Ascending Aorta to Right Pulmonary Artery Anastomosis

Immediate Results in 123 Patients and One Month to Six Year Follow-up in 74 Patients

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SUMMARY

An intrapericardial ascending aorta-to-right pulmonary artery anastomosis was performed in 123 patients with cyanotic congenital heart disease associated with pulmonary stenosis or atresia during the eight year period from 1964 through 1971. While there were 20 early postoperative deaths (16%), 90% (93 patients) of the 103 survivors were symptomatically improved.

In the postoperative evaluation, 74 patients (72% of the survivors) returned for follow-up examination, which included cardiac catheterization in 87. The anastomosis was nonfunctioning in ten (13.5%) of these 74 patients and in an additional 13 (17.5%) patients, clinically silent right pulmonary artery stenosis was demonstrated angiographically at the anastomosis site. Clinical improvement had been maintained in the 64 patients with an open anastomosis (audible continuous murmur or angiographically demonstrated patency) including the 13 patients with acquired pulmonary stenosis at the operative site. Persistent right pulmonary artery stenosis was demonstrated in nine of the 14 patients who had cardiac catheterization after intracardiac repair of the congenital heart defect and closure of the anastomosis, but this was insignificant in four of these nine.

The ascending aorta-to-right pulmonary artery anastomosis provides symptomatic improvement for most patients with cyanotic congenital heart disease associated with pulmonary stenosis but may produce right pulmonary artery obstruction requiring attention at the time of total intracardiac repair.

Additional Indexing Words:
Cyanosis Congestive heart failure Aorto-pulmonary shunt
Cyanotic congenital heart disease

THE ASCENDING AORTA-TO-RIGHT PULMONARY ANASTOMOSIS, as described by Waterston in 1962 and by Cooley and Hallman in 1966, was devised to provide an operation which would be easier to perform in the small infant than the Blalock-Taussig operation and less complicated to close at the time of intracardiac repair than the Potts anastomosis. This report details our experience with 123 patients who had an intrapericardial ascending aorta-to-right pulmonary artery anastomosis for cyanotic congenital heart disease associated with pulmonary stenosis or atresia at the Texas Children’s Hospital and the Texas Heart Institute in the eight year period between January, 1964, and December, 1971, and the 74 patients who were evaluated one month to six years postoperatively at our institutions.

Methods

The age of the patients at the time of operation ranged from 22 hours to 29 years. Each of the operations employed the intrapericardial approach described by Cooley and Hallman. The anomaly was tetralogy of Fallot in 77 patients, transposition of the great arteries (TGA) with pulmonary stenosis (PS) in 21 patients, pulmonary valve atresia with intact ventricular septum and diminutive right ventricle in 13, and tricuspid atresia with pulmonary stenosis or atresia in ten (tables 1 and 2).

Of the 123 patients, 104 underwent cardiac catheterization before the anastomosis was performed. Systemic arterial oxygen saturation was obtained in each of these 104
patients. Main pulmonary artery (MPA) pressure was obtained in only 47 patients as the anatomic defect prevented entry of the catheter into the MPA in the remaining 57 patients. Surgery was performed in patients with recurrent cyanotic spells or systemic artery saturation below 80%. In 19 patients, operation was performed urgently on the basis of clinical diagnosis.

Postoperative cardiac catheterization was performed on 57 patients. The percutaneous sheath technique was utilized in 50 of these patients and was employed on all patients who weighed 12 pounds or more. Systemic arterial oxygen saturation was obtained and pulmonary artery pressure was measured in the MPA via the right ventricle (RV) or right pulmonary artery (RPA) via the shunt. Special attention was directed toward evaluating the shunt by recording pressures during withdrawal of the catheter from RPA to MPA and from MPA to ascending aorta when the pulmonary artery could be entered only from the aorta by way of the anastomosis. Patency of the RPA at the anastomosis site was evaluated by injecting contrast material both into the MPA when possible and into the ascending aorta, and estimating the relative flow to both the right and left pulmonary arteries.

