery was recommended.

Corrective surgery. Forty-eight patients were judged to have pulmonary arteries of adequate size to permit a corrective operation at the time of the initial surgery. There was one death in this group of 48 patients. The type of outflow reconstruction is shown in Table 3. In general, when a right ventricular outflow patch was used, the patch was made as narrow as possible to evoke the smallest amount of enlargement of the right ventricular outflow tract. When the patch was carried across the anulus, an attempt was made to divide the usual bicuspid pulmonary valve at its anterior commissure and widen its anulus the least possible amount to effectively lower right ventricular pressure to one-half of the systemic pressure. Pericardial tissue was used for the patch in 31 patients and synthetic material was used in 10.

In three patients, the anterior descending coronary artery arose from the right coronary artery and traversed the right ventricular outflow tract. Thus, the usual linear incision could not be made through the outflow tract. In these instances, an incision was made distal to the coronary artery by incising into the pulmonary artery. Valve stenosis was relieved and if the pulmonary anulus required enlargement, a patch of pericardial tissue was extended from the pulmonary artery across the anulus as far proximal as possible without damaging the abnormal coronary artery. An incision was made in the body of the ventricle, and resection of muscle and closure of the ventricular septal defect were accomplished from beneath the area of the coronary artery. One of these patients required reoperation 18 months later for residual pulmonary stenosis. A second patient with normal coronary anatomy also required reoperation for residual pulmonary stenosis 1 year after corrective surgery. There were no instances of heart block associated with these repairs.

Table 2. Status of the 248 Infants at Time of Admission to the Hospital

	Number of Patients
In extremis	22
Intubated	16
On drug therapy	
Prostaglandin B_1	41
Cardiotonic	12
Stable condition	192

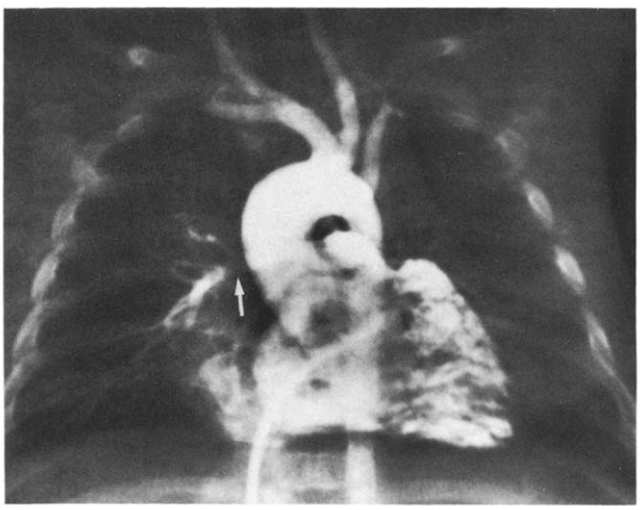
Table 3. Type of Right Ventricular Outflow Tract Operation in 62 Patients With Tetralogy of Fallot

	Number of Patients
Correction	48
Patch	41
Across pulmonary anulus	24
No patch	7
Palliation	
RVOT patch only	14

RVOT = right ventricular outflow tract.

Initial palliative surgery. In 14 patients (Fig. 1), the pulmonary arteries were judged inadequate to accept total right heart efflux, and a right ventricular outflow patch was constructed across the pulmonary anulus to the bifurcation of the pulmonary arteries. There were two early deaths in this group due to inadequate pulmonary blood flow and persistent cyanosis. In both cases, the pulmonary arteries

Figure 1. Right ventriculogram of a 3 day old infant with severe tetralogy of Fallot. The size of the pulmonary arteries is quite small, and the right pulmonary artery just proximal to the **arrow** is clearly less than one-third the diameter of the aorta. This infant was treated with a right ventricular outflow tract patch and later underwent closure of the ventricular septal defect.



were less than 20% of the diameter of the ascending aorta. Six of the 14 patients required subsequent reoperation for modification or attempted correction. Two of the six later died from their second operation. In these two, the pulmonary artery was still small and an attempt to connect a valved conduit to the right and left pulmonary arteries was not tolerated. The four successful corrective operations were in patients in whom good pulmonary artery growth had occurred and the right pulmonary artery/aorta ratio was above one-third at the time of the corrective procedure.

