Ascending Aorta-Right Pulmonary Artery Anastomosis

Clinical Experience with Thirty-five Patients with Cyanotic Congenital Heart Disease

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SUMMARY

The results of ascending aorta-right pulmonary artery anastomosis in 35 patients are reported. The patients ranged in age from 18 hours to 12 years; 16 were less than 12 months old. Seventeen had tetralogy of Fallot, six had pulmonary atresia and a ventricular septal defect, and eight had tricuspid atresia with hypoplasia of the right ventricle. In six infants under 4 months of age with tricuspid atresia, an atrial septal defect was made by the balloon catheter technique prior to the shunt operation. Four patients initially required digitalis for left heart failure. Three patients died in the postoperative period. The remaining 32 patients have been followed up to 3 years and have done well and maintained adequate shunts.

Additional Indexing Words:
Cyanosis Tetralogy of Fallot Tricuspid atresia Pulmonary atresia Ventricular septal defect Congenital heart disease Balloon atrioseptostomy

PALLIATIVE OPERATIONS continue to play an important role in the management of patients with cyanotic congenital heart disease and inadequate pulmonary blood flow. Older patients with tetralogy of Fallot can be managed by complete correction of the intracardiac defects, but in infants and small children some form of shunt operation to increase pulmonary blood flow is indicated. In addition, certain lesions such as tricuspid atresia with hypoplasia of the right ventricle or single ventricle with pulmonic stenosis are not amenable to total correction at present.

In children with these lesions, the subclavian artery-pulmonary artery anastomosis described by Blalock and Taussig has been the palliative operation of choice since its introduction in 1944. An anastomosis between the descending aorta and the left pulmonary artery as described by Potts and his associates has been preferred by most surgeons in infants.

In 1962 Waterston described a technique for creation of a systemic-to-pulmonary artery shunt by anastomosing the ascending aorta to the right pulmonary artery. Subsequently, Cooley and Hallman reported on the operation in more detail. In 1966 Edwards and his associates reported the results of a similar operation on five patients. The present report describes our experiences with 35 patients who had a variety of congenital malformations of the heart associated with cyanosis and inadequate pulmonary blood flow in
whom an ascending aorta-right pulmonary artery anastomosis was created.*

Methods

Thirty-five patients ranging in age from 18 hours to 12 years were treated surgically (table 1). Sixteen were 12 months old or less; seven were between 1 and 2 years. Seventeen patients had tetralogy of Fallot, and an additional six patients had pulmonary atresia associated with a ventricular septal defect (table 2). Eight patients had tricuspid atresia with hypoplasia of the right ventricle and pulmonic stenosis or atresia. Three infants had a common ventricle associated with transposition of the great arteries and pulmonic stenosis or atresia. Two of these had dextrocardia. One patient had a double-outlet right ventricle with pulmonic stenosis.

All patients had a history of paroxysms of cerebral hypoxia or progressively increasing cyanosis and decreasing tolerance to exercise. In the

*Three additional patients with pulmonary atresia and intact ventricular septum had an ascending aortic-pulmonary artery anastomosis after a balloon atrial septostomy. Since these infants, in general, have not been successfully treated in the past by a systemic-to-pulmonary artery shunt, they will be presented in a separate report to be published.

Table 1

<table>
<thead>
<tr>
<th>Age</th>
<th>Number of patients</th>
<th>Deaths</th>
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</thead>
<tbody>
<tr>
<td>Under 1 year</td>
<td>16</td>
<td>2</td>
</tr>
<tr>
<td>1 to 2 years</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>Over 2 years</td>
<td>12</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>35</td>
<td>3</td>
</tr>
</tbody>
</table>

Table 2

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Number of patients</th>
<th>Deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot</td>
<td>17</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary atresia and ventricular septal defect</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>Common ventricle, pulmonary stenosis, and transposition of great vessels</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Double-outlet right ventricle and pulmonary stenosis</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>35</td>
<td>3</td>
</tr>
</tbody>
</table>

patients with tetralogy of Fallot total correction was not carried out either because of their young age or their small size or both. In those patients over 3 years of age, an ascending aorta-pulmonary artery shunt was chosen over a Blalock anastomosis because of the presence of stenosis of the left subclavian artery in association with a right aortic arch (two patients), because a right aortic arch was first recognized at the time of operation (one patient), or because of a previous left thoracotomy for cardiac arrest in another patient with a right aortic arch. In two other patients a previous Blalock anastomosis had closed. Another older boy had only a right pulmonary artery and many collateral vessels in the mediastinum.

