SURGICAL AORTOPULMONARY SHUNTS- A THIRTY-SEVEN YEAR EXPERIENCE IN A SOUTH AFRICAN TERTIARY INSTITUTION

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A research report submitted to the Faculty of Health Sciences, University of the Witwatersrand, Johannesburg, in partial fulfilment of the requirements for the degree of Master of Medicine in Paediatrics

Johannesburg 2019

Declaration

I, Nontobeko Charity Dladla-Mukansi declare that this research report is my own work. It is being submitted for the degree of Master of Medicine in Paediatrics at the University of the Witwatersrand, Johannesburg. It has not been submitted before for any degree or examination at this or any other University.

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The 13th of November 2019

Dedication

I dedicate this work to my parents Anthony and Hlengiwe Dladla, who have dedicated their lives for the benefit of their family. I thank God for my parents' unconditional love and support.

ABSTRACT

Introduction

The surgical aortopulmonary shunt is a valuable palliative procedure in the management of congenital heart diseases. There is a paucity of data regarding aortopulmonary shunts in the developing world, including South Africa.

Objectives

The primary objective was to describe the demographic, clinical and echocardiographic characteristics of children between ages 0 and 14 years that underwent surgical aortopulmonary shunts. The secondary objectives were to describe trends in aortopulmonary shunt designs, outcomes in terms of morbidity and mortality, progression to definitive surgery and to assess patency of shunts.

Material and Methods

A retrospective clinical audit of patient files who underwent an aortopulmonary shunt between 01 January 1980 to 30 December 2016 was undertaken at Chris Hani Baragwanath Academic Hospital (CHBAH) in Soweto, Johannesburg. The study period was divided into 3 stages and for descriptive purposes as follows: 1980-1991 refers to period 1, 1992-2003 refers to period 2 and 2004-2016 refers to period 3.

Results

A total of 177 aortopulmonary shunts were done over the 37-year study period. Of these 177 patients, 165 (93.2%) patient files were available.

Fifty-six percent of the patients included in the study were male. The majority of patients were from the Gauteng Province (76.8%). The four most common diagnoses across the entire study period were tricuspid atresia (26.0%), pulmonary atresia with VSD (23.7%), tetralogy of Fallot (23.2%) and complex cardiac lesions (16.9%), with no particular trend in the proportion of these diagnoses presenting across this study period. There was no statistical difference between period 1 and 2 (p-value a=0,328) and between period 1 and 3 (p-value b=0,548).

The total number of all surgeries done over the entire study period was 2145, of which 8.3% were aortopulmonary shunts. Period 1 had the highest percentage [35 (10.9%)] of aortopulmonary shunts compared to the total number of surgeries performed. There was a decline in the number of aortopulmonary shunts performed over the study periods 1-3. With no statistical difference across periods as shown in table 1 with p-value a and b.

Of the different types of aortopulmonary shunts, most patients [157 (88.7%)] had a modified Blalock-Taussig shunt (BTS). The remainder of the shunts included 3 (1.7%) classic BTS, 12 (6.8%) central shunts and 5 (2.8%) unknown BTS. The percentage of modified BTS done increased from 80% in period 1 to 87.3% in period 2 and to 95.2% in period 3.

Period 1 had the most complications (28.6%) compared to 11.4% in period 2 and 19.1% in period 3. Sepsis as a complication following surgery increased over the study period from 2.9% in period 1 to 3.8% and 7.9% in periods 2 and 3 respectively.

Early mortality was 17.1%, 26.6% and 25.4% from periods 1-3 respectively. Late mortality declined from 17.0% in period 1 to 11.4% and 0% in periods 2 and 3 respectively. Only 37 (20.9%) patients were documented to have further surgery after the initial aortopulmonary shunt. Across all three study periods, no blocked shunts were documented.

Conclusions

This study describes the characteristics and outcomes of aortopulmonary shunts over a 37-year period in a tertiary care resource limited low to middle income country setting. The commonest cardiac lesions for which aortopulmonary shunts are performed are tricuspid atresia, pulmonary atresia with VSD, tetralogy of Fallot and other complex cyanotic cardiac lesions. The frequency of aortopulmonary shunts compared to total surgeries has decreased over the study period, which may imply a trend towards early corrective surgery for these cardiac lesions. The modified BTS is the most frequently performed aortopulmonary shunt used for palliative surgery in our setting, which is a similar trend in developed countries.

The morbidity and mortality in this study is higher than developed countries, with sepsis being the most common complication. Attention to infection control practises need to be emphasized peri- and post-operatively in our hospitals.