Tricuspid Atresia and Single Ventricle With Diminished Pulmonary Blood Flow

Twenty-eight patients had a form of tricuspid atresia or single ventricle with pulmonary stenosis or diminished pulmonary blood flow. All 28 presented with cyanosis. It was considered that no direct corrective procedure was possible during infancy in these patients and that most would be a candidate for corrective surgery of the Fontan type in the future.

Enlargement of ventricular septal defect with or without pulmonary valvotomy. Twelve patients who had a rudimentary chamber attached to an anterior pulmonary artery that received blood by way of a ventricular septal defect from the main ventricle (type 1B tricuspid atresia without transposition) underwent enlargement of the ventricular septal defect with or without pulmonary valvotomy as the initial palliative procedure (Fig. 2). These 12 patients were placed on cardiopulmonary bypass and a small incision was made in the rudimentary chamber attached to the pulmonary artery. The pulmonary valve was incised, if stenotic, and the ventricular septal defect was enlarged on the left superior aspect of the defect. A small piece of pericardial tissue was used to close the ventriculotomy. One death occurred in the 12 infants. The patient was in extremis and showed no improvement in the immediate postoperative period. Even though oxygenation was better, metabolic acidosis was not reversed and this infant died 6 hours after surgery. There were no instances of heart block. One patient had increased pulmonary flow requiring treatment with diuretic drugs and digitalis. Four of these patients subsequently underwent a successful Fontan type modification.

Central shunt between aorta and pulmonary artery. In the remaining 16 patients in this group the pulmonary artery was posterior or no rudimentary chamber was attached directly to it. The primary goal in these patients was to create a central shunt between the aorta and the main pulmonary artery. The central shunt was accomplished in 12 patients. In 10, the anastomosis was directly between the aorta and the main pulmonary artery. In three of these, a Gortex graft was used to connect the main pulmonary artery to the aorta. In two patients, a Waterston type anastomosis was made, and a Blalock-Taussig anastomosis was made in one patient. In all 28 of these infants, the main attempt was to increase flow into the main pulmonary artery to ensure bilateral growth of the pulmonary vessels.

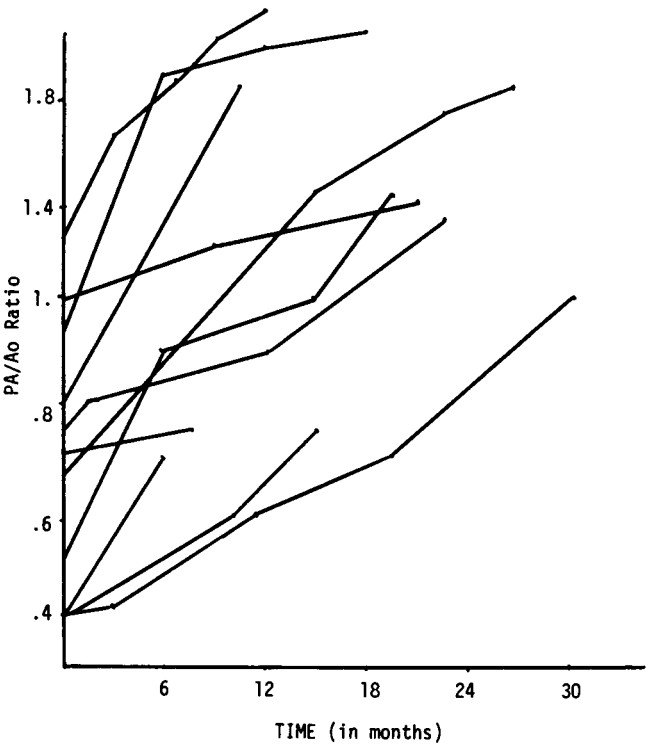


Figure 2. Pulmonary artery growth in tricuspid atresia. Pulmonary artery to aorta ratio (PA/Ao) in the 11 living patients who underwent enlargement of the ventricular septal defect as a method of palliation of tricuspid atresia. Growth has been documented by echocardiography and cardiac catheterization.

