Risk factors for mortality in patients with pulmonary atresia undergoing modified Blalock–Taussig shunt

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Background
Pulmonary atresia (PA) is one of the most difficult pathologies to manage, and the surgical procedures related are associated with high mortality and morbidity. A single approach is impractical because of the wide spectrum of right morphology which makes a uniform surgical approach impossible. Anatomical criteria, such as the degree of tricuspid valve or right ventricular hypoplasia or whether a right ventricular infundibulum is present, have been used to assign the infants to single-ventricular or biventricular repair tracks. Palliative procedure to provide a more stable source of pulmonary blood flow is often required before undergoing a more definitive palliation or repair. Despite its widespread use, modified Blalock–Taussig shunt (MBTS) has been associated with high morbidity and mortality
Aims
The aim of the study was to specify the risk factors for mortality in patients with PA undergoing MBTS operation.
Patients and methods
A prospective nonrandomized study was conducted on 32 patients with PA who underwent MBTS operation between August 2019 and January 2022. Data including demographic data, preoperative status, and morphological factors were correlated to the outcome. Surgery was performed through a posterolateral thoracotomy or sternotomy according to surgeon preference. Nonstretchable ringed Gore-Tex grafts were used to create MBTS between the subclavian artery or innominate artery and the right or left pulmonary artery. Hospital mortality and related risk factors were studied.
Results
The mean age at operation was 13.32 ± 8.36 months. The study involved 17 (53.1%) males and 15 (46.9%) females. Mean weight was 6.86 ± 2.71 kg. Posterolateral thoracotomy was done in 19 (59.4%) patients. Patent ductus arteriosus (PDA) was ligated on 12 (37.5%) patients. The mean total hospital stay was 11 ± 5.94 days. Hospital mortality was seven (21.9%) patients. Isolated risk factor for hospital mortality showed that weight at the time of operation was a statistically significant risk factor (P=0.02). Complications were statistically significant in relation to mortality in patients who had reoperations of the MBTS and low cardiac output.
Conclusions
MBTS for patients with PA had a high mortality especially in smaller weights. PDA stenting should be studied as an alternative option in selected cases.

Keywords:
modified Blalock–Taussig shunt, mortality, pulmonary atresia

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Introduction

Pulmonary atresia (PA) is one the most difficult pathologies to manage, and the surgical procedures related are associated with high mortality and morbidity. The lesion is morphologically heterogeneous, with varying degrees of right ventricular and tricuspid valve hypoplasia. Aberrations of the coronary circulation are common [1].

Natural history of PA shows a high early mortality of 68% in first year of life in all types of PA without any type of interventions. Moreover, in patients who survive without interventions, they have limited exercise capacity [2].

A single approach is impractical because the wide spectrum of right heart morphology makes a uniform surgical approach impossible. Anatomical criteria, such as the degree of tricuspid valve or right

ventricular hypoplasia or whether a right ventricular infundibulum is present, have been used to assign the infants to single-ventricular or biventricular repair tracks [1]. A palliative procedure to provide a more stable source of pulmonary blood flow is often required before undergoing a more definitive palliation or repair. A surgical shunt, most commonly the modified Blalock–Taussig shunt (MBTS), introduced by De Leval *et al*. [3] has been a widely used as an initial palliative surgical procedure to provide those with ductal-dependent pulmonary flow. Despite its widespread use, the MBTS has been associated with high morbidity and mortality [4].

Morbidity and mortality after MBTS is associated with several factors including the baseline age, lower weight, cardiac anatomy, chromosomal abnormalities, smaller pulmonary artery diameter, and patent ductus arteriosus (PDA) ligation [5–7].

There is scarce literature that focus on the outcomes of MBTS on this subgroup of patients, especially in limited resource settings. Herein, incidence and predictors of hospital mortality of PA after MBTS were studied.

Patients and methods

Methods

After ethical approval of the institutional board, a prospective nonrandomized study was conducted on 32 patients with PA who underwent MBTS operation between August 2019 and January 2022. PA with intact interventricular septum (PA IVS) and PA with ventricular septal defect (PA VSD) were included in the study. All patients with PA as an associated lesion not as the main pathology as in tetralogy of Fallot with PA subtype, PA with right ventricular-dependent pulmonary circulation, and patients undergoing MBTS surgery as part of the Norwood operation were excluded.

Data including demographic data, preoperative saturation, and presence of concomitant noncardiac abnormality, morphological factors including pulmonary artery size, presence of VSD, and major aortopulmonary collateral arteries (MAPCAS) were correlated to the outcome.

