Ascending Aorta-Right Pulmonary Artery Shunt in Infants and Older Patients with Certain Types of Cyanotic Congenital Heart Disease

By William F. Bernhard, M.D., Jimmy E. Jones, M.D., David Z. Friedberg, M.D., and S. Bert Litwin, M.D.

SUMMARY
A side-to-side anastomosis between the ascending aorta and the right pulmonary artery was created in 80 infants (less than 1 year of age) and in 61 older children with a variety of cyanotic cardiac abnormalities in which there is pulmonary stenosis or atresia. Seventy-one per cent of the infant group and 90% of patients over 1 year of age were long-term survivors (up to 6 years).

Tetralogy of Fallot was the most commonly encountered anomaly in all 141 patients (66%); transposition of the great vessels and pulmonary stenosis occurred in 18%; and tricuspid atresia with pulmonary stenosis in 10%.

The presence of an excessively large shunt anastomosis (55% of deaths) and additional (unrecognized) other anomalies (26%) were major causes of postoperative deaths.

In patients in whom a systemic-pulmonary artery anastomosis is required, this operation is the procedure of choice in infants under 1 year of age. It is also of value in older patients if a Blalock-Taussig shunt cannot be performed.

Additional Indexing Words:
Infant shunts

The systemic-to-pulmonary arterial anastomosis remains an important method of palliative treatment of critically ill infants with an inadequate pulmonary blood flow due to pulmonary stenosis or atresia and ventricular septal defect. While the Blalock-Taussig operation provides effective palliation for most patients over 1 year of age, the surgical results in infants have not been satisfactory.1-3 A new operative solution to this problem was described by Waterston in 1962, who created a shunt between the posterior wall of the ascending aorta and the anterior aspect of the right pulmonary artery.4 This procedure is carried out intrapericardially, since at this location the vascular structures are of sufficient size to permit satisfactory anastomosis in the small infant. In addition, the shunt is centrally located in the mediastinum and can be closed with little difficulty at the time of open correction of the cardiac defects.5,6

This report summarizes our experience with the side-to-side ascending aorta-right pulmonary artery anastomosis in a series of 141 patients admitted to the Children's Hospital Medical Center during the period 1964-1969. There were 79 males and 62 females; 80
The surgical mortality of side-to-side ascending aorta-right pulmonary artery anastomosis in 80 patients less than 1 year of age. 64 babies (80% of the infant group) were between 1 day and 6 months of age at the time of operation, and each was severely hypoxic.

Indications for Surgery

Severe hypoxemia (arterial oxygen saturation less than 50%) prompted immediate operation in the infant group (80 patients), of whom 64 (80%) were between the ages of 12 hr and 6 months (fig. 1). Cardiac catheterization and angiography were accomplished preoperatively in 75% of these patients. In the others, the diagnosis was made either by clinical means, at operation, or by postmortem examination.

In 61 older children, surgery was undertaken on an elective basis. It was necessary because of the closure of an existing Blalock-Taussig anastomosis in 45 of these patients. In the others, the presence of anatomic abnormalities prevented the construction of a satisfactory subclavian-pulmonary artery anastomosis.

Incidence of Cardiac Defects

Tetralogy of Fallot was the most common congenital anomaly encountered in the entire series of 141 patients (66%); transposition with pulmonary stenosis occurred in 18%; and tricuspid atresia with pulmonic stenosis in 10%. The remaining 10 patients had pulmonary atresia with intact ventricular septum (five patients), single ventricle and pulmonic stenosis (three patients), and dextrocardia, ventricular septal defect with pulmonic stenosis (two patients).

Results of Surgery

In the infant age group, 62 patients (78% of the infants) survived operation and were discharged from the hospital. There were five late deaths, three of which were apparently unrelated to the functioning shunt (table 1). Of the 18 hospital deaths, an excessively large shunt across the anastomosis caused acute left ventricular failure in 10 babies (55%), while intraoperative technical errors accounted for the deaths of three others. Three infants (18%) expired secondary to an unrecognized associated defect. One had mitral stenosis; one, mitral valve atresia; and the third, biliary atresia. Two other deaths resulted from overwhelming bacterial pneumonitis.

Three of the five late deaths occurred in babies with properly functioning shunts. One of these, a 6-week-old patient with tetralogy

<table>
<thead>
<tr>
<th>Anomaly</th>
<th>No. operated upon</th>
<th>No. alive</th>
<th>Survivors (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot (including VSD with main pulmonary artery atresia)</td>
<td>47</td>
<td>37</td>
<td>79</td>
</tr>
<tr>
<td>Transposition of the great vessels with pulmonary stenosis</td>
<td>15</td>
<td>10</td>
<td>67</td>
</tr>
<tr>
<td>Tricuspid atresia with pulmonary stenosis</td>
<td>11</td>
<td>7</td>
<td>64</td>
</tr>
<tr>
<td>Pulmonary valve atresia with intact ventricular septum</td>
<td>5</td>
<td>2</td>
<td>40</td>
</tr>
<tr>
<td>Hypoplastic right ventricle with pulmonic stenosis and ventricular septal defect</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Single ventricle and pulmonic stenosis</td>
<td>1</td>
<td>1</td>
<td>100</td>
</tr>
<tr>
<td>Total</td>
<td>80</td>
<td>57</td>
<td>71</td>
</tr>
</tbody>
</table>
of Fallot, died suddenly, presumably due to occlusion of the anterior descending branch of the left coronary artery by diffuse arteritis. A second infant with tetralogy of Fallot had unsuspected, extensive subendocardial fibroelastosis, and expired in congestive failure. The third death occurred in a baby with correctly diagnosed tricuspid atresia but undetected total anomalous pulmonary venous drainage to the portal vein. In two patients, death was secondary to pneumonitis and ventricular arrhythmias 3 to 6 months postoperatively, but the underlying cause probably was related to the excessive size of the shunt.

