Systemic to Pulmonary Shunting in 93 Cyanotic Neonates

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Background: **Systemic to pulmonary shunting is still the main palliation option in many neonates with cyanotic heart disease.**

Design: A Retrospective Study.

Setting: The Mohammed bin Khalifa bin Salman Al-Khalifa Cardiac Center, Bahrain Defence Force Hospital, Bahrain.

Method: All infants from January 1995 to December 2008 who had undergone systemic to pulmonary shunting procedures were included in the study.

Result: Ninety-three infants underwent 108 shunt type procedures, 39 females and 54 males. One hundred five Modified Blalock-Taussig Shunts (MBTS) and three classic Blalock-Taussig shunts were performed. Tetralogy of Fallot/Double Outlet Right Ventricle (DORV) with right ventricular outflow tract obstruction was the most common diagnosis, 51 (55%).

Early postoperative mortality was 1 (1%). Late postoperative mortality was 4 (4%); sepsis was the major cause. Early (<1 month postoperatively) shunt failure/occlusion occurred in 6 (6%) patients. Under 14 days of age at the time of shunting and shunt size of 5mm (18% versus 4.6%) were risk factors for early shunt occlusion/failure. Eight (8%) patients required late shunt revision at two months to 5 years post initial shunting. Fifty-one patients had undergone final corrective surgery.

Conclusion: MBTS shunting provides effective and safe palliation in small infants with complex cyanotic heart disease. Early shunting (<14 days of age), and 5mm shunt size are additional risk factors for early shunt failure.

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Systemic to pulmonary shunting provides palliation to neonates and small infants with many complex forms of cyanotic heart disease. It is usually the first step palliation in these neonates who are either unsuitable for total correction or due to the higher pulmonary vascular resistance are unable to undergo bidirectional Glenn anastomosis early in life.

The modified Blalock-Taussig Shunt (MBTS) is the most commonly used systemic to pulmonary artery shunt whereby a synthetic conduit of Polytetrafluoroethylene (PTFE) is anastomosed between a subclavian artery and a pulmonary artery branch. MBTS provides adequate and regulated blood flow to the pulmonary circulation in these critically ill cyanotic neonates.

The aim of this study is to evaluate the morbidity and mortality of MBTS performed on infants.

METHOD

All infants from January 1995 to December 2008 who had undergone systemic to pulmonary shunting procedures were included in the study.

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All the infants had an initial diagnosis made by cross-sectional echocardiography. No patient required initial preoperative cardiac catheterization as the assessment of the anatomy was appropriate by echocardiography alone.

In the MBTS technique, the diameter of the Gortex tube graft used was directly related to the size of the pulmonary artery branch to which it was anastomosed. If the pulmonary artery branch was 4 mm or smaller by cross-section echocardiography a 4mm diameter Gortex tube graft was used.

Heparin (100units/kg) was continued 6-12 hourly postoperatively for 24-48 hours. All patients who underwent MBTS also received aspirin postoperatively at 3-5mg/kg/day indefinitely or until final corrective/palliative surgery was performed.

Shunt failure was defined as either clinical evidence of insufficient pulmonary blood flow with the absence of a shunt murmur and/or echocardiographic evidence of occlusion or insufficiency within one month of operation. Shunt revision was defined as either clinical evidence of insufficient pulmonary blood flow in the presence of a shunt murmur and/or echocardiographic evidence of insufficiency more than one month postoperatively. Clinical and cross-section echocardiographic follow up to evaluate patency of the MBTS was performed routinely in all patients until final corrective/ palliative surgery was planned.

RESULT

Ninety-three infants underwent 108 shunt procedures, 39 females and 54 males. The majority of infants were of Bahraini origin, see figure 1. Forty-eight (51.6 %) were less than one month of age at initial shunting procedure, see figure 2. One hundred five MBTS were constructed, and three classic Blalock-Taussig type shunts were performed. Ninety-nine (91%) of the procedures were performed in the two hospitals in the Kingdom of Bahrain, see figure 3. All three classic types of shunts were performed elsewhere and thus were not included in this analysis; only the modified shunts were included in the study. The size of the 105 MBTS is shown in figure 4 with a 5mm shunt being the most common size. Initial shunting was always performed on the contralateral side of the aortic arch unless technical/anatomical considerations required shunting on the same side as the aortic arch.