All patients had diagnostic cardiac catheterization before the shunting operation. In six patients under 4 months of age with tricuspid atresia and inadequate pulmonary blood flow, an atrial septostomy using the balloon catheter technique was employed after completion of the diagnostic study. These latter patients were severely anoxic and were managed on an emergency basis; the anastomosis was created within a few hours following septostomy.

The anastomosis was performed through a right lateral thoracotomy in the fourth intercostal space except in those with dextrocardia in whom left thoracotomy was done. The pericardium was opened anterior to the phrenic nerve and the atrial appendage retracted inferiorly. The right pulmonary artery was exposed from its origin at the main pulmonary artery to its bifurcation into the lobar vessels. A heavy silk ligature was placed around the artery distally. A curved arterial clamp was placed on the ascending aorta, the posterior blade lying behind the right pulmonary artery (fig. 1). A 4-mm incision was made in the occluded portion of the ascending aorta and a similar incision was made transversely in the pulmonary artery. The latter was made slightly shorter than the aortic incision since the pulmonary incision tended to enlarge. The posterior row of the anastomosis was constructed with a continuous 6-0 Tevdek suture. The anterior portion was completed with interrupted sutures. After release of the clamp and ligatures from the vessels and re-expansion of the lung, the amount of flow through the anastomosis was gauged by palpation of the pulmonary artery. If the continuous thrill felt coarse, suggesting an excessively large shunt, an additional suture was placed in the corner of the anastomosis and its size reduced.

Results

An ascending aorta-right pulmonary artery anastomosis was accomplished successfully in

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35 patients. Three patients died in the postoperative period. One, an 18-hour-old male with tricuspid atresia and inadequate pulmonary blood flow, died 48 hours after operation because of increasing bronchoalveolar obstruction of the lung. It was evident that the shunt had been functioning well and there was no heart failure. At autopsy the lungs showed diffuse cystic changes. Another infant who was 2 weeks old and had tricuspid atresia and inadequate pulmonary blood flow died from unrecognized peritonitis 4 days following a successful septostomy and anastomosis. The cause for the peritonitis was not found at autopsy. The third death occurred in a physically underdeveloped 9-year-old girl with tetralogy of Fallot, who as an infant required open chest cardiac resuscitation following separation from a conjoined twin. Six days after operation this child died suddenly of unknown cause following a smooth initial course. Before death she had unequivocal signs of an effective shunt and no evidence of heart failure. At autopsy the aortic-pulmonary anastomosis was patent, and no other abnormalities were found to explain the death.

A 6-month-old infant with pulmonary atresia and ventricular septal defect had a prolonged postoperative course complicated by atelectasis of the right lower lobe and an emphysematous right upper lobe. However, he gradually improved with bronchoscopy and antibiotic therapy. In the other 31 patients clinical improvement was maintained as long as 3 years following operation. Cyanosis was either slight or absent, activity tolerance was normal, and none of the patients had recurrence of hypoxic spells.

In the immediate postoperative period 32 of 35 patients had a grade I to III/VI continuous murmur, best heard to the right of the midsternum or under the right clavicle.
In most instances it was also audible posteriorly in the right interscapular space. Several months postoperatively, the continuous murmurs were frequently of greater intensity and more diffusely heard than earlier. In four patients a continuous murmur was never heard. In one this was due to the small size of the anastomosis in a patient with a very small pulmonary artery. The other three patients died in the postoperative period, and in each instance a widely patent anastomosis was found at postmortem examination.