Transposition of the Great Arteries

Total correction in simple transposition. One hundred thirty-four patients presented with a form of transposition of the great arteries. Of these, 82 were believed to have simple transposition with no, or only a small, ventricular defect that did not require operative repair. Total correction was accomplished in these by using either a Mustard or a Senning type repair. Profound hypothermia with total circulatory arrest was used in 61 patients, and standard cardiopulmonary bypass with cannulation of each vena cava was employed in 21 patients. One of these 82 patients underwent Blalock-Hanlon atrial septectomy followed 1 week later by a Mustard repair. This patient died as did one other patient with a very small left atrium and probable small mitral valve (Table 4).

Total correction in complete transposition. Thirty-four patients presented with a complex form of transposition of the great arteries in which either pulmonary stenosis, ventricular septal defect or pulmonary atresia was present. All 34 patients underwent a corrective form of operation and 28 survived. The ventricular septal defect was closed or pulmonary stenosis was relieved, or both, in conjunction

Table 4. Type of Transposition of the Great Arteries in 134 Patients Presenting in the First Year of Life

	Number of Patients	Operative Deaths
Simple	82	2
Complex		
Correction	34	6
Palliation	18	2

with the atrial baffle procedure. The ventricular septal defect was closed through the tricuspid valve and the subpulmonary stenosis resected through the pulmonary anulus.

Palliative shunt procedure in complex transposition. Eighteen patients were considered unsuitable for corrective procedures during infancy. These patients had pulmonary atresia, severe pulmonary stenosis, small-sized pulmonary arteries or multiple ventricular septal defects associated with diminished pulmonary blood flow. A shunt procedure was performed in all 18. There were two deaths associated with the operative shunt relating to continued cyanosis unrelieved by attempts to increase pulmonary blood flow. In both cases, the patients had small, almost diminutive, pulmonary arteries. Placement of a large (5 to 6 mm) Gortex graft between the pulmonary artery and aorta failed to change the degree of cyanosis, suggesting pulmonary vascular disease and diminutive arteries.

Pulmonary Atresia

Thirty-six patients presented with some form of pulmonary atresia in which no continuity between the transverse pulmonary artery and the heart was noted. Eight of these patients were considered to have good-sized pulmonary arteries and underwent a shunt procedure without mortality. In 16, the pulmonary artery size as estimated by angiogram was intermediate and the shunt was accomplished with one death (Table 5). In general, a Gortex shunt was placed between the bifurcation point of the pulmonary arteries and the ascending aorta. In the small infants, a 4 mm shunt was used and in the older infants, a 5 to 6 mm shunt was accomplished. In 12 patients, the pulmonary artery size was extremely small, and 2 of the 12 died after a shunt procedure. In these infants even though the shunt was patent, the pulmonary artery size was so small that increasing pulmonary flow was not accomplished. No breakdown of the type of intracardiac anomaly in this group of patients with pulmonary atresia was presented because this did not enter into the decision for the type of palliative procedure attempted.

Table 5. Pulmonary Artery Size as Estimated by Angiogram in 36 Infants With Pulmonary Atresia

	Number of Patients	Operative Deaths
Good	8	0
Intermediate	16	1
Small	12	2

Effect of shunting procedure on pulmonary arteries. All eight patients with good-sized pulmonary arteries have shown progressive enlargement in the size of their pulmonary vessels, and three have undergone successful Rastelli type corrections. None of the 10 patients surviving the shunting with so-called small pulmonary arteries have shown progressive growth in these vessels although the shunt is patent and has functioned adequately to allow the child to survive at least 18 months after the initial operation.