Intracardiac repair of the defect and closure of the anastomosis was carried out in 28 patients. In 14 of these, cardiac catheterization was repeated with special attention to measurement of pressures during withdrawal from RPA to MPA, as well as the angiographic appearance of the right main branch of the pulmonary artery.

### Results

**Mortality**

Ascending aorta-to-right pulmonary artery anastomosis was accomplished in all 123 patients without any intraoperative deaths. One hundred and three patients (84%) survived the immediate postoperative period and were discharged from the hospital. Of the 20 hospital deaths, 11 resulted from intractable congestive heart failure (CHF), five from hypoxemia, and four from causes unrelated to the shunt (sepsis in two patients, disseminated intravascular coagulopathy (DIC) in one, and sudden unexplained death in one previously well postoperative child). An additional 17 deaths occurred between one month and five years postoperatively, four from CHF, five from hypoxemia and seven from unrelated causes (sepsis in one patient cerebrovascular accident in one patient, Goodpasture's syndrome in one patient and causes unknown to us in four patients). At autopsy the shunt was nonpatent in seven of the ten patients who died from hypoxemia. The remaining three had a stoma of only 1.5 mm or less in diameter. Of the 15 patients who died from CHF, 11 had an opening greater than 4 mm in diameter and four (all infants less than six months of age) had a stoma which measured between 3 mm and 4 mm in diameter. One additional patient with CHF died three months after the procedure during revision of an excessively large shunt. The total shunt-related mortality was 21% (26 of 123).

Of the patients under six months of age at the time of operation, 64% (34 of 53) survived both the early and late postoperative periods (tables 2, 3). The sur-

### Table 1

<table>
<thead>
<tr>
<th>Mortality Related to Defect</th>
<th>No. patients</th>
<th>No. deaths</th>
<th>Survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot</td>
<td>77</td>
<td>19</td>
<td>75</td>
</tr>
<tr>
<td>Transposition of the great arteries with pulmonary stenosis</td>
<td>21</td>
<td>6</td>
<td>71</td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td>15</td>
<td>1</td>
<td>93</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>10</td>
<td>0</td>
<td>100</td>
</tr>
<tr>
<td>Total</td>
<td>123</td>
<td>26</td>
<td>79</td>
</tr>
</tbody>
</table>

### Table 2

<table>
<thead>
<tr>
<th>Effect of Age and Defect on Early and Late Shunt-Related Mortality</th>
<th>Age at surgery (months)</th>
<th>No. of patients operated</th>
<th>Died &lt;1 week postop</th>
<th>Died &gt;1 week postop</th>
<th>Total shunt-related mortality (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot</td>
<td>0–6</td>
<td>32</td>
<td>9</td>
<td>4</td>
<td>41</td>
</tr>
<tr>
<td></td>
<td>&gt;6</td>
<td>45</td>
<td>3</td>
<td>3</td>
<td>13</td>
</tr>
<tr>
<td>TGA with PS</td>
<td>0–6</td>
<td>6</td>
<td>3</td>
<td>2</td>
<td>83</td>
</tr>
<tr>
<td></td>
<td>&gt;6</td>
<td>15</td>
<td>1</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td>0–6</td>
<td>11</td>
<td>0</td>
<td>1</td>
<td>9</td>
</tr>
<tr>
<td></td>
<td>&gt;6</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>0–6</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>&gt;6</td>
<td>6</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>All patients</td>
<td>0–6</td>
<td>53</td>
<td>12</td>
<td>7</td>
<td>36</td>
</tr>
<tr>
<td></td>
<td>&gt;6</td>
<td>70</td>
<td>4</td>
<td>3</td>
<td>10</td>
</tr>
</tbody>
</table>

Abbreviations: TGA = transposition of the great arteries; PS = pulmonary stenosis.