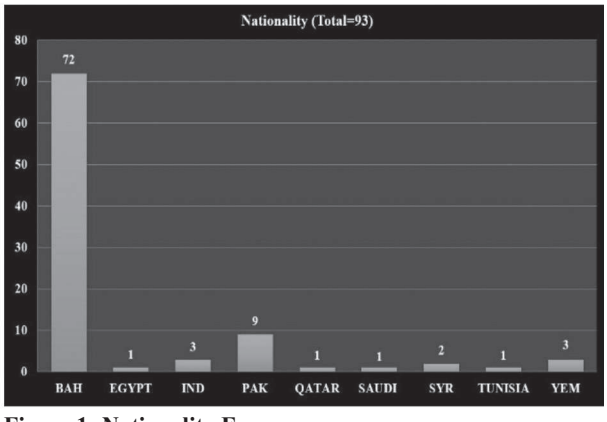


Figure 1: Nationality Frequency

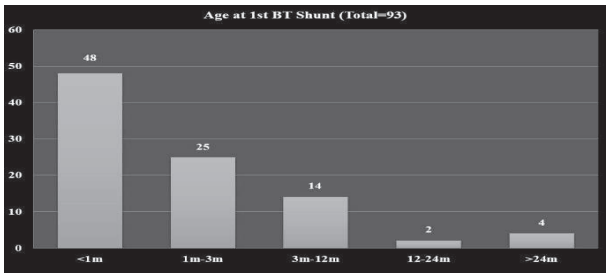


Figure 2: Age Distribution

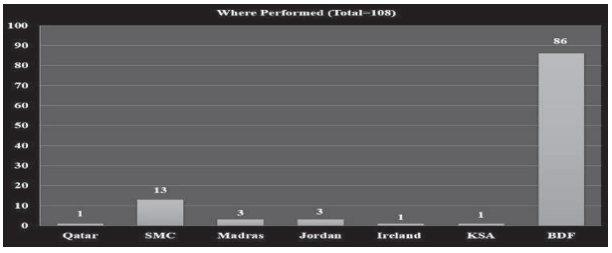
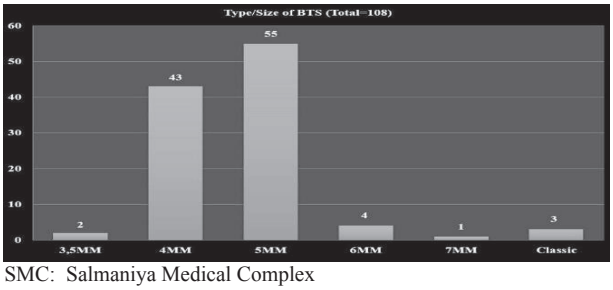


Figure 3: The Setting of the Procedure



SMC: Salmaniya Medical Complex
KSA: Kingdom of Saudia Arabia
BDF: Bahrain Defence Force Hospital

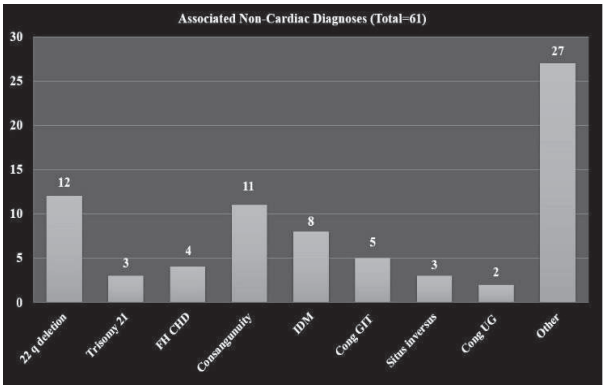
Figure 4: Type and Size of BTS

A wide variety of complex cyanotic lesions were encountered, see table 1. Tetralogy of Fallot/Double Outlet Right Ventricle (DORV) with right ventricular outflow tract obstruction (RVOTO) was the most common diagnosis, 51 (47.2%). Two patients had 3.5mm MBTS as part of Stage 1 Norwood procedure for hypoplastic left heart syndrome. Sixty-one (56.5%) infants had significant non- cardiac diagnosis, see figure 5.