Surgery was performed through a posterolateral thoracotomy or sternotomy according to surgeon preference. Nonstretchable ringed Gore-Tex grafts were used to create MBTS between the subclavian artery or the innominate artery and the right or left pulmonary artery. Sizes of the graft used ranged from 3.5 to 5 mm. The patients were followed up postoperatively until hospital discharge after recording the complications. Hospital mortality and related risk factors were studied.

Statistical analysis

Statistical analysis was done using IBM SPSS statistics for windows, Version 23.0 (Armonk, NY: IBM Corp.). Quantitative data were presented as minimum, maximum, mean, and SD. Qualitative data were presented as count and percentage. Student's *t* test was used to compare quantitative data between two independent groups. χ^2 (or Fisher's exact) test was used for comparison of qualitative data between different groups. *P* value less than or equal to 0.05 was considered statistically significant.

Results

The mean age at operation was 13.32 ± 8.36 months. The study involved 17 (53.1%) males and 15 (46.9%) females. Other demographic data and preoperative risk factors are described in Table 1.

Surgically, posterolateral thoracotomy was done in 19 (59.4%) patients. Left subclavian artery was connected to the left pulmonary artery in all thoracotomy approaches, while the innominate artery was connected to the left pulmonary artery through sternotomy approach. A shunt size of 3.5 was used in three patients, size 4 in 19 patients, and size 5 in 10 patients. PDA was ligated on 12 (37.5%) patients. Cardiopulmonary bypass was not needed in any patients.

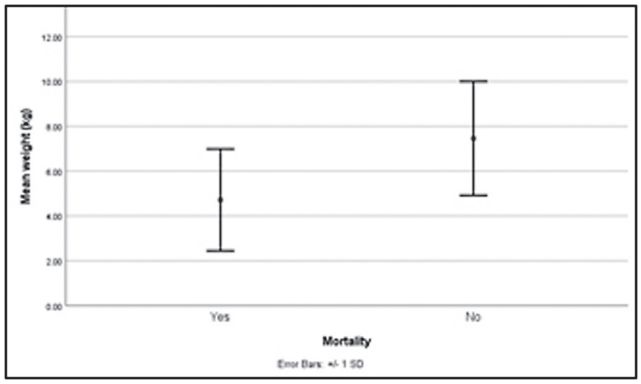
The mean duration of postoperative mechanical ventilation was 48.97 ± 43.42 h and ICU stay was 6.47 ± 3.55 days and the mean total hospital stay was 11 ± 5.94 days. The mean immediate postoperative saturation was 82% on mechanical ventilation on fraction of inspired oxygen of 0.4 and 79.74% on room air. Postoperative heparin infusion was used in 20 (62.5%) patients. Hospital mortality was seven (21.9%) patients (Figs 1,2). Patients died after reoperation due to shunt thrombosis, three patients after low cardiac output, and two patients due to shunt malfunction. Postoperative complications are described in Table 2.

Table 1 Clinical characteristics and preoperative variables

Variables	*n* (%)/mean±SD
Weight (kg)	6.86 ± 2.71
Preterm	2 (6.3)
Oxygen saturation (%)	60.09 ± 7.59
PA size (mm)	4.64 ± 0.82
PA VSD	18 (56.3)
PA IVS	14 (43.8)

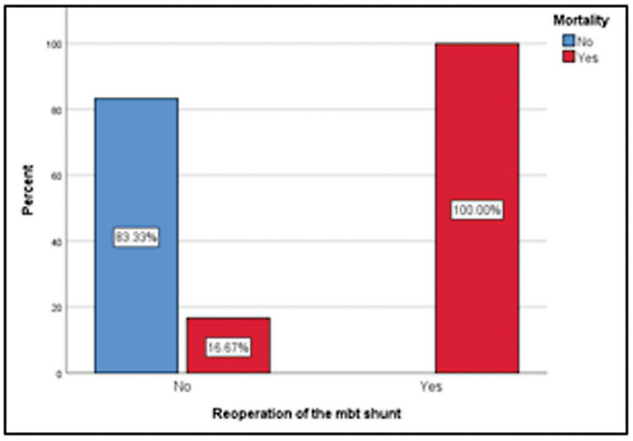
IVS, interventricular septum; PA, pulmonary atresia; VSD, ventricular septal defect.

Figure 1



Risk analysis of the weight and mortality.

Figure 2



Risk analysis of reoperation of the MBTS and mortality. MBTS, modified Blalock–Taussig shunt.

Table 2 Total number of postoperative complications (*N*=32)

Complications	*n* (%)
Bleeding	2 (6.3)
Sepsis	2 (6.3)
Pneumonia	3 (9.4)
Shunt malfunction	6 (18.8)
Low COP	3 (9.4)

COP, cardiac output.