In the 61 patients over 1 year of age, there were only seven deaths (89% survival). Technical difficulties during operation accounted for four of these, while two children succumbed in the hospital with congestive heart failure, probably on the basis of an excessively large shunt. One patient died at home of unknown cause.

**Surgical Complications**

Symptoms of mild left ventricular failure developed within 24 hr following operation in 28 of the 141 patients (20%), and 15 of these with a moderate degree of heart failure were less than 1 year of age. A second operation to narrow an overly large anastomosis was undertaken in nine patients, seven of whom were babies under 6 months of age. Only four of these secondary operations were successful (fig. 2).

An inadequate shunt stoma was created in one patient, who remained cyanotic postoperatively. Successful enlargement of the anastomosis was effected 12 hr later. Late spontaneous closure of the shunt (6 months to 6 years postoperatively) occurred in 11 other patients (7%), and eight of these survived another systemic-to-pulmonary artery shunt: four patients had a Blalock-Taussig shunt, three patients had a Potts’ shunt, and one patient required a graft.

The mean systemic oxygen saturation in all surviving patients was 87%, compared to a preoperative mean value of 47% in the infant group and 68% in children over 1 year of age.

**Discussion**

A side-to-side anastomosis between the ascending aorta and right pulmonary artery has proven to be the most satisfactory palliative operation for patients under 1 year of age with an inadequate pulmonary blood flow. The success of this procedure is dependent upon the creation of a sufficiently small anastomosis (3.0 mm in diameter) to prevent flooding of the pulmonary vasculature.

*Circulation, Volume XLIII, April 1971*
and rapid development of left ventricular failure.\(^7\) Errors in the surgical creation of a proper stomal size were responsible for 40% of the deaths in this series of 141 patients and should be avoided. However, as flow is proportional to the fourth power of the anastomotic radius, a small change in stomal size has a large effect on the total blood flow. Even in the presence of an anastomosis of proper size, there frequently is uneven distribution of flow between the two lungs. The lung on the side of the anastomosis receives about 75% of the blood flowing through the anastomosis, while the opposite lung receives most of the existing flow through the pulmonary artery.\(^8\)

Determination of arterial oxygen saturation before and immediately after establishing a shunt (ventilating the lungs with the same oxygen concentration) has been helpful in gauging the correct size of the anastomosis. An arterial oxygen saturation in excess of 90%, irrespective of the initial saturation, signifies the presence of excessive pulmonary blood flow, prompting the surgeon to narrow the anastomosis.

The volume of a surgically created left-to-right shunt in the neonate may increase as the pulmonary vascular resistance diminishes, resulting in a gradual rise in systemic arterial oxygen saturation over a period of 5 to 7 days. For this reason, intraoperative arterial oxygen saturation in this age group should be limited to 90%. Despite this precaution, if respiratory distress or right-sided pulmonary consolidation develops within 12 to 24 hr following surgery, re-exploration is mandatory to permit narrowing of the shunt. Since this aggressive attitude toward this problem has been assumed, deaths from congestive heart failure have been reduced. In the past year, no patients have required a second operation for overperfusion of the lungs.

The presence of unrecognized associated congenital abnormalities accounted for six of the 23 deaths in this series and underlines the need for careful preoperative evaluation. One infant died with biliary atresia. The other five patients were discovered to have additional cardiac anomalies at postmortem examination, including mitral stenosis, mitral atresia, total anomalous pulmonary venous drainage, coronary arteritis, and endocardial fibroelastosis. In view of the critically ill state of these infants, detailed pursuit of a complete diagnosis is often unjustified. Indeed, 16 infants underwent surgery without angiographic study because they were too ill, and two of them died on the basis of unrecognized lesions.

Exploration of the chest through a right lateral thoracotomy permits the establishment of a limited diagnosis of pulmonary stenosis and the execution of necessary palliative procedure. Creation of an atrial septal defect was performed additionally in three babies with transposition and pulmonary stenosis.

Because of the anatomic relationship of the great vessels, a side-to-side anastomosis was performed between the ascending aorta and the main pulmonary artery in five neonates, with three survivors. In all instances, transposition was present, with the main pulmonary artery located to the right and posterior to the ascending aorta.

Since the majority of these patients have large pulmonary blood flows after this type of systemic-pulmonary artery shunt, digitalis administration is carried out routinely for 12 months postoperatively. In most patients, cardiac compensation can be achieved in this manner, without the necessity of diuretics.

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_Circulation_. 1971;43:580-584
doi: 10.1161/01.CIR.43.4.580
_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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