Table 1: Type of Cardiac Lesions

AVSD/TA	1
DTGA/LVOTO	5
DORV/RVOTO	11
EBSTEIN	2
LTGA PA	3
PA	7
TOF	40
TRIC ATRESIA	14
UVH	10
Total	**93**

AVSD TA: atrioventricular septal defect with tricuspid atresia
DTGA/LVOTO: Dextro transposition of the great arteries with left ventricular outflow tract obstruction
DORV/RVOTO: double outlet right ventricle with right ventricular outflow tract obstruction
LTGA/PA: Levo transposition of the great arteries with pulmonary atresia
PA: pulmonary atresia
TOF: Tetralogy of Fallot
UVH: univentricular heart



FH CHD: Family History of Congenital Heart Disease
IDM: Infant of Diabetic Mother
Cong GIT: Congenital Gastro-Intestinal Tract Anomaly
Cong UG: Congenital Uro-Genital Anomaly

Figure 5: Associated Non-Cardiac Diagnoses

Five (4.6%) deaths after initial MBTS were recorded. None were directly related to the procedure. One (1%) early death (12 days post MBTS) was due to a blocked endotracheal tube in an infant 2.7kg with tricuspid atresia. Four (4%) late deaths were encountered. Two at 2 and nine months postoperatively with proven sepsis, and two at 4 and nine months postoperatively with presumed sepsis. There were no deaths, early or late, in any patients who underwent second shunt operation.

Eight (8%) patients required late shunt revision two months to 5 years after initial MBTS operation, see figure 6. None of these late revisions were due to acute shunt occlusion, but rather chronic anastomotic stenosis and/or visceral outgrowth of the initial shunt.

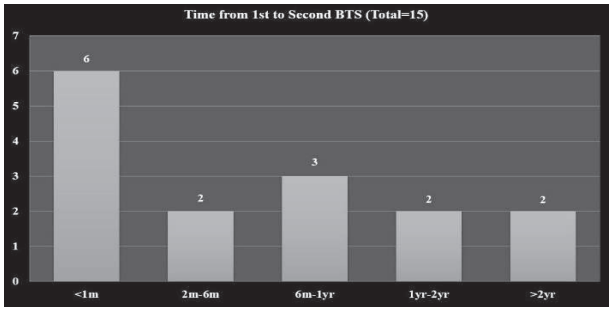


Figure 6: Time from First to Second BTS

Fourteen (13%) shunts needed re-shunting. Six (6%) had early shunt failure (less than one month postoperatively). All the six shunts were done before 14 days of age. Of the total shunt failure/occlusion, 10/55(18%) were 5mm in diameter, and 2/43 (4.6%) were 4mm in diameter, see figure 7.

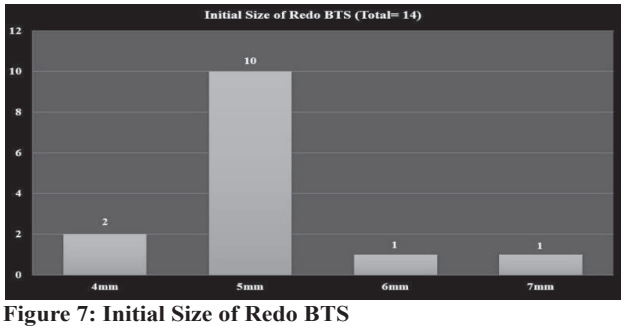


Figure 7: Initial Size of Redo BTS

Fifty-one patients have undergone final corrective/palliative procedures, see figure 6. Intracardiac repair of Tetralogy of Fallot/DORV with right ventricle outflow tract obstruction (RVOTO) is the most common procedures performed (63%). The rest of the infants are awaiting the second/final stage palliation or correction.

DISCUSSION

Recent advances in surgical techniques have resulted in many palliative procedures becoming obsolete. The systemic to pulmonary arterial shunts have however remained as mainstay palliation in many ill infants with ductal-dependent pulmonary circulation. It is often the first step in a multi-stage approach. This study confirms that there are still a significant number of cyanotic infants in whom early definitive care is not possible and will require shunting.

The MBTS has been found to be the preferred technique for such palliation, and the classic technique is now seldom used[1,2,3,4]. Shunting is a relatively low-risk procedure with perioperative mortality ranging from 3%-35%[5,6,7,8]. Improved mortality rates are probably due accurate diagnosis, earlier referral, liberal prostaglandin use, improved surgical/anesthetic techniques and postoperative care. This study showed that deaths were more common in the initially shunted infants with sepsis (proven/suspected). There were no deaths in infants undergoing a repeat shunt procedure.

Early shunt failure/occlusion (<1month postoperatively) remains a serious concern. Low-birth-weight(<2kgs), pulmonary artery size <4mm, not using postoperative heparin and/or low dose aspirin, low initial pH and preoperative mechanical ventilation have all been identified as risk factors for early failure/occlusion[4,9]. We have identified that shunt size of 5 mm as an additional risk factor (18% versus 4.6%). Late failure rates are low considering expected somatic growth. Some authors have attributed this to the use of aspirin although no comparisons to a control group have been made[4].

CONCLUSION

MBTS shunting provides effective and safe palliation in small infants with complex cyanotic heart disease. Tetralogy of Fallot/Double Outlet Right Ventricle (DORV) with right ventricular outflow tract obstruction is the most common diagnosis. Procedure mortality is low (1%) and sepsis appears to be contributing factors to late mortality. Early shunt failure occurred in 6% of infants and early shunting (<14 days) and 5mm shunt size are additional risk factors for early shunt failure. Late shunt failure is rare and no late revisions were due to acute shunt occlusion, but rather chronic anastomotic stenosis and/or visceral outgrowth of the initial shunt.

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Competing Interest: None.

Sponsorship: None.

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Ethical Approval: Approved by the Mohammed bin Khalifa bin Salman Al-Khalifa Cardiac Center, Bahrain Defence Force Hospital, Bahrain.

REFERENCES

1. Ullom RL, Sade RM, Crawford FA Jr, et al. The Blalock-Taussig Shunt in Infants: Standard versus Modified. Ann Thorac Surg 1987; 44(5):539-43.
2. Kumar DA, Kumar RN, Rao PN, et al. Systemic to Pulmonary Arterial Shunts in Neonates. Ind J Thorac Cardiovasc Surg 2003; 19:159-162.
3. Gold JP, Violaris K, Engle MA, et al. A Five-Year Clinical Experience with 112 Blalock-Taussig Shunts. J Card Surg 1993; 8(1):9-17.
4. Al Jubair KA, Al Fagih MR, Al Jarallah AS, et al. Results of 546 Blalock-Taussig Shunts Performed in 478 Patients. Cardiol Young 1998; 8(4):486-90.
5. Guyton RA, Owens JE, Waumett JD. The Blalock-Taussig Shunt. Low Risk, Effective Palliation and Pulmonary Artery Growth. J Thorac Cardiovasc Surg 1983; 85(6):917-922.
6. Arciniegas E, Blackstone EH, Pacifico AD, et al. Classic Shunting Operations as Part of Two-Stage Repair for Tetralogy of Fallot. Ann Thorac Surg 1979; 27(6):514-8.
7. Edmunds LH Jr, Stephenson LW, Gadzik JP. The Blalock-Taussig Anastomosis in Infants Younger than 1 Week of Age. Circulation 1980; 62(3):597-603.
8. Neches WH, Naifeh JG, Park SC, et al. Systemic-Pulmonary Artery Anastomosis in Infancy. J Thorac Cardiovasc Surg 1957; 70:91-92.
9. Alkhulaifi AM, Lacour-Gayet F, Serraf A, et al. Systemic Pulmonary Shunts in Neonates: Early Clinical Outcome and Choice of Surgical Approach. Ann Thorac Surg 2000; 69(5):1499-504.