Fifteen of the 35 patients were treated with digoxin in the postoperative period. Of these only four had definite evidence of congestive heart failure. These latter patients responded promptly to digitalization. Digoxin was given to the remaining 11 patients because of tachycardia without overt evidence of congestive heart failure. In all 32 survivors, a small increase in heart size was noted roentgenographically during the first 2 weeks after operation. However, even in the four patients with definite heart failure, the degree of cardiac enlargement was not marked.

One patient developed chylothorax 4 weeks after operation. She was treated by multiple thoracenteses with excellent results.

**Discussion**

In patients with cyanotic congenital heart disease and inadequate pulmonary blood flow, the effectiveness and low risk of a pulmonary artery-systemic artery anastomosis is well established. Total correction when possible in lesions such as tetralogy of Fallot carries a high mortality risk in infants and small children. Therefore, a palliative procedure has been advocated if surgical treatment is necessary prior to the age of five. The Blalock-Taussig anastomosis has been the operation of choice for this purpose. The Potts anastomosis is less desirable since it is more difficult to estimate the optimal size of this anastomosis and because its obliteration at the time of subsequent total correction has often been difficult.

However, in certain situations the subclavian artery-pulmonary artery anastomosis is difficult or impossible to perform: (1) in infants with small subclavian and pulmonary arteries, (2) in patients with a retro-esophageal subclavian artery, and (3) in patients with stenosis or atresia of the subclavian artery.

The introduction of the ascending aorta-right pulmonary artery shunt has provided a solution to many problems related to the Blalock and the Potts anastomoses. The technique is simple to perform, requiring minimal dissection. Complications such as a Horner's syndrome are avoided. The arterial supply to the arm remains intact. Furthermore, the ascending aorta is larger than the descending aorta used in a Potts anastomosis and, therefore, can more readily be partially occluded in infants without significantly obstructing aortic flow. Closure of the anastomosis at the time of total correction of a tetralogy of Fallot can be readily accomplished by opening the ascending aorta and placing several sutures into the defect.

The Waterston shunt is more centrally located between the right and left pulmonary arteries as compared to the other anastomoses. It therefore was expected to supply both lungs with more nearly equal flows of blood. However, chest roentgenograms obtained in three of our patients suggested a greater flow to the right lung than the left, perhaps due to tenting of the pulmonary artery at the anastomosis site.

The main difficulty encountered with the use of the new anastomosis has been the accurate determination of its size. Most of the anastomoses have measured 4 mm or less in diameter in patients under 2 years, and 5 mm in older children. Because of the threat of serious complications such as left heart failure, marked cardiomegaly and pulmonary hypertension with shunts that are too large, this point of surgical technique requires special consideration.

Because the ascending aorta-right pulmonary artery anastomosis offers a number of advantages over a Potts anastomosis, there are now few indications for the latter operation. However, in view of the difficulty in

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judging the proper shunt-size with the Waterston operation, the Blalock anastomosis should be used as the procedure of choice in those patients over 3 years of age who have no anatomic contraindications to the performance of this procedure.

An aortic-pulmonary artery anastomosis (Waterston six, Potts one) preceded by an atrial septostomy has been performed in seven severely hypoxic infants under 4 months of age with tricuspid atresia. Cardiac catheterization data in conjunction with a right atrial dye injection and cineangiocardio gram suggested partial obstruction to flow from the right atrium to the left. One operative death occurred in an infant who had severe obstructive lung disease as an associated problem. Another infant died of peritonitis 4 days following operation. No cause for the peritonitis was found at autopsy. In older patients with tricuspid atresia and hypoplasia of the right ventricle, a low pressure shunt as described by Glenn is preferable to a high pressure shunt. However, it can rarely be successfully accomplished in infants under 6 months of age. After a systemic-pulmonary artery anastomosis, however, the elevated pressure and flow in the pulmonary artery may make a subsequent superior vena cava-right pulmonary artery anastomosis possible.

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