Status of Systemic Artery to Pulmonary Artery Shunts in Complex Congenital Heart Disease

By

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Introduction:

Primary repair of cyanotic congenital heart disease has become increasingly popular over the past decade. This is mainly due to technological advances which have allowed cardiopulmonary bypass and postoperative care of the sick neonate to reach an advanced level of sophistication. Blalock-Taussig Shunts which since 1945 (1) had been the mainstay of palliative surgical management in babies with cyanotic heart disease are now done much less frequently. Because of the small caliber of the vessels involved, many surgeons who wish to palliate these neonates have often used direct aorto-pulmonary shunts (2, 3, 4). Despite recent reports from many centers (5,6) advocating primary repair as the treatment of choice in infancy, we believe that the systemic artery to pulmonary artery shunts, notably the Blalock-Taussig shunt and its modification using an expanded polytetrafleuroethylene (PTFE) graft, play an important role in the early management of cyanotic heart disease where the complexity of the anomaly or lack of facilities preclude primary repair in infancy. Where a Blalock-Taussig shunt fails, direct aorto-pulmonary aretry shunts can still be done but these are not the shunts of choice because of their high incidence of congestive heart failure and increased pulmonary vascular disease (7,8). This report reviews our experience with infants and children with congenital heart disease associated with inadequate pulmonary blood flow.

Clinical Material:

In the four year period between July 1979 and June 1983, a total of 35 shunts were done in 32 infants and children. 17 were classical Blalock-Taussig shunts. 16 were modified Blalock-Taussig shunts and 2 were central aorto-pulmonary shunts. There were 19 boys and 13 girls. 18 patients were one month of age, 8 were infants between one month and one year of age and 6 children were between one year and 5 years of age. The youngest patient was 5 hours old at the time of the shunting procedure. The age distribution of the patients at the time of the shunting is given in Fig. 1.

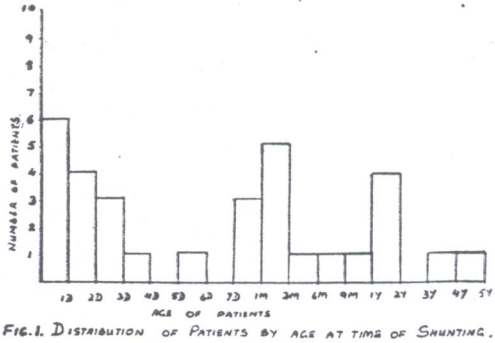

FIG. 1. DISTRIBUTION OF PATIENTS BY AGE AT TIME OF SHUNTING.

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The frequency of the congenital heart defects for which the shunts were done is given in Table 1. The most common anomaly

Table I: Defects Palliated by Systemic Artery-to-Pulmonary Artery Shunts

Anomaly	*No. of Pts.*
Complex TOF	14
Tricuspid Atresia	3
AV Canal with PS	2
TGA with PS/Hypoplastic LV	6
Univentricular Heart with PA	3
DORV with PS	1
PA with IVS	1
PA with VSD	2

TOF: Tetralogy of fallot
AV Canal: Atrio-ventricular canal
PS: Pulmonic stenosis
TGA: Transposition of great arteries
LV: Left ventricle
PA: Pulmonary Atresia
DORV: Double outlet right ventricle
IVS: Intact ventricular septum
VSD: Ventricular septal defect

was Tetralogy of Fallot. Most of these with TOF had complex forms of the disease which prevented primary repair in infancy. Of the 18 patients less than one month of age only 6 had TOF. The indications for shunting were cyanosis, hypoxic spells and decreasing exercise tolerance.

