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Does an undersized modified Blalock–Taussig shunt according to weight do better in Tetralogy of Fallot with pulmonary atresia?

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Abstract

Introduction

The Blalock–Taussig shunt (BTS) is regarded as a safe and effective means of increasing pulmonary blood flow for cyanotic heart conditions. The evaluation of shunt size for postoperative hemodynamics and until second stage remains difficult. Our objective was to determine the outcome of an undersized M-BTS according to weight on immediate and after 12 months of follow-up in patients who had Tetralogy of Fallot with pulmonary atresia as a first step for total correction of biventricular repair.

Patients and methods

A total of 28 patients were recruited from January 2013 to December 2016. Clinical variables, such as age, sex, and weight, and perioperative variables, such as the diameter of the graft and branch pulmonary artery size, were reviewed. The mortality, morbidity, and O_2 saturation have been chosen as the outcome variables in immediate and 12-month follow-up results.

Results

A total of 28 M-BTSs were done with no mortality. All patients were done with right lateral thoracotomy. The mean age was 34 ± 31.9 months and weight was 12.2 ± 8.7 kg. There were 10 male and 18 female cases. The mean size of the shunt is 4.1 ± 0.9 mm. We used 5-mm shunts in eight cases, 4-mm shunts in 10 cases, 3.5-mm shunt in six cases, and 3-mm shunts in four cases. The mean size of the main pulmonary artery was 5.6 ± 1.5 mm, left pulmonary artery was 4.5 ± 1.6 mm, and right pulmonary artery was 6.5 ± 5.6 mm. Two patients had postoperative bleeding.

Conclusion

Undersized M-BTSs according weight reflect better immediate and short-term results in patients with Tetralogy of Fallot with pulmonary atresia.

Keywords: Modified Blalock–Taussig shunt, pulmonary atresia, Tetralogy of Fallot, undersized

INTRODUCTION

In spite of the ongoing trend toward the primary surgical repair of complex congenital heart diseases, modified Blalock–Taussig shunts (M-BTSs) are still required as a palliative surgical management with diminutive pulmonary blood flow before surgical repair [1]. In Tetralogy of Fallot (TOF) and its related cardiac anomalies, Blalock–Taussig shunts (BTS) have been indicated for only a limited number of patients with anoxic spells, ductus-dependent pulmonary circulation, severe pulmonary stenosis, or a combination of these conditions [2].

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The purpose of a BTS is to create a reliable path of blood flow to the lungs when the pulmonary blood flow is restricted and the patient is not suitable for total repair [3,4].

In spite of continuous improvement of operative and postoperative management, overall mortality after MB-T shunts from neonatal to older patients ranged from 2.3 to 16.0% [5].

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Although flow through the shunt is controlled by the size of the subclavian artery (SCA) in M-BTS, the length and diameter add an additional resistance to the blood flow. Other considerations in determining the size of the graft selected include the presence of an additional source of pulmonary blood flow (pulmonary stenosis versus pulmonary atresia) and an estimate of pulmonary vascular resistance [6].

In principle, the shunt size is titrated to around 1.16(0.9-1.6) mm/kg body weight [7]. The general rule of thumb is to select a shunt size of 3 mm for body weight up to 3 kg, and 3.5 mm is the optimal size to use in a term neonate of ~3.5 kg.

If a 3.0-mm shunt is used, the risk of sudden thrombosis and acute obstruction in the post shunt period is increased. Early institution of anticoagulation with low-dose heparin 10–20 units/kg/h is important once the hemostasis is secured. Another significant issue with a small shunt is the likelihood of the child outgrowing the shunt size causing progressive hypoxemia and cyanosis. On the contrary, if a large shunt is used in a neonate (>4 mm), ensuing excessive pulmonary blood flow may compromise systemic perfusion, causing Left Ventricle volume overload, failure, and prolonged ICU and hospital stay [8].

PATIENTS AND METHODS

Ethics committee approval was taken. A total of 28 cases were treated from January 2013 to December 2016. All cases underwent right lateral thoracotomy.

All patients had TOF with pulmonary atresia. Excluded from this study were patients with pulmonary atresia who underwent a simultaneous valvotomy and/or right ventricular outflow tract augmentation, and any other patients undergoing associated intracardiac procedures in addition to a M-BTS.

A total of 28 M-BTSs were done for patients with cyanosis and small pulmonary arteries of TOF with a Mc-Goon ratio less than 1.6, and we had no mortality. We chose the shunt size according to the weight, with a shunt size of less than or equal to 1 mm/kg body weight, which were relatively small shunts to avoid prolonged ventilation and use of inotropic supports. These were primary procedures before the total correction in a biventricular repair for patients with TOF with pulmonary atresia, which is usually done after one year. Mortality, morbidity, and O_2 saturation were the risk predictors.

All patients underwent preoperative transthoracic echocardiography; routine preoperative laboratory tests including complete blood picture, liver and renal function tests, and O_2 saturation; and multislice computed tomography scan for the pulmonary arteries.

Operative technique

The patient was put in right lateral position in a standard fashion. Right standard thoracotomy was done. The SCA was dissected free of adventitial tissue, with its proximal part sufficient for clamping.

The corresponding right pulmonary artery (RPA) was dissected between ascending aorta and superior vena cava

sufficient for clamping and avoiding upper lobe branch compromise.

Heparin is routinely administered in all patients at a dose of 1 mg/kg intravenously before clamping the SCA.

An interposition Gor-Tex polytetrafluoroethylene (PTFE) tube graft (WL Gore, Elkon, Maryland, USA) of a suitable size is selected, and the proximal end is beveled.

A C-clamp was applied to the SCA, and an arteriotomy is performed at the inferior aspect of the artery; anastomosis was done with Gor-Tex (PTFE) tube graft using 7/0 proline continuous suture. The graft is cut to final length. A C-clamp is applied to the right PA.

The RPA is opened longitudinally and the distal anastomosis by 8/0 proline suture between the PTFE graft and the RPA is completed. The clamp is released to allow back bleeding from the PA. Hemostasis is achieved and antegrade flow is permitted by releasing the clamp on SCA. An open shunt should result in a drop in systolic blood pressure and an increase in heart rate and arterial oxygen saturation (80–85%).

Heparin was not reversed. A single chest tube is placed. The thorax was closed in a standard fashion. Postoperatively, all patients were mechanically ventilated with adjustment of fraction of inspired oxygen as low as possible down to 30% which maintains oxygen saturation of more than 75%. They were extubated when hemodynamically stable with satisfactory arterial blood gas levels.

Use of inotropic support may be indicated to maintain adequate systemic perfusion pressure during and after the surgery to prevent early shunt thrombosis. A diastolic pressure of greater than 30 mmHg is recommended to maintain coronary perfusion. Epinephrine 50–150 ng/kg/min infusion was used to maintain systolic pressures over 100 mmHg to ensure shunt patency, which was assessed by oxygen saturation, auscultation of shunt murmur either over the chest or over the opening of the endotracheal tube, and also blood gas levels and echocardiography.

Postoperative heparin was continued for 24 h in all patients, especially with small shunts (3-3.5 mm), where it was continued for 48 h. All patients were discharged on aspirin 5-10 mg/kg/day.

Patients who tolerate the procedure well and exhibit improved oxygenation at the end of the shunt were mostly subjected to early extubation and fast tracking.

Clinical variables (age, sex, and weight) and perioperative variables, such as the diameter of the graft and branch pulmonary artery size, were reviewed. The mortality, morbidity, and O_2 saturation have been chosen as the outcome variables based on their importance on reflecting the shunt function.

RESULTS

A total of 28 shunts were done with no mortality (0%). The mean age was 34 ± 31.9 months and the mean weight was

 12.2 ± 8.7 kg. There were 10 (35.7%) male and 18 (64.3%) female cases.

All shunts were selected in size less than or equal to 1 mm/kg body weight; the mean size of the shunt was 4.1 ± 0.9 mm, and we used 5-mm shunts in eight (28.6%) cases, 4-mm shunts in 10 (35.7%) cases, 3.5-mm shunts in six (21.4%) cases, and 3-mm shunts in four (14.3%) cases.

The mean size of the main pulmonary artery was 5.6 ± 1.5 mm, the mean size of the left pulmonary artery was 4.5 ± 1.6 mm, and the mean size of the RPA was 6.5 ± 5.6 mm.

A 4-mm right M-BTS to the RPA through right lateral thoracotomy was done for one case after one year and a half of previous central 5-mm left M-BTS to the main pulmonary artery with the continuation to left pulmonary artery; the RPA did not grow and she needed a redo surgery, as in Fig. 1 and Table 1.

All patients had postoperative oxygen saturation ranged from 75–85% immediately and 70–80% after 12-month follow-up. Two (7.1%) patients had postoperative bleeding and were reopened. Two (7.1%) cases had accessory left anterior descending (LAD) crossing in front of the right ventricular outflow tract (RVOT).

Two cases had restricted shunt flow after 12 months of follow-up: one of them had immediate total correction and the other one had another shunt after 3 months as the Mc-Goon ratio was still 1.3 and pulmonary arteries still did not grow well.

Six cases were done as redo after previous median sternotomy shunts owing to one of the following causes: pulmonary

Table 1: Demographic characteristics	of the study group
	n (%)
Right lateral thoracotomy	28 (100)
Mortality	0 (0)
Mean age (months)	34±31.9
Mean weight (kg)	12.2±8.7
Sex	
Male	10 (35.7)
Female	18 (64.3)
Redo	6 (21.4)
Size of shunt	4.1±0.9 mm
5 mm	8 (28.6)
4 mm	10 (35.7)
3.5 mm	6 (21.4)
3 mm	4 (14.3)
Size of pulmonary artery and its branches	
MPA	5.6±1.5 mm
LPA	4.5±1.6 mm
RPA	6.5±5.6 mm
Bleeding	2 (7.1)
Redo after previous shunt	6 (21.4)
Accessory LAD crossing RVOT	2 (7.1)
Restricted shunt flow postoperatively	2 (7.1)

LPA, left pulmonary artery; MPA, main pulmonary artery; RPA, right pulmonary artery.

arteries were not grown adequately, restricted flow through previous shunts, cyanosis, and small Mc-Goon ratio for total biventricular repair.

DISCUSSION

Primary correction is the preferred approach in neonates and young adults. With pulmonary atresia and reduced pulmonary flow, an initial palliative arterial systemic-to-pulmonary shunt is mandatory. Use of PTFE graft offers major advantages: greater pulmonary artery growth with less distortion of pulmonary arteries, lower shunt failure rate, preservation of SCA, and fewer technical problems including ease of insertion and take down [9].

In the earlier era, before the improvement in perioperative management, the studies reported relatively high mortality. The study done by Tamisier *et al.* [10] reported an overall mortality of 21%. Although El Midany *et al.* [1] had better results, the mortality was relatively high 13.8%.

According to the recent annual reports in Japan [11], the 30-day mortality and hospital death rates of the systemic-to-pulmonary shunt were 2.0 and 2.9%, respectively, and even in neonates, the 30-day mortality and hospital death rates were 1.7 and 2.3%, respectively. In our study, there was no mortality (0%) postoperatively and after 12 months of follow-up; however, the number of cases in our study was relatively small.

In most series including ours, patients with TOF and pulmonary atresia tend to do better [12]. Therefore, we select all patients with TOF and pulmonary atresia to fix the pathology and to prove that concept.

Evaluation of shunt size for good postoperative hemodynamics remains difficult. The effect of shunt size on outcome is still a debatable issue. Nosseir stated that mortality was lower in large shunt group in single ventricle physiology [13].

However, Dirks *et al.* [7] studied 32 patients. They reported 9% mortality, and they concluded that bigger shunt size per kg of body weight was a significant risk factor for mortality.

This meets with what we found in our series; we did not prefer large shunts and chose shunt size according to the weight, which was relatively small to avoid prolonged ventilation and use of inotropic supports.



Figure 1: A right modified Blalock–Taussig shunt through right thoracotomy with shunt size of 4 mm after previous central shunt through median sternotomy.

The same study was done by Rao *et al.* [14], and they studied 46 patients. They used 3.5 mm in 13 patients, 4 mm in 31 patients, and 5 mm in two patients and reported one case of shunt blockage in each shunt size group. They concluded that smaller grafts do not increase the incidence of shunt blockage [14]. In our study, we had no shunt blockage, with only restricted shunt flow after 12 months in two patients, although we used relatively undersized shunts.

El Midany *et al.* [1] had surgical preference toward smaller shunt sizes (3.5 or 4 mm) to avoid overshunting and to minimize the pulmonary artery distortion that may occur with larger shunt sizes. The small shunt size did not affect the short-term outcome in intervention for shunt blockage [1]. This meets the reason for our preference of using smaller shunts, and we avoided larger ones.

Shibata *et al.* [15] stated that a small shunt of 3 mm is safe and provides adequate pulmonary blood flow. They had 18 patients weighing an average 3.5 kg who underwent M-BTS, where 10 patients had 3-mm shunts and 8 patients had 3.5-mm shunts. There were no hospital deaths or shunt occlusion and no differences in postoperative saturated O_2 or body weight. One patient with 3.5-mm shunt had cardiogenic shock owing to excessive pulmonary blood flow [15].

Mullen *et al.* [16] conducted a study on 23 patients who underwent M-BTSs and they suggested that avoidance of postoperative heparin is a safe practice and may reduce bleeding problems and the incidence of perigraft seromas. They suggested that shunt thrombosis is more likely related to intraoperative technical difficulty or extremely small pulmonary artery size [16].

However, Rao *et al.* [14] stated that shunt block can be a problem even in a technically well-performed shunt, and other factors may play a role. They recommended the judicious use of postoperative heparin in patients with small pulmonary arteries or those with a high hematocrit to reduce the incidence of shunt blockage [14].

We used postoperative heparin for 24 h in all patients and for 48 h in only 10 (35.7%) cases with small-sized shunts (3–3.5 mm). We had no shunt blockade and only two (7.1%) cases of postoperative bleeding.

CONCLUSION

Undersized M-BTS according weight reflects better immediate and short-term results in patients with TOF with pulmonary atresia.

Limitations

The current study has several limitations that should be considered. This is a small prospective cohort, single study. These limitations, as it represents our preliminary experience with a small number of patients, need to be confirmed by larger series.

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Conflicts of interest

There are no conflicts of interest.

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