Mid and long-term evaluation of left modified blalock-taussig shunt after cavopulmonary shunts: A retrospective cohort study

Original Article

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ABSTRACT

Background: Congenital heart diseases are the most common type of congenital malformations. Some of them may pass unnoticeable in the first days or maybe years of life, but the others are pretty serious and life-threatening due to major malformations that lead to the mixing of oxygenated and deoxygenated blood. Most of these cases are not repairable and require separating the venous and arterial blood to help improve oxygen saturation (SpO_2).

Aim: The study aims to evaluate the mid and long-term effects of left modified blalock-taussig (MBT) shunt after bidirectional cavopulmonary shunt.

Patients and Methods: A retrospective cohort study was conducted on 20 patients who underwent MBTS after Bidirectional Glenn. We evaluated the mid and long-term effects of MBTS in those patients, especially SpO_2 and echocardiographic findings. The study considered the ethical principles of the Helsinki Declaration approval from the research ethics committee at our institution (MS 510/2023).

Results: There was a significant increase in postoperative SpO_2 in comparison to preoperative SpO_2 ($P<0.001$) with a mean difference of 20.95%.

Conclusion: Congenital cyanotic heart disease represents a life-threatening condition where the main problem is desaturation due to the mixing of arterial and venous blood. Bidirectional Glenn is a step to separate the oxygenated and deoxygenated blood. But later on, SpO_2 continues to decline and further intervention is needed. In this study, we found that MBTS provides a postoperative significant increase in SpO_2 in a wide group of those patients even who are not candidates for other operations without significant complications.

Key Words: Cavopulmonary shunts, congenital heart diseases, modified blalock-taussig shunt.

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INTRODUCTION

Congenital heart diseases are the most common type of congenital malformations[1]. Some of them may pass unnoticeable in the first days or maybe years of life, but the others are pretty serious and life-threatening due to major malformations that lead to the mixing of oxygenated and deoxygenated blood. Most of these cases are not repairable and require separating the venous and arterial blood to help improve oxygen saturation (SpO_2)[2].

As the main problem here is desaturation due to the mixing of arterial and venous blood, trials were made to create another pathway to help improve SpO_2 and separate the arterial and venous blood. Cavopulmonary shunts were described as a possible solution for long-standing cyanosis[3].

Bidirectional Glenn (BDG) is one of the cavopulmonary shunts that aims at improving SpO_2 where the superior vena cava (SVC) is transected at its attachment with the right atrium and anastomosed to the pulmonary artery[4]. Although BDG relieves the cyanosis for some time, most cases develop desaturation with advanced age, and further intervention is needed[5].

Fontan operation is performed as a second stage of repair where the IVC with its junction with the right atrium is connected to the pulmonary artery. This procedure results in complete separation of pulmonary and venous return (total cavo-pulmonary shunt)[6] but it has many restrictions and many complications, especially in the long term. Most of the complications are caused by chronic systemic venous hypertension due to the lack of a pumping sub-pulmonary ventricle. The most frequent complications are arrhythmias, thromboembolism, protein-losing enteropathy, heart failure, and plastic bronchitis[7]. The pathogenetic mechanisms are not completely understood but they are all connected to chronic systemic venous hypertension. Quality of life and life expectancy are deeply impaired by the onset of such complications[8].

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While searching for options for dealing with the effects of such complex congenital heart problems, mainly desaturation, it was observed that patients with complex cardiac anomalies in addition to patent PDA have better blood oxygenation than those without patent PDA[9].

Depending on the fact that arterial shunts could improve SpO_2 and regarding the restrictions and complications of the Fontan procedure, the idea of performing an arterial shunt like MBT shunt in patients with BDG suffering from cyanosis came up.

The Blalock-Taussig's shunt (BTS) was initially introduced by Alfred Blalock and Vivien Thomas[10], who used the subclavian artery as an anastomosis to create a canine model of PA hypertension. This idea was then adopted as a last-ditch effort to increase pulmonary blood flow in tetralogy of Fallot patients[11].

Over time, a major improvement of the traditional BTS was devised, avoiding the subclavian artery by using a synthetic vascular prosthesis. Compared with the original BT shunt, this modification had a number of benefits, including a decreased propensity to deform hypoplastic PAs, a reduced requirement for mediastinal dissection, the preservation of upper extremity blood flow, a steady shunt flow, and an appropriate length[12]. The MBTS is still the most popular systemic PA shunt in use today because of these important benefits[13]. Additionally, the hepatic factors in the lungs are preserved by MBT shunt following BDG, which lowers the risk of AV malformation[14].