*Circulation. Volume XLIX, May 1974*
Table 3

<table>
<thead>
<tr>
<th>Age at operation</th>
<th>No. of patients</th>
<th>No. deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Under 6 months</td>
<td>53</td>
<td>19</td>
</tr>
<tr>
<td>6 – 12 months</td>
<td>20</td>
<td>2</td>
</tr>
<tr>
<td>12 – 24 months</td>
<td>24</td>
<td>2</td>
</tr>
<tr>
<td>Over 24 months</td>
<td>26</td>
<td>3</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td><strong>123</strong></td>
<td><strong>26</strong></td>
</tr>
</tbody>
</table>

Survival for patients over six months of age at the time of anastomosis was 90% (63 of 70).

Follow-up

Symptomatic improvement was observed in 93 of the 103 surviving patients (90.3%) with decreased cyanosis and cessation of hypoxemic spells. Seventy-four of the patients who underwent ascending aorto-RPA anastomoses had continued follow-up at our institutions. All patients with patent shunts had grade II to IV/V continuous murmurs best heard at the right sternal border in the second intercostal space. Congestive heart failure as manifested by cardiomegaly, engorged pulmonary vascularity, and hepatomegaly was diagnosed in 36 of the 74 patients (49%). Of these 36 patients, 14 had right-sided pulmonary edema and the remaining 22 had no radiographic evidence of pulmonary edema although they did have cardiomegaly and hepatomegaly. Of the remaining 38 patients without CHF, 22 (57%) had radiographic signs of increased perfusion of the right lung and 16 had equal vascularity in both lung fields.

Cardiac catheterization

Cardiac catheterization was performed in 57 patients between one month and six years after operation. In 36 patients, routine cardiac catheterization was performed 14 to 72 months postoperatively (mean 39.3 months) to evaluate the status of the anastomosis prior to total intracardiac corrective surgery. Increasing cyanosis was the indication for cardiac catheterization in 15 patients one month to 36 months (mean 19.0 months) after anastomosis. In six patients with intractable CHF, the cardiac catheterization was performed two to 48 months (mean 18.5 months) postoperatively.

The anastomosis was nonfunctional in ten of the 57 patients (17%) catheterized postoperatively. In 13 patients, there was no angiographic evidence of a communication between the MPA and RPA, and all of the blood flow through the shunt was directed to the right lung (fig. 1). Blood flow to the left lung was accomplished either by direct flow from the RV to MPA and LPA (fig. 2) or via bronchial collaterals. In another 13 patients, kinking of the RPA without a significant pressure gradient was seen angiographically at the site of the anastomosis.

The mean preoperative arterial oxygen saturation in 104 patients was 63.5 ± 1.5%. The 46 patients with patent anastomoses at the second catheterization had a mean arterial saturation of 80.4 ± 1.2% (P < 0.001) while those 11 with nonfunctioning shunts had a mean saturation of 64.5 ± 3.3%. Patients with tricuspid atresia had the least improvement in systemic arterial saturation. The changes in oxygen saturation for each type of cardiac defect studied are summarized in table 4.

Pulmonary artery pressures were measured in 47 patients prior to anastomosis and in 45 patients.

Figure 1

Obstruction of the RPA proximal to the anastomosis. Contrast material in the aorta opacifies only the RPA.

Figure 2

Obstruction of the RPA by the anastomosis. All blood flow from the MPA is to the LPA.
postoperatively. Of the 45, the MPA only was entered in nine patients, the RPA via the anastomosis in 14 patients, and both the MPA and RPA in 22 patients. Preoperatively, mean peak systolic MPA pressure was $16.6 \pm 0.08$ mm Hg and following surgery the pressure rose significantly in the MPA ($24.6 \pm 2.9$ mm Hg, $P < 0.005$) and in the RPA ($26.8 \pm 2.6$ mm Hg, $P < 0.001$). Eight patients developed elevated RPA pressure (>40 mm Hg peak systolic) with a range from 41/25 to 75/45 and two of these had significant RPA and MPA hypertension (75/40). All of these patients had acquired stenosis of the RPA proximal to the shunt and had CHF with either unilateral or bilateral pulmonary edema for several months after the operation was performed.

