Current Considerations in the Surgical Management of Tetralogy of Fallot*

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Tetralogy of Fallot has been treated by aortic-pulmonary anastomosis, and by total correction utilizing deep hypothermia. Total open heart correction is currently performed utilizing total cardiopulmonary bypass. More recently there has been a trend toward early total open heart correction in small infants and neonates utilizing deep hypothermia and circulatory arrest, and away from the palliative anastomotic operations in these infants. The critical role of the degree of anatomic abnormality of the right ventricular outflow tract, the place and optimal age for complicated outflow tract reconstruction, and the relative merits of the single stage early correction versus the two-stage procedure (early systemic-pulmonary anastomosis and later total correction) are reviewed in the light of current experience.

Tetralogy of Fallot was among the first cyanosis-producing lesions to be treated by aortic-pulmonary anastomosis.1 It was the first cyanosis-producing lesion to be successfully treated by open heart repair under hypothermia and inflow occlusion,2 with cardiopulmonary support utilizing cross-circulation,3 and total cardiopulmonary bypass.4 The recent revival of interest in deep hypothermia and circulatory arrest in the management of small infants with this condition has brought us full circle.

Over these 27 years, a number of new palliative procedures have been advocated, new techniques for total correction of complicated lesions have been developed, and the age of patients at operation has steadily declined. Meanwhile longterm follow-up data on patients surviving palliative operations, total correction, or both as staged procedures are becoming available. As this experience is ongoing and dynamic, disagreement exists in certain areas. It is important, therefore, to examine the surgical alternatives and their expected accomplishments in perspective, in an effort to reduce some of the confusion arising from so many separate and conflicting reports.

Natural History

Patency of the ductus arteriosus may prevent severe arterial hypoxemia, but in almost every case the ductus closes in the early weeks of life. Collateral channels to the pulmonary circulation via bronchial or other systemic arteries may develop and be responsible for improvement in some children.6,7 They are particularly prominent in patients surviving to adulthood.8 But they are of little help in most patients, the usual patient experiencing progressive cyanosis and polycythemia, as outflow tract obstruction increases9 and thrombosis ensues in the pulmonary vascular bed.10 The average age

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at death without surgical treatment is approximately 12 years.8

These considerations aside, there is no question that the severity of hypoxemia and the resultant symptoms, the ultimate of which are cyanotic "spells," are directly related to the severity of right ventricular outflow tract obstruction. There is also no doubt that those children having symptoms earliest in life bear the most distorted outflow tract anatomy. Forty-two8 to 50 percent11 of infants with symptoms in the first six months of life have complete outflow tract obstruction (pseudotruncus aorticus). This fact is important as we consider the role of palliative versus totally corrective operations in neonates and infants.

**INDICATIONS FOR SURGICAL TREATMENT**

Cyanotic spells are a pressing indication for surgical treatment. (Propranolol therapy may be efficacious in decreasing spells, but it has little effect on fixed obstruction at the infundibular or valvular level, and may even be hazardous.)

Growth failure, fatigueability, dyspnea, increasing polycythemia and increasing squatting are indications of progressive arterial oxygen unsaturation and indicate the necessity for surgical intervention.

Finally, the diagnosis of tetralogy of Fallot in itself is an indication, ultimately, for surgical treatment in childhood,12 since the hemodynamic liabilities increase with age,5,13 and the prognosis without treatment is poor.8

**PALLIATIVE OPERATIVE PROCEDURES**

*The Brock Procedure*

Closed heart transventricular widening of the pulmonary valve and infundibulum offers palliation to some children with tetralogy of Fallot,14 and is considered the palliative procedure of choice by some for children over one year of age.18 It is clearly of little help when there is infundibular atresia, an atretic or hypoplastic valve and annulus or severe pulmonary artery coarctation. This procedure has found few adherents in this country, especially with the increasing trend toward early total correction in children with favorable outflow anatomy.

*Systemic-to-Pulmonary Artery Anastomoses*

The principle of augmenting pulmonary blood flow by anastomosis of the subclavian to the pulmonary artery? was a surgical milestone. Anastomosis of the left pulmonary to the descending thoracic aorta18 added another form of shunt to the surgical armamentarium, but neither was uniformly applicable to neonates and small infants. Anastomosis of the main pulmonary artery to the ascending aorta was attempted in small infants17 but is rarely useful in the tetralogy of Fallot because the pulmonary artery is so small.11 Anastomosis of the ascending aorta to the right pulmonary artery extrapericardially18 or from within the pericardium18,20 can be done regardless of age or size.11 An end-to-side modification is possible, although it is not ideal.11 While surgical mortalities as high as 3021 and 32 percent22 have been reported for infants in the first six months of life, other series show that these operations can be performed with an extremely low mortality in all age groups.11,23,24 Published results indicating a mortality of 43 percent, "with continued cyanosis or congestive heart failure in the survivors"25 do not provide an accurate appraisal of the potential of these operations.

Clinical improvement can be sustained for years, but the majority of patients will ultimately again become symptomatic and require further surgery, usually a corrective operation. Those who require such palliative procedures early in life tend to require second operations at an earlier age.29

There is no question that these shunt operations increase arterial oxygen saturation,26,30 reduce polycythemia and allow for improvement in growth and activity.26-28 Whether or not they increase the capability of the left ventricle to sustain the systemic cardiac output after total correction is still under debate.31-34 Recent evidence suggests that left ventricular size is not affected.35 Elevation of arterial oxygen saturation above 90 percent necessitates excessive shunting through the anastomosis and predisposes to high output cardiac failure.26

If the anastomosis is made no larger than 3 mm in infants, and not made larger than 4 mm in the larger children, or if the subclavian-to-pulmonary anastomosis is used, this will rarely occur.11

Pulmonary vascular obstruction secondary to medial thickening in the small caliber pulmonary arteries occurs in less than 5 percent of children after systemic to pulmonary artery anastomoses, and is not clearly related to the degree of shunting.26,36 Acquired pulmonary atresia at the valvular or infundibular level may occur after these shunts.37 These two problems do not occur with sufficient frequency to negate the great value of these procedures, especially in neonates and infants.