Aim

The study aims to evaluate the mid and long-term effects of left MBT shunt after bidirectional cavopulmonary shunt.

PATIENTS AND METHODS:

Study setting

This study is a retrospective cohort study from January 2023 to December 2023 on 20 patients conducted at the Department of Cardiothoracic Surgery Ain Shams University Hospital.

Inclusion criteria

Patients greater than 5 years old, patients with cavopulmonary shunt operation, patients with sizable left pulmonary artery, patients with normal anatomy of the left subclavian artery, patients with SpO_2 less than 75%.

Exclusion criteria

Patients less than or equal to 5 years old, patients with dysfunction cavopulmonary shunt, patients with other debilitating diseases that may affect life expectancy.

Procedures and tools

Regarding medical records of the patients

Demographics (age, sex, BMI), complete history taking including past medical history of any other illness, general examination including: vital signs, collection of preoperative data: BMI, SpO2, and echocardiography.

Surgical technique of MBTS

The patient is put in the left lateral position for the posterolateral thoracotomy to be done, dissection over the left subclavian artery and the main pulmonary artery and its left branch, vascular clamps are applied and Side to side anastomosis is done using a gortex tube graft of suitable size, removal of vascular clamps and checking for bleeding from the anastomosis, placement of chest tube and primary closure in layers.

Collection of post-operative data

SpO_2, NYHA class, echocardiography, postoperative morbidity, and mortality.

Statistical analysis

Statistical analysis was done using IBM SPSS statistics for windows, Version 20.0. Armonk, NY: IBM Corp. Categorical data were expressed as number and percent. Continuous data were expressed as mean, SD, median, minimum, and maximum. Comparisons of pre-operative and post-operative continuous data were performed using paired-samples T-test. A value of *P* less than 0.05 was considered statistically significant.

RESULTS:

The results are discussed in detail and will evaluate the outcome of mid and long-term effects of left MBT after bidirectional cavopulmonary shunt.

Table 1: Sex of the studied patients

Sex	Number (%)
Male	14 (70%)
Female	6 (30%)

The study included 20 patients underwent MBTS following CPS via thoracotomy approach. There were 14 (70%) males and six (30%) females (Table 1).

The indications of MBTS were hypoplastic left ventricle in 30%, TGA+pulmonary atresia in 15%, D-TGA in 15%, Transposition of great arteries (TGA)+ Double outlet right ventricle (DORV) in 15%, Double inlet left

ventricle (DILV)+ Pulmonary stenosis (PS) in 10%, DORV+ Ventricular septal defect (VSD) in 5%, DORV in 5%, and TOF with hypoplastic left ventricle in 5%.

Preoperative left ventricular ejection fraction (LVEF) ranged between 52 and 65% with a median of 58.50% and a mean of 58.90±4.25%.

ICU stay ranged between 4 and 8 days with a median of 5 days and a mean of 5.40±1.23 days. Postoperative hospital stay ranged between 8 and 22 days with a median of 13.50 days and a mean of 14.35±4.15 days (Table 2, Fig. 1).

Postoperative SpO_2 ranged between 85 and 94% with a median of 89% and a mean of 88.9±2.40% (Table 3).

There was a significant increase in postoperative SpO_2 in comparison to preoperative SpO_2 (*P<0.001*) with a mean difference of 20.95% (Table 4).

The postoperative course was uneventful in the majority of cases. There was no mortality, re-operation, or shunt occlusion. There were minimal postoperative complications reversed on medical treatment in 3 (15%) cases including thrombosis treated by anticoagulants, wound infection, and pneumonia in one patient for each (5%)

Table 2: Durations of postoperative ICU and hospital stays

Statistics	ICU stay (days)	Hospital stay (days)
Mean	5.40	14.35
Median	5	13.50
SD	1.23	4.15
Minimum	4	8
Maximum	8	22