### Postsurgical repair

Total intracardiac repair and transaortic closure of the anastomosis was performed in 28 patients. In none was any attempt made to reconstruct the RPA. Cardiac catheterization was performed in 14 patients six to 21 months postoperatively (mean 14 months). In all 14 patients, the cardiac catheterization was part of the routine postoperative evaluation. There were various degrees of stenosis of the RPA at the anastomosis site in nine of the 14 patients studied. One of these nine patients had complete obstruction of the RPA at the site of the anastomosis and the only blood flow to the right lung was through bronchial collaterals. The MPA and LPA pressures in this patient were 50/5 mm Hg. In four patients, there was RPA stenosis at the site of the anastomosis (fig. 3) with systolic pressure gradients of 20 to 70 mm Hg across the area. An additional four patients had angiographic evidence of kinking of the RPA, but the pressures were the same on both sides of this area.

### Discussion

The Blalock-Taussig anastomosis has classically been the operation of choice for improving pulmonary blood flow. However, this anastomosis is often difficult to perform in infants with small subclavian arteries, in infants with retroesophageal subclavian artery, and in infants in whom creations of the shunt on the same side as the aortic arch may produce kinking of the subclavian artery. In children older than two years, however, the mortality and morbidity of this operation are considerably less and the technical problems are fewer so this approach may be the one of choice.

The Potts anastomosis is infrequently used at our institutions if the cardiac defect is amenable to total correction at a later date. Pulmonary hypertension may develop if the shunt is too large. In addition, closure of the shunt through an anterior approach at the time of total intracardiac repair is technically difficult.

The Glenn procedure (anastomosis between the superior vena cava and the right pulmonary artery) generally fails in the newborn infant and reconstitution of the right pulmonary artery and superior vena cava at the time of intracardiac repair is difficult.

### Table 4

<table>
<thead>
<tr>
<th></th>
<th>Pre-Shunt</th>
<th>Shunt Open Post-op</th>
<th>Shunt Closed Post-op</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No. patients</td>
<td>O$_2$ saturation (%)</td>
<td>No. patients</td>
</tr>
<tr>
<td>All patients</td>
<td>104</td>
<td>63.5 ± 1.5</td>
<td>47</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>63</td>
<td>66.6 ± 1.8</td>
<td>34</td>
</tr>
<tr>
<td>TGA with PS</td>
<td>21</td>
<td>61.4 ± 3.3</td>
<td>3</td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td>13</td>
<td>54.0 ± 3.6</td>
<td>6</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>7</td>
<td>61.9 ± 7.2</td>
<td>4</td>
</tr>
</tbody>
</table>

Abbreviations: TGA = transposition of the great arteries; PS = pulmonary stenosis.
The recent success reported by Barratt-Boyes and associates\textsuperscript{16, 17} with early intracardiac correction of many defects under hypothermia has not been duplicated in many centers. This approach may not be practical in cases in which a rapid increase in pulmonary blood flow would be life-saving. In addition, certain defects are not presently amenable to total correction and shunt procedures are the only chance for growth and development.