Discussion

Corrective surgery in early infancy. With improvements in overall clinical care of infants, early recognition of those with cyanotic heart disease is becoming more common. Thus, our diagnostic and therapeutic regimens are directed to a greater proportion of patients in the first month of life than previously (7,8). Obviously, many infants still die after early recognition and diagnosis of congenital heart disease because of improper planning and poor selection of operative procedures; therefore, reassessment of proper therapy needs to be considered in the management of all infants with cyanotic heart conditions. Mortality cannot be the only standard for comparing palliative procedures with corrective operations. Initial therapy cannot be considered successful if an infant survives and shows improvement with a palliative operation, but is left with a damaged or hypoplastic pulmonary artery that cannot be reconstructed at a second operation (Fig. 3). From our experience, we have concluded that age alone does not determine survival when corrective procedures are employed. It should be possible to use cardiopulmonary bypass or ancillary techniques, such as circulatory arrest or hypothermia, with the same success irrespective of the patients' size and age. Clearly, the only determinant over which the surgeon has no influence is the basic anatomic configuration or genetic disposition of the anomaly.

Selection of corrective or palliative surgical procedure. It seems clear that many infants born with hypoplastic or small pulmonary arteries may never have these arteries enlarged irrespective of the therapeutic approach. Possibly repeated balloon dilation may have a role in these patients with small pulmonary arteries. The attempt to cause the most blood to flow uniformly to both pulmonary arteries should be an important goal at selection of the initial palliative procedure. This is especially important in patients with forms of congenital heart disease that are now considered uncorrectable. Thus, many infants with anomalies of the single ventricle, tricuspid atresia type must become candidates for a Fontan type of separation of the circulation if

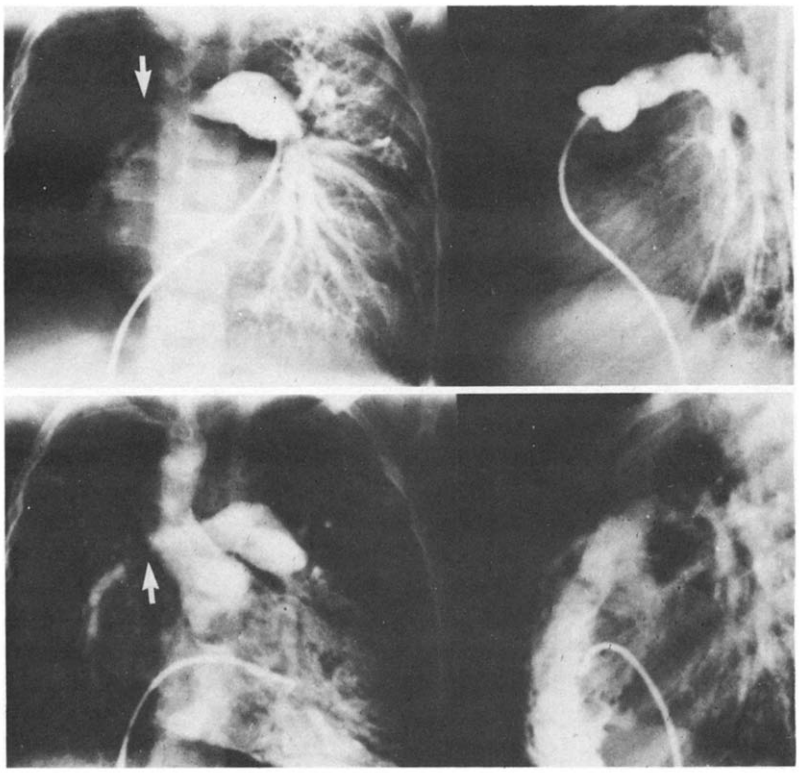


Figure 3. Two patients who had a previous Waterston shunt as palliation for tetralogy of Fallot in infancy. Note the marked stenosis and narrowing of the right pulmonary artery at the point of the **arrows** in each case. Not only has the left pulmonary artery grown in a normal fashion, but the right pulmonary artery seems to have been permanently damaged by the Waterston anastomosis. **Upper panel**: anteroposterior (left) and lateral (right) pulmonary angiograms. **Lower panel**: anteroposterior and lateral right ventricular angiograms.