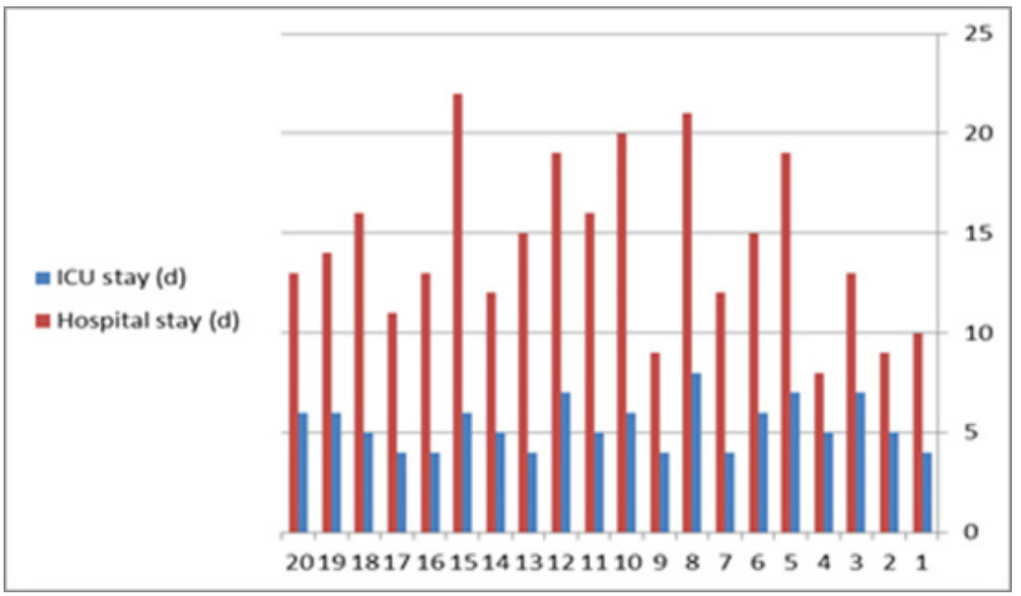

Fig. 1: Postoperative ICU and hospital stay by days for each patient.

Table 3: Postoperative oxygen saturation (SpO_2)

Statistics	Postoperative SpO_2
Mean	88.90
Median	89
SD	2.40
Minimum	85
Maximum	94

Table 4: Change in postoperative oxygen saturation (SpO_2)

Statistics	Postoperative-preoperative SpO_2
Mean difference (%)	20.95
P value	<0.001*

DISCUSSION

Structural anomalies of the heart or intrathoracic great vessels that happen during fetal development are known as congenital heart disorders (CHD). The most prevalent kind of birth abnormality and the main reason why children with congenital abnormalities die is congenital heart disease (CHD)[1]. There are two types of CHD: cyanotic CHD, often known as critical CHD, and noncyanotic CHD[15].

The main problem in cyanotic CHD is desaturation due to the mixing of arterial and venous blood. Cavopulmonary shunts were described as a possible solution for long-standing cyanosis as they can provide another pathway to separate the arterial and venous blood[3].

BDG is one of the cavopulmonary shunts that aims at improving the SpO_2 where the SVC is transected at its attachment with the right atrium and anastomosed to the pulmonary artery[4]. Although BDG relieves the cyanosis for some time, most cases develop desaturation with advanced age, and further intervention is needed[5].

Fontan operation is performed as a second stage of repair and it results in complete separation of pulmonary and venous return[6], but it has many restrictions and complications, especially in the long-term.

However, in the event of a subpulmonic pumping chamber's absence, the unphysiological Fontan circulation leads to a decrease in cardiac output and an increase in central venous pressure, which can lead to a number of short- or long-term problems.

These complications include[16]:

One common cardiac complication in Fontan circulation is atrioventricular valve regurgitation, which has a negative effect on long-term results by raising the likelihood of mortality or heart transplantation[17].

Following Fontan surgery, arrhythmias such as bradyarrhythmia, supraventricular tachyarrhythmia, and atrial tachyarrhythmia are frequently seen. They might result in cardiac failure, thrombogenesis, atrioventricular valve regurgitation worsening, or even abrupt death[18].

Ventricular dysfunction, either diastolic or systolic, is a frequent consequence that worsens long-term mortality and morbidity. There are several contributing factors, including repeated operations, pressure, and volume overloading, and persistent hypoxia[19].

In Fontan circulation FSV patients, thromboembolic events are a major cause of morbidity and death. Despite the lack of clarity around the genesis and etiology of thromboembolic events, endothelial dysfunction, aberrant hemodynamics, and hypercoagulability are the three components of Virchow's triad that are implicated. Endothelial dysfunction specifically refers to the loss of prothrombotic products and vasoconstrictors, along with a rise in prothrombotic factors and antithrombin, Protein C, S, and antithrombin, and vasodilators such as nitric oxide, which ultimately leads to a prothrombotic state[20].