Isolated risk factor for hospital mortality showed that weight at the time of operation was a statistically significant risk factor (P=0.02). The mean weight in the mortality cases was less than the surviving patient (4.71 ± 2.27 vs. 7.46 ± 2.55 kg) (Fig. 1). Mean age was less on the mortality group than the surviving patients (8.18 ± 7.38 vs. 14.76 ± 8.17 months) (*P*=0.07). All other demographic and anatomical risk factors showed no statistically significant association with mortality (Table 3).

Ligation of the duct had no significant differences on the outcome. Four (30.8%) patients who had sternotomy as an approach died while three (15.8%) died after the thoracotomy approach (*P*=0.40). Immediate saturation after the surgery was 69 ± 21.41% on the mortality group versus 85.7 ± 3.63% in the surviving patients (*P*=0.08).

Complications were statistically significant in relation to mortality in patients who had reoperations of the MBTS and low cardiac output (Figs 2,3). Complications are described in Table 4.

Discussion

PA is one of the most complex pathologies in the cyanotic heart disease as there is a variation in anatomical subtypes and significant morphological heterogeneity [8]. Initial palliation to patients with PA is still the most common trend aiming to promote pulmonary artery growth by several techniques such as the MBTS, central shunt, and catheter interventions, which could increase the success rate of total repair. It is also more used in patients with very low oxygen saturation, nonconfluent pulmonary arteries, and small right ventricular size [9]. Recently, there is a shift to repair at an earlier stage in selected cases with or without prior intervention [10].

There is scarce literature dedicated to discuss the outcomes of MBTS operation on PA patients in particular.

Dorobantu *et al*. [11] reported that mortality in patients who had MBTS in different pathologies was doubled to 10% in the era between 2000 and 2013 because of the decrease in the trend of using shunt for tetralogy of Fallot and increase of using it in PA. Another study by Maghur *et al*. [12] on 93 patients who underwent MBTS reported six mortality, 50% of them were patients with PA, and the mortality in PA patients alone was 33%.

PA IVS was studied by Petrucci *et al*. [4], and they reported 15.6% mortality in 179 patients with a statistically significant mortality out of the 7.2% overall mortality in 1273 patients. This was explained with possible undiagnosed ventricular coronary artery fistulas or sinusoids.

In a study by Alsoufi *et al*. [13,14], 23% mortality on 53 PA IVS was reported and that was statistically significant as a risk of hospital death on analysis of 174 patients.

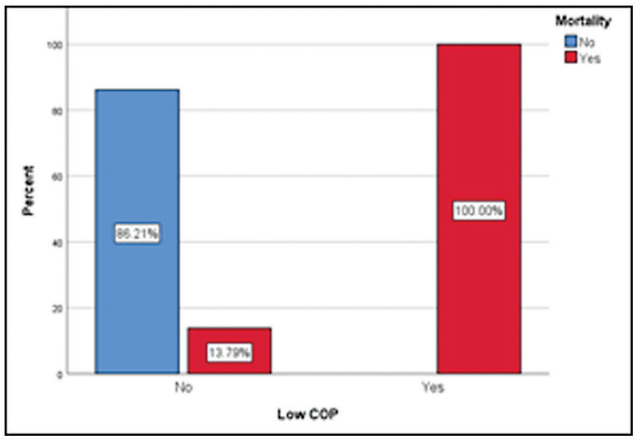
Mortality in this study was seven (21.9%) patients, which is similar with the previous study but on the other hand Fenton and McKenzie reported a lower mortality.

Table 3 Risk analysis of demographic, clinical, and anatomical data and mortality

	Mortality				*P* value
	Yes (*N*=7)		No (*N*=25)		
	Mean/*n*	SD/%	Mean/*n*	SD/%	
Sex					
Female	4	26.7	11	73.3	0.68
Male	3	17.6	14	82.4	
Preterm					
No	6	20.0	24	80.0	0.40
Yes	1	50.0	1	50.0	
Preoperative oxygen saturation	59.00	8.64	60.40	7.44	0.67
Noncardiac abnormalities					
No	7	24.1	22	75.9	1.00
Yes	0	0.0	3	100.0	
PA size	4.20	0.67	4.76	0.83	0.11
PA IVS	4	28.6	10	71.4	0.67
PA VSD	3	16.7	15	83.3	
MAPCAS					
Negative	7	23.3	23	76.7	1.00
Positive	0	0.0	2	100.0	

IVS, interventricular septum; PA, pulmonary atresia; VSD, ventricular septal defect.

Figure 3



Risk analysis of low COP and mortality. COP, cardiac output.