The ascending aorta-to-right pulmonary artery shunt proposed independently by Waterston\textsuperscript{1} and Cooley\textsuperscript{2} has found favor as an effective means of improving pulmonary blood flow in cyanotic infants and children. The procedure is relatively easy to perform and can be done with a minimum of dissection. Often, extracardiac anatomy can be clarified by inspection of the heart during operation and in those malformations in which it is indicated, a Blalock-Hanlon atrial septectomy can be performed utilizing the same incision. The approach is always via the right chest (unless there is L-TGA) whether the aortic arch is on the right or left and irrespective of anomalies of the subclavian arteries or bronchi. The ascending aorta is larger than the descending aorta and therefore can more easily be partially occluded during creation of the anastomosis without obstructing systemic blood flow. Closure of the anastomosis can be easily accomplished at the time of total repair when the aorta is cannulated.\textsuperscript{2}

The relatively high mortality in the group of patients less than six months old at the time of operation reflects the extremely moribund condition of many of these infants. Ten of the 53 patients under six months of age had a history of cardiopulmonary arrest before going to surgery for the anastomosis. Four of these patients subsequently died. In six cases the patients were taken directly from the catheterization laboratory to the operating suite in extremely critical condition.

The major postoperative problems encountered with this anastomosis have been directly related to the size of the shunt. In the neonate and young infant, a stoma of 3 mm is sufficient to produce adequate pulmonary blood flow without signs of CHF.\textsuperscript{11-14} In small children, the stoma may be enlarged to 4 mm while in older children a 5 mm opening is satisfactory.\textsuperscript{11} If the stoma is too small, it may close spontaneously and cyanosis and hypoxic spells will reappear.

An excessively large shunt stoma often results in CHF which may be refractory to treatment. Bernhard et al.\textsuperscript{12} have suggested that all infants undergoing shunt procedure be routinely digitalized for 12 months postoperatively. Pulmonary hypertension is also an important sequel to an excessively large shunt.\textsuperscript{6, 11} In eight of the patients in our series, there was a significant elevation of RPA pressure.

Kinking of the RPA was identified in almost half the patients (26 of 57) in our series who were recatheterized. In larger children, it can be avoided by making the anastomosis as posterior as possible, but in infants this may be difficult due to size limitations. Proper identification of the posterior wall of the aorta by a suture and alignment with another suture on the anterior wall of the RPA prior to the placement of the partial occluding clamps will identify the site of the anastomosis and may reduce the incidence of kinking. The pressure gradient across the obstructed area may not be readily apparent upon withdrawal of the catheter as there may be equal pressures in the RPA (from the shunt) and in the MPA (from the RV) and a gradient may not present until after closure of the shunt. Thus, selective angiographic study of the RPA may be required to identify the kink. If there is significant obstruction of the RPA at the site of the anastomosis, surgical repair of the involved area at the time of total intracardiac correction is imperative to avoid the development of MPA and LPA hypertension. Ebert et al.\textsuperscript{18} have proposed separation of the RPA from the aorta and pericardial patch arterioplasty of the RPA in those cases in which RPA obstruction has been identified by cardiac catheterization prior to total correction.

Roentgenographic evidence of either right-sided pulmonary edema or pleural effusion was noted in almost one-fourth of our patients. This has also been seen with the Blalock-Taussig anastomosis on either side\textsuperscript{16} or with the Potts anastomosis on the left side.\textsuperscript{6} In the ascending aorta-to-right pulmonary artery anastomosis, an excessively large anastomosis or kinking of the RPA proximal to the anastomosis may cause overperfusion of the right lung. In patients with complete obstruction of the RPA from the MPA, the entire flow through the shunt is diverted to the right lung.

Ascending aorta-to-right pulmonary artery anastomosis is effective in improving pulmonary blood flow and systemic arterial saturation in the patient with cyanosis due to pulmonary stenosis or atresia. However, distortion and kinking of the RPA just proximal to the anastomosis may result in excessive flow to the right lung suggesting that the anastomosis is too large. At the time of intracardiac repair of the cardiac anomaly and closure of the anastomosis, an unrecognized and unrelieved stenosis of the RPA at the anastomotic site may lead to elevated MPA and LPA pressure. Cardiac catheterization and selective pulmonary artery angiography are required to demonstrate the presence and severity of the stenotic RPA.
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_Circulation_. 1974;49:952-957
doi: 10.1161/01.CIR.49.5.952

_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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