With a remitting and relapse course, protein-losing enteropathy is an uncommon but devastating complication. It is characterized by massive protein loss into the digestive tract, which results in low levels of serum albumin and total protein, hypercoagulability, and immunodeficiency due to enteric loss of lymphocytes and immunoglobulins[21].

Like PLE, plastic bronchitis (PB) is a rare but potentially catastrophic condition that causes cohesive and obstructive bronchial casts to form and expectorate in Fontan patients[22].

Liver fibrosis, cirrhosis, and possibly cancer are among the congestive hepatopathies that make up Fontan-associated liver disease (FALD)[23].

At rest, the kidneys' blood supply normally makes up 20–25% of the heart's output. Here, renal impairment, including acute kidney injury (AKI) and chronic kidney disease (CKD), may result from the Fontan hemodynamic change[24].

In the review conducted by Johanna *et al*. Many patients with Fontan circulation experienced serious long-term complications such as difficulty in areas of cognition related to attention and executive functioning, visual-spatial reasoning, and psychosocial development. They were also at high risk for mental health morbidities, particularly anxiety disorders and depression[25].

It was an interesting remark that patients with patent PDA who did not undergo PDA closure during the cavopulmonary shunt operation had more acceptable saturation than patients with closed PDA, and they did not need further intervention[9]. Depending on the fact that arterial and pulsatile blood supply to the lung can help to avoid severe progressive desaturation, hence came the idea of doing arterial shunts after BDG.

Not all patients who have had a Glenn shunt are candidates for Fontan, in addition to the serious complications already mentioned. This is because many of these patients have significant A-V valve regurgitation, systemic ventricular dysfunction, pulmonary artery hypoplasia, and high pulmonary vascular resistance[26].

Therefore, many approaches were being tested to help individuals with BDG who experienced increasing desaturation increase their SpO_2. Veno-venous collaterals have been reported to worsen the prognosis in these individuals by lowering their arterial SpO_2. Transcatheter closure for those collaterals was carried out in the Doaa *et al*. trial in an effort to raise the SpO_2[27].

Creating radial and axillary arteriovenous (AV) fistulae to augment pulmonary blood flow across the preexisting cavopulmonary shunt was another strategy to aid in increasing SpO_2 following BDG[28].

According to a study by William *et al*., a 13-year-old patient with tricuspid atresia who had a superior vena cava-right pulmonary artery (SVCRPA) shunt at age 3 could benefit from an A-V fistula as a useful way to supplement blood flow to the right lung. This was achieved by increasing the flow through the shunt, giving it a pulsatile character, and ultimately lowering the blood's viscosity[29].

Patients with complicated cardiac abnormalities who have a patent PDA tend to have greater SpO_2 than those who do not. Furthermore based on earlier research mentioning the benefit of A-V fistulae in raising SpO_2 in patients who had previously had cavopulmonary shunts. The notion of executing MBTS subsequent to the BDG was raised.

Previously referred to as the standard or classic BT shunt, the MBTT shunt is a palliative surgical operation used to treat patients with cyanotic cardiac disorders defined by reduced pulmonary artery flow[30].

The goal of the BT shunt is to deliver enough blood flow to the pulmonary artery to alleviate cyanosis without causing pulmonary over circulation. Through a lateral thoracotomy, the subclavian artery was divided and anastomosed end-to-side to the pulmonary artery in the standard BT shunt surgery. After numerous modifications, the initial procedure became the MBTT shunt, which creates a systemic-pulmonary shunt without compromising any of the brachiocephalic tributaries or the subclavian arteries by using an interposition polytetrafluoroethylene (PTFE) graft[30].

The benefits of pulmonary artery shunts were listed in the review paper written by Pankaj *et al*. as follows[31]:

Enhancement of hematological parameters and systemic SpO_2: The creation of an axillary arterio-venous fistula for the purpose of correcting hypoxemia after a first BDG was documented by Glenn and Fenn in 1972[29]. After a BDG, patients who have a systemic to pulmonary artery shunt have higher systemic SpO_2 due to increased pulmonary blood flow, increased recruitment of the pulmonary capillary bed, and decreased pulmonary vascular resistance. This leads to a reduction in the risk of cerebral and peripheral venous thrombosis, a progressive decrease in hematocrit, blood viscosity, and coagulopathy, and an increase in SpO_2 from 8% to 25%[32].