any technique to eliminate intracardiac shunting is ultimately to be employed. To be a candidate for a Fontan procedure, one must have adequate pulmonary arteries with low vascular resistance. It is preferable if the distribution of flow is to both lungs rather than to a single lung. Potentially a Blalock-Taussig, Waterston or Potts type of shunt may lead to unilateral enlargement or scarring and kinking of one or the other branch pulmonary arteries. Shunting into the main pulmonary artery itself, or at least to the bifurcation, offers the theoretical advantage that if the arteries grow, they may grow bilaterally and with more uniformity.

Total repair of tetralogy of Fallot. In attempting total repair of tetralogy of Fallot in infancy, one must be able to obtain a mortality rate comparable to that achieved with simple palliative operations. There seems to be no distinct disadvantage to total correction if comparable mortality values can be obtained. A small number of patients most likely will have recurrent pulmonary stenosis irrespective of the type of repair because, in the small infant, the intense infundibular muscle accumulation often has not yet occurred. In our own experience, one patient clearly did not have muscle bands or unusual muscular connections in the outflow tract at the time of initial repair. A low pressure was present in the right ventricle and he was considered to have had a good surgical result. Approximately 18 months later, he was reoperated on for recurrent pulmonary stenosis. The recurrence was probably due to well developed muscle accumulations that were part of genetic predisposition of the lesion rather than a result of failure to divide these accumulations at the time of the initial procedure. The frequency of this occurrence does not seem great enough to recommend either alternate approaches or more extensive patching of the outflow tract. A second intracardiac operation does not appear to carry a greater risk of potential damaging effect to the heart or its pulmonary circulation than an initial palliative shunt procedure plus a corrective operation.

Total versus palliative surgery for transposition. The results reported in the treatment of transposition of the great arteries in early infancy have been extremely gratifying, and continued improvement in precision in surgical technique will likely make these results even better with time (9–11). Instances of arrhythmia seem to be diminishing and the long-term functional state of the right ventricle remains satisfactory. Therefore, palliative procedures such as operative atrial septectomy do not appear to be indicated in the treatment of the simple forms of transposition. The success of the Rashkind septostomy is excellent. Only the group of patients with complicated transposition and pulmonary stenosis or hypoplastic pulmonary arteries remain candidates for some type of palliative operation. Even in this group, growth of the pulmonary arteries is necessary for potential long-term success. Pulmonary valvotomy has had varying success because it is often difficult or impossible to resect the fibrotic subvalvular stenosis that may accompany transposition with either a hypoplastic anulus or extensive subvalvular fibrous accummulations. Because many in this group

may be candidates for a Rastelli approach, a resection of infundibular pulmonary stenosis is not necessary for a good result with the Rastelli operation.

It is clear that in these patients, operative mortality will continue to diminish. However, proper planning and selection of operative procedures that result in bilateral growth of pulmonary vessels and avoidance of kinking and injury to the pulmonary tree should ensure a more optimistic long-term outlook for these infants. It is no longer feasible to speak only of mortality in conjunction with the management of infants with cyanotic heart disease. One must plan from the time of initial diagnosis to obtain the optimal long-term results because many of these infants with complicated lesions must be candidates for a procedure in which blood is pumped by either the right atrium or venous pressure through the lungs. Thus, the selection of the palliative operation becomes the most important decision in determining the long-term prognosis.

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