ACKNOWLEDGEMENTS

I gratefully acknowledge and thank:

. Almighty God for granting me the strength and perseverance to commence and complete this research project.

. My supervisors, Professor Cilliers, Dr Vijay Mammen and Dr Kathy Vanderdonk.

. All patients whose data contributed to this study.

. The Medical Advisory Committee and the management of Chris Hani Baragwanath Academic Hospital for allowing me to conduct this study.

. The staff of the Records Department at Chris Hani Baragwanath Academic Hospital for their assistance.

. The staff of the Faculty of Health Sciences of the University of the Witwatersrand, particularly the Department of Paediatrics and Child Health.

TABLE OF CONTENTS

List of Tables

ABBREVIATIONS

BTS = Blalock Taussig shunt

CHF = Congestive heart failure

CMJAH = Charlotte Maxeke Johannesburg Academic Hospital

CHBAH = Chris Hani Baragwanath Academic Hospital

HREC = Human Research Ethics Committee

ICU = Intensive care unit

LV = Left ventricle

MBTS = Modified Blalock Taussig shunt

PA = Pulmonary artery

PHT = Pulmonary hypertension

PR = Pulmonary regurgitation

TA = Tricuspid atresia

TOF = tetralogy of Fallot

PDA = Patent ductus arteriosus

1.0 INTRODUCTION

The surgical aortopulmonary shunt is a valuable palliative procedure in the management of congenital heart diseases (CHD) [1]. The procedure has been successfully performed on cyanosed children since 1944, with countless patients having benefited from this procedure [1,2]. It is indicated in patients with congenital heart disease associated with decreased pulmonary blood flow if corrective surgery cannot be performed immediately [3]. The common types of CHD in which this procedure is indicated are tetralogy of Fallot, tricuspid atresia, pulmonary atresia, Ebstein anomaly and certain types of single ventricle patients [1,3,4]. Approximately 21 different palliative procedures for the surgical management of congenital heart defects have been described [5].

Palliative surgeries are being done less frequently in the modern era due to improved extracorporeal circulation methods, modern perioperative supportive care and improvement in operative techniques allowing for full correction of complex cardiac lesions in small children. [3]. The procedure is technically challenging and to be done safely, requires an experienced appropriately trained paediatric heart surgeon [5,6].

There are 5 main types of shunts which are described below and that have evolved over time since the first operation that was designed by Drs Blalock and Taussig and perfected by their laboratory technician Mr Vivian Thomas [7].

The five main types of shunts:

1.) The original shunt or classic Blalock-Taussig Shunt (BTS) requires transection of the subclavian artery and its anastomoses to the ipsilateral pulmonary artery (PA) through a thoracotomy approach. Most commonly a right-sided shunt is recommended for a left sided aortic arch and a left-sided shunt is recommended for a right-sided aortic arch [8,9]. An advantage of the original classic shunt is that it provides continuous blood flow. In addition, because the tissue used is the patient's own subclavian artery, the vessel grows with the patient. It was previously recommended for infants older than 3 months and is less frequently done now because it sacrifices the subclavian artery resulting in a small risk of ischaemia and shortening of the affected arm. This shunt is seldom used in the modern era [4].

2.) A modified version of the original shunt was introduced following the observations of the disadvantages of the classic BTS. This modified BTS (MBTS) is now preferred by most surgeons. It was first reported by Gazzaniga and associates in 1976 [4]. It is the preferred BTS for patients younger than 3 months though it can be done at any age. It is regarded as one of the more eminent palliative procedures for congenital heart defects and has proven to have a highly satisfactory long-term clinical application [5]. The surgery for the MBTS involves the placement of a Gore-Tex interposition graft between the subclavian artery or innominate artery and ipsilateral pulmonary artery (PA) [9,10].

   The advantages include preservation of the circulation to the affected arm, regulation of the shunt flow by the size of the systemic artery to

which it is sutured, a high early patency rate with minimal tissue ingrowth of even a small-diameter expanded Gore-Tex, and finally an adequate shunt length is guaranteed. The procedure is occasionally complicated by leakage of serous fluid through the Gore-Tex material, which may lead to prolonged chest tube drainage [6].

The surgical approach in modified BTS is a left or right thoracotomy or midline sternotomy [2,8]. In the last 10 years, the median sternotomy has been the preferred surgical access because of the technical difficulties in dividing the shunt at the time of corrective surgery [4].

3.) The Waterson shunt procedure involves the placement of a connection between the ascending aorta and the right PA via sternotomy or right thoracotomy [11]. It is seldom used by congenital heart surgeons due to the high incidence of surgical complications usually related to the poor control of the size of the shunt inserted. It invariably leads to excessive pulmonary blood flow with the danger of causing pulmonary hypertension [8,9].