Increase in pulmonary artery size

Ishikawa *et al*.[33] have demonstrated significant increases in pulmonary artery size on the ipsilateral side following a BTS, especially if the shunt was performed within the first year of life, despite conflicting data regarding the increase in pulmonary artery size following the creation of a systemic pulmonary artery shunt. It is unclear why many patients who have had a systemic to pulmonary shunt do not see a significant increase in the size of their pulmonary arteries after the procedure, but it is plausible that a relatively small shunt is not able to pump enough blood into the pulmonary circulation to match the normal cardiac output. An innate flaw in the pulmonary arteries or the integration of ductal tissue inside the pulmonary artery are two other potential causes.

Regression of pulmonary arteriovenous collaterals (malformations; AVM)

The majority of patients experience pulmonary AVM 5–10 years after BDG; this results in a worsening of pulmonary function and an increase in cyanosis. The exact cause of these AVMs is unknown, but it is thought to be caused by a lack of hepatic factor circulating through the lungs after antegrade pulmonary blood flow is interrupted after a BDG[34]. According to a study by Doff B. *et al*., these AVMs regress and are prevented from growing further[35].

Improvement in ventricular function

Six patients who had undergone a BDG before and were deemed inappropriate for TCPC because of ventricular dysfunction were palliated using a brachial AV fistula in the Quarti *et al*. research[36]. After a brachial AV fistula was created, all six patients' ventricular function improved during follow-up, and they all underwent successful TCPC procedures within 6 years. It is unclear how precisely the systemic to pulmonary artery shunt in these individuals improved ventricular function. Ventricular function may have improved, nevertheless, because of factors such as decreased blood viscosity from a lower hematocrit, increased myocardial preload, and increased SpO_2 from better oxygen delivery to the myocardium[36].

Out of 320 children with cyanotic congenital heart malformations who had previously undergone cavopulmonary shunt operations, 11 were deemed unsuitable for definitive repair, a Fontan procedure, or other palliation due to their increasing cyanosis and exercise intolerance, as reported by A Magee *et al*.[35]. Three individuals had a prior bidirectional cavopulmonary connection, and eight had a prior Glenn shunt. Ten patients underwent ipsilateral axillary AV fistula formation to increase pulmonary blood flow. Before surgery, mean SpO_2 were 80% +/-2%, immediately after surgery, they were 85% +/-2%, and after a mean follow-up interval of 7.4 years (range 0.1–15.5 years), they were 84% +/-3%. This suggested that when alternative choices are limited, establishing an axillary AV fistula to increase pulmonary blood flow following a cavopulmonary shunt offers helpful palliation for complicated cyanotic heart disease.

In this study, we reported the mid- and long-term postoperative outcomes, particularly SpO_2, after we retrospectively evaluated the data of 20 patients who underwent BDG in our hospital and received MBTS.

The preoperative SpO_2 had a mean of 67.95±3.56% with a range of 60–73%. The median was 68.50%. The range of postoperative SpO_2 was 85–94%, with a mean of 88.9±2.40% and a median of 89%. With a mean difference of 20.95%, there was a substantial rise in postoperative SpO_2 compared with preoperative SpO_2 ($P < 0.001$). Therefore, it was statistically significant that in patients who have had a prior cavopulmonary shunt and are not candidates for other procedures or cannot tolerate their consequences, left MBTS effectively raises SpO_2.

Worldwide, a midline sternotomy has become the standard procedure for managing MBTS. It is also said that doing a MBTS via thoracotomy is technically more difficult. On the other hand, thoracotomy surgery has been suggested to put patients at lower risk for wound infections, postoperative respiratory impairment, shorter hospital stays, and cosmetic issues.

The sole side effect reported in the A Magee *et al*. research[35] was a patient's slight arm edema distal to the fistula; this patient's distal vein was left unlocated and she needed another procedure after 19 days.

Most of the patients in our dataset had unremarkable postoperative courses. No shunt obstruction, death, or re-operation occurred. Three (15%) cases had modest postoperative problems that were resolved with medical treatment; they included one (5%) patient's, wound infection, and pneumonia, as well as thrombosis treated with anticoagulants.

CONCLUSION

Congenital cyanotic heart disease represent a life-threatening condition where the main problem is desaturation due to the mixing of arterial and venous blood. BDG is a step to separate the oxygenated and deoxygenated blood. But later on, SpO_2 continues to decline and further intervention is needed. In this study, we found that MBTS provides a post-operative significant increase in SpO_2 in a wide group of those patients even who are not candidates for other operations without significant complications.

CONFLICT OF INTEREST

There are no conflicts of interest.

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