**TOTAL OPEN HEART CORRECTION**

Total correction provides excellent longterm clinical and hemodynamic results in the majority of patients.31,38-44 The major obstacle to success in-
volves reconstruction of the difficult right ventricular outflow tract. The number and variety of recently proposed solutions to its management indicate that the problem is far from solved.13,22,45-53

A previous systemic-to-pulmonary anastomosis may be responsible for a higher surgical mortality at the definitive repair.44 Other experience indicates that there may be a lower mortality if the shunt causes reduction in polycythemia33 or if it increases left ventricular size and work potential in children with initially small left ventricles.31,33 Most of us feel that except for the Potts anastomosis, the results of total correction are similar in children with or without previous systemic-to-pulmonary anastomoses.32,34,42

Some31,32,55 feel that the risk of corrective surgery is higher if the patient is under five years of age, and that there is a less satisfactory outflow reconstruction and a higher incidence of heart block and residual ventricular septal defect in children operated on before this age. Others44 suggest the age of three years as the lower limit for total correction, some42 four years, some34 10 kg in weight and some15 13 kg in weight. Those56,57 who advocate open repair of other lesions in neonates feel that systemic-to-pulmonary anastomoses are preferable for neonates and for infants with tetralogy of Fallot.

The longterm hemodynamic results of total correction in children undergoing repair at two years of age and under are not yet available. Of a recently published study of a group of 13 patients operated on under the age of four years with heart recatheterization over one year later, only one was under two years old at the initial operation. Tricuspid regurgitation and a large ventricular septal defect were present in this child at recatheterization.48 Of five children under age two later operated on by this same surgical group, all survived, and had acceptable intraoperative measurements. These have been shown to be closely related to late hemodynamic results.40 Recent data indicate that there may be a recurrence of significant right ventricular outflow obstruction in some patients.44 It is not possible to predict whether or not this incidence will be higher in children with surgical repair in infancy. The attainment of 100 percent survival with good results in nine infants under the age of 18 months, utilizing deep hypothermia and circulatory arrest for total correction15 is unsurpassable. On the other hand, other reported series, often quoted as indicating excellent results in small children, indicate a 33 percent mortality, a 33 percent incidence of heart block and 33 percent with continued cyanosis (open ventricular septal defect and continued severe outflow gradient). Only one infant did well,54 and there was a 40 percent mortality in children two years old or under.90 These latter authors90 agree that palliative anastomoses are preferable in infants weighing 7 or 8 kg.

In our experience, if the right ventricular outflow tract is favorable, surgical repair can be accomplished in children of 7 kg in weight or over, with as low a risk (less than 10 percent) as older children. If the outflow tract is hypoplastic, we prefer a systemic-to-pulmonary anastomosis. No one would disagree that those children whose outflow tracts are atretic or so severely hypoplastic as to require some form of conduit reconstruction should have a palliative shunt and corrective operation after age five.31-47,50,52,53,54 The early results of outflow reconstruction by means of these valve-bearing homograft conduits have been excellent,32,47,49,53 not only in tetralogy of Fallot, but also in complicated transposition of the great vessels. The longterm results, especially relative to homograft degeneration, are impossible to predict at this early date.

The Management of Unusual Problems

Absence of One Pulmonary Artery

The left pulmonary artery is more often absent than the right. An apparent absence may be an angiographic artifact due to preferential flow to the right pulmonary artery, especially when there is stenosis at the orifice of the left pulmonary artery.43 In such cases repair may be simplified. When discontinuity is present, a more distal and sizable left pulmonary artery may be illustrated by late phase aortography, since there are usually bronchial collaterals to it. Discontinuity of the left pulmonary artery from the main pulmonary artery may be iatrogenic, produced by the performance of an end-to-end left Blalock anastomosis. Despite some experience to the contrary,44 unless continuity can be re-established or stenosis relieved, and this has been possible in some cases,42-46,65 the results of corrective surgery have been poor.40,42,63 It is not, in our opinion, feasible to repair such a lesion in the first two years of life. An anastomotic procedure is preferable. An aortic-pulmonary anastomosis in a child with a single pulmonary artery cannot be performed in the standard manner. Anastomosis of the ascending aorta to the main pulmonary artery may be performed without circulatory support, but, as a rule, the main pulmonary artery is too small to allow an anastomosis with partial occlusion.11,17 Any other anastomosis requires deep hypothermia or circulatory support by cardiopulmonary bypass.65

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The latter alternative has been successful in our hands.

**Absence of the Pulmonary Valve**

These infants can be treated with digitalis, diuretics and salt restriction for right heart failure, which is usually diminished by the first month of life. Compression of the left main bronchus with obstructive emphysema and pneumonia is a lethal chain of events. Since compression is caused by aneurysmal dilatation of the main pulmonary artery anteriorly and the descending thoracic aorta (or the left side of the vertebral column when a right arch is present), no relief of the compression has thus far been accomplished by surgical means. Total open heart correction with insertion of a heterograft pulmonary valve does not seem feasible in a neonate, nor does pneumonectomy appear to be a successful alternative.

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