4.) The Potts-shunt operation is another variation that is seldom used in the modern era, whereby an anastomosis from the descending aorta to the left pulmonary artery is created via a left thoracotomy. It also has the potential to cause an increased in pulmonary blood flow and cause pulmonary hypertension as does the Waterston shunt [11].

5.) The 5^{th} variation is a central shunt that is mostly used in patients with very small pulmonary arteries. A Gore-Tex tube is connected between the ascending aorta and the main pulmonary artery [3].

The BTS is the most common aortopulmonary shunt that is performed in the modern era, with the central shunt type being used when there are small branch pulmonary arteries [1,12]. The classic BTS, Waterston shunts and Potts shunts are currently seldom used due to their associated complications. [4,8,9,11,15-17]. In the recent times, the BTS is being reserved in western countries as an emergency measure in children who are very ill and not able to tolerate full correction using cardiopulmonary bypass support [1,18].

The largest retrospective review of BTS surgery was done at a single centre over 6 decades in Baltimore (USA), from November 1944 to December 2006. The study compared two different eras from 1940 to 1970 and 1970 to 2000. They showed that 2016 BTS were performed on 1880 patients originating from 35 different countries. Data was available for 94.4% (1774 of 1,880) of patients. The cardiac diagnosis in most patients was TOF (72%) although the diagnosis was acknowledged to be imprecisely recorded in the early part of the study. A small number of patients (9%) had a single ventricle diagnosis and underwent an initial BTS followed by a succession of other palliative procedures [1].

They showed a mean annual decrease of BTS surgery (66 per year in the first era and 9 per year in the second era) due to full corrective surgery becoming possible in younger children. This procedure was still used frequently in patients with single ventricle anatomy with the proportion of single-ventricle application of the BTS increasing (5% versus 34%) between the two study periods [1].

The BTS design of the series included classic BTS in 75% patients, a modified BTS in 7% patients (which only gained acceptance in the 1970's), 9% unknown and 8% other [1]. As surgical techniques improve and experience with total correction of tetralogy of Fallot has grown, surgeons have opted to undertake total correction earlier in life, using the BTS as a palliative choice for neonates, extremely low birth weight babies, those who are unstable at the time of presentation or those with lesions that mitigate against early total correction. [1].

In addition, operative mortality decreased between the two study periods (16% versus 9%). The overall operative mortality for all BTS patients (first and successive shunts) was 14%. The operative mortality decreased to 11% for a second BTS and 0% for a third BTS. Of note, the incidence of prolonged intubation, sepsis, and wound infection increased in the latter stages of the study (1990's and 2000's) reflecting the increased complexity of patients undergoing BTS during this decade. The most common cause of death was heart failure and haemorrhage, mainly in the early period of the study, followed by cardiac arrest, stroke, and brain abscess [1].

In resourced challenged countries, patient circumstances and poor socioeconomic conditions may dictate the type of surgery undertaken. These patients commonly present to hospital at a much older age and if they live in areas remote from a central hospital transport delays can affect their outcome [9,19,20-21].

A retrospective review of BTS surgery performed in Libya from May 1992 to May 1998 showed that during the 6-year period, 100 MBTs were performed on 94 children with a median age of 12 months. Out of 100 MBTs, 89 were done using the left-sided shunts and 11 had right-sided shunts. Interposition tubes of 6 millimetres in diameter were used in 68 cases and 4 millimetres in 32 cases [12].

Sixty-three (76%) patients in this study had tetralogy of Fallot. The remaining 31 patients (33%) included 13 (41%) tricuspid atresia, 9 (29%) pulmonary atresia, 6 (19%) univentricular heart lesions and 3 (9%) other diagnoses. The early operative mortality prevalence was 6% (n=6). The risk factors for operative mortality were patients in the neonatal period, diagnosis other than tetralogy of Fallot and emergency surgery. The late mortality or deaths were 12 patients, of which 6 were due to sepsis [12].

The causes of early death reported in this article from Libya included acute shunt thrombosis, congestive cardiac failure, pulmonary oedema secondary to sudden increase in pulmonary flow, kinking and distortion of the PA resulting in hypoxia, ventricular fibrillation, and sepsis which is similar to the publication by Krassermann et al [21].

Late complications comprise infective endocarditis, pseudo-aneurysm, and late shunt failures [12]. Other complications of BTS observed in Switzerland include

chylothorax, phrenic palsy, necrotising enterocolitis, abdominal bleeding, and residual stenosis post-reconstruction [22]. Generally, the complication rate in developing countries compares favourably with first world countries [1].

A study from China undertaken between January 1986 and