Operative Technique:

Classical Blalock-Taussig shunt: The classical Blalock-Taussig shunt was performed through a posterolateral thoracotomy incision and the chest was entered through the fourth intercostal space. Almost all initial Blalock-Taussig shunts were done on the side where the subclavian artery arose from the innominate artery to allow better flow characteristics through the downwardly directed subclavian artery. However, with minor modifications of technique, the shunt could be performed almost as easily on the side where the subclavian artery originated directly from the aortic arch (9). The pulmonary artery was dissected and vascular controlled achieved beyond the first bifurcation of the RPA or LPA. The subclavian artery was dissected up to the vertebral artery. The vertebral artery was ligated and after proximal clamping the subclavian artery was transected in such a manner that its distal end flared from inclusion of the origin of the vertebral artery. Sufficent dissection around the innominate and common carotid arteries was required that when the transected subclavian artery was brought down anterior to the recurrent laryngeal nerve, it had enough length and mobility that the anastomosis to the pulmonary artery could be carried out without tension. Heparinization of the patient was neither required nor desirable. The anastomosis was then carried out between the subclavian artery and the ipsilateral pulmonary artery. The whole anastomosis was done with a running 6-0 prolene suture and because of the flared distal end of the subclavian artery, the diameter of the anastomosis was always greater than that of the subclavian artery proximally. Major complications with this procedure are rare and none were encountered in the present series.

Modified Blalock-Taussig Shunt: In this procedure, instead of transecting the subclavian artery and redirecting it downwards for direct anastomosis with the pulmonary artery,

an expanded polytetrafluoroethylene (PTFE) graft (usually 4-6 mm. in diameter) was used. The proximal end of the graft was anastomosed to the subclavian artery a short distance distal to its origin and the distal end of the graft was anastomosed to the ipsilateral pulmonary artery. The diameter of both the proximal and distal anastomosis is wider than that of the subclavian artery and also of the pulmonary artery and helps maintain long term patency of the shunt. It should also be noted that the continuity of the subclavian artery is not interrupted so perfusion of the arm is not jeopardized.

Results:

In this series 35 shunts were done in 32 patients. There were 17 classical Blalock Taussig shunts and 16 modified (PTFE) Blalock-Taussig shunts. Two central aorto-pulmonary shunts were required after failure of classical BTS. There were no intraoperative deaths.

Of the 17 patients with classical BTS, all patients were initially relieved of their cyanosis. There were three early failures at one day, 3 days and 19 days. The first patient with TOF and pulmonary atresia was operated at 5 hours of age and had thrombosis of the shunt after one day. He was reoperated on with performance of a central aorto-pulmonary shunt but died two days later. The second patient with TOF and virtual infundibular atresia had thrombosis of a right classical BTS on the third day and was revised by interposition of PTFE graft between the right subclavian and pulmonary artery and is doing well. The third patient with TOA, single ventricle, mitral and pulmonary atresia had a right classical BTS and a Blalock-Hanlon procedure at one day of age; the shunt subsequently became inadequate so a central ascending aorta to main pulmonary artery shunt was done 19 days later. The patient is doing well. There were four early deaths in the 17 patients with classical BTS; one is indicated above; three others were in patients with complex congenital heart lesions, who died with patent shunts. Three other patients died from this group following subsequent definitive repair of the congenital anomaly. Thus there was a total of 7 deaths among the 17 patients who were initially palliated with a classical BTS.

Of the 16 patients with a modified Blalock-Taussig shunt, there was only one death. The patient had severe tricuspid atresia and a hypoplastic right ventricle with pulmonary atresia. He was shunted at four days of age and a 5 mm. PTFE graft was used. The diameter of the native subclavian artery was 2 mm. He died two days later of low cardiac output. The remaining 15 patients continue to do well with functioning shunts.

Table 2 gives the mortality in this series related to type of shunt.

Table 2: Mortality Related to Type of Shunt

Type of Shunt	*No. of Pts. Shunted*	*No. of Pts. Died After Shunt*	*No. of Pts. Died After Definitive Repair*	*% Mortality*
Modified BTS	16	1	0	6%
Classical BTS	17	4	3	41%

Overall 24 patients are currently alive with adequate shunts. Shunt patency in these patients is assessed by the presence of an audible shunt murmur and lack of abnormal symptoms in each patient.

Comment:

Improved results of corrective surgery have reduced the need for palliative shunts which are now applied mainly to neonates and infants with complex congenital cardiac defects which preclude primary repair in infancy, or where facilities for such complex corrective procedures are not available.

The Waterston shunt (ascending aorta to main pulmonary artery) has been used in the neonatal period because it is technically easier to perform (2,3). However, it has the distinct disadvantage of being associated with excessive pulmonary blood flow causing congestive cardiac failure (7,8,10) and may also seriously deform the pulmonary artery. The Potts anastomosis (descending aorta to left pulmonary artery) is also associated with excessive pulmonary blood flow (11), and is difficult to take down at the time of intra-cardiac correction. The Glenn shunt (superior vena cava to right pulmonary artery) has a prohibitive mortality in early infancy and is no longer used (12). The Blalock-Taussig shunt (subclavian to pulmonary artery) has become the shunt of choice in the older infant but was previously avoided in the neonate because of difficulty with the small subclavian artery. With the improvements in techniques of micro-vascular surgery this shunt is however being applied to smaller infants (13). A modification of the Blalock-Taussig shunt is utilization of an expanded PTFE graft between the uninterrupted subclavian artery and the pulmonary artery. This shunt has several advantages. 1) It allows persistent arm perfusion by not interrupting the subclavian artery, 2) It has better flow characteristics with no kinking of the subclavian artery as might occur with a classical BTS on the side where the subsclavian artery originates directly from the aorta, 3) A 4-6 mm. shunt is used whenever possible so that as the child grows the subclavian artery can increase in size and increase the flow through the shunt, 4) Also, since it is a wide diameter shunt, the chances of thrombotic occlusion are .smaller than with a classical BTS using a small diameter subclavian artery.

The superiority of the modified BTS is borne out by our data with no shunt failures in 16 patients. In comparison, 3 of 17 shunts failed in the classical BTS group.

It must also be noted that this shunt can be constructed where abnormal anatomy or other technical considerations may make a classical BTS impossible to construct. These shunts are easy to ligate at the time of a subsequent reconstructive procedure and do not cause kinking of the pulmonary artery. The theoretical disadvantage of using a foreign material has not been a problem in our patients or in other reported series (14,15).

Since the followup of these patients has been relatively short, however, we are restricting the use of the PTFE shunt to infants and neonates who do not have ideal anatomy for a Blalock-Taussig shunt or where a classical BTS has failed.

Our late mortality is disappointingly high but was not related to shunt failure.

In conclusion, a classical Blalock-Taussig shunt can be performed with acceptable mortality in the neonate and provides effective and long term palliation. The modified BTS using the PTFE graft offers several advantages over the classical procedure but because of a relatively short follow-up, its use is restricted at present to patients where unfavorable anatomy precludes a classical BTS or a classical BTS has failed.

Summary

We reviewed our experience with systemic artery—pulmonary artery shunts in infants and children at University of Maryland Hospital and compared the classical Blalock-Taussig shunt (BTS) to modified BTS.

A total of 35 shunts were done in 32 infants and children. 16 were modified BTS, 17 were classical BTS and 2 were central aorto-pulmonary shunts. 18 patients were less than 1 month of age, 8 were infants between 1 month and 1 year of age, and 6 children were between 1 year and 5 years of age. The indications for shunts in this series include complex forms of TOF, tricuspid atresia, AV canal with pulmonic stenosis, TGA with pulmonic stenosis/hypoplastic left ventricle, univentricular heart with pulmonary atresia, double outlet right ventricle with pulmonary stenosis, pulmonary atresia with IVS and pulmonary atresia with VSD. In 24 patients the shunt was the initial procedure while in 8 it followed a previous procedure (Waterston shunt 2, previous BTS 5, and pulmonary valvotomy 1).

Of the 16 patients with modified BTS, only 1 with severe tricuspid atresia and hypoplastic right heart died. The remaining 15 have patent shunts and are doing well. Of the 17 classical BTS, 3 failed initially. One required a modified BTS and 2 a central aorto-pulmonary shunt. Another four of these 17 patients died from complex congenital anomalies with patent shunts and three died following subsequent attempts at definitive repair. Overall 24 patients are currently alive with functioning shunts.

Based on this experience we conclude that the modified BTS is often preferable to the classical BTS. The advantages are: A) it allows persistent arm perfusion, B) has better flow characteristics with no kinking of subclavian artery and C) is a wide diameter shunt. In our experience this has resulted in superior early patient survival and more satisfactory long term results.

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