Table 4 Risk analysis of complications and mortality

	Mortality [*n* (%)]		*P* value
	Yes (*N*=7)	No (*N*=25)	
Bleeding			
No	7 (23.3)	23 (76.7)	1.00
Yes	0	2 (100.0)	
Sepsis			
No	7 (23.3)	23 (76.7)	1.00
Yes	0	2 (100.0)	
Pneumonia			
No	7 (24.1)	22 (75.9)	1.00
Yes	0	3 (100.0)	
Shunt malfunction			
No	5 (19.2)	21 (80.8)	0.59
Yes	2 (33.3)	4 (66.7)	
Reoperation of the MBTS			
No	5 (16.7)	25 (83.3)	0.04
Yes	2 (100.0)	0	
Low			
No	4 (13.8)	25 (86.2)	0.01
Yes	3 (100.0)	0	

MBTS, modified Blalock–Taussig shunt.

Fenton *et al*. [15] reported 5.7% hospital mortality in their study on a similar number to this study on 35 patients with PA IVS on during a period of 11 years. A mortality of 8.8% was found in a study for 16 years by McKenzie *et al*. [16] on 97 patients with PA. Another result was obtained by Alsoufi *et al*. [14], who found a 6.9% mortality in 58 patients with PA VSD.

The high mortality of this study could be explained by many reasons: late time of diagnosis and presentation due to lack of national screening programs for such complex cyanotic heart disease.

No statistically significant difference was found in mortality between PA VSD and PA IVS. However, Dorobantu *et al*. [11] found that PA IVS is a risk factor for high mortality, which was 16.5% in PA IVS and 6% in PA VSD. Meanwhile, PA VSD was found to be a risk factor for increased reintervention.

In their study to compare the results of MBTS to total repair, Kwak *et al*. [17] found a mortality of 10.3% in 29 patients with PA VSD.

This study demonstrated that patients' weight at the time of procedure' is a significant risk factor for death after MBTS. This finding is in accordance with the findings reported by several collective studies [18–21].

Petrucci *et al*. [4] concluded that low weight (<3 kg) is a risk factor for mortality in MBTS in general [1] Small weight (<3 kg) showed an estimated median survival probability of 71.3%, but in patients of more than 3 kg this was 81.2%.

In this study, the mean age was 13.32 ± 8.36 months but this was statistically nonsignificant (*P*=0.07). However, Dorobantu *et al*. [11] found that age is a significant risk factor for early death in MBTS operation, in general on his study on 1993 patients. PDA ligation has no relation to mortality in our study and this could be explained by the small sample size. Zahorec *et al*. [22] reported that patients with a surgically closed arterial duct had a higher mortality (9.7 vs. 0%, *P*=0.038). They explained it as the management of overpulmonary circulation is easier than hypoxic event after closing the duct.

No significant difference was found between the outcomes of sternotomy or the thoracotomy approach on this study, but in a larger study by including different pathologies, mortality was significantly higher in sternotomy than thoracotomy (*P*=0.03); this may be attributed to a more proximal insertion shunt site in sternotomy than thoracotomy, which may also increase pulmonary circulation and subsequently intubation time [18].

In addition to the high mortality, MBTS also affects the future repair. Amark *et al*. [23] studied the results of total repair in PA VSD and they found that initial placement of a systemic-pulmonary artery shunt was an independent risk factor of death after surgery.

Several literatures agreed that catheter intervention with right ventricular outflow tract (RVOT) or duct stent is a safe option to these patients with a comparable outcome in terms of mortality and pulmonary artery growth and superiority in terms of ICU, hospital stay, and complications [24,25]. However, it was reported that they have more intervention and 17% of patients required MBTS eventually due to failed PDA stenting [26]. On the basis of this, we suggest that PDA stenting could be a safe option for patients with PA presented with low weight. When the baby reaches a larger body weight, MBTS or Glenn shunts are other valid options, if total repair is not feasible in properly selected cases.

Limitations

The small number of cases is the most significant limitation that contributed to have many risk factors becoming nonsignificant in comparison to the findings of other big studies on MBTS on different pathologies. That is also the main reason for including the results of the two types of PA on one study as separating them will decrease the total number in this group of patients. There is scarce literature on this group of patients. The lack of follow-up for these patients after hospital discharge results is another drawback.

Conclusion

Our study contributed to knowledge regarding the outcomes of MBTS in patients with PA in Egypt. MBTS for patients with PA had a high mortality especially in smaller weights. PDA stenting should be studied as an option in selected cases. More studies are needed to report a larger number of patients on each subtype separately with midterm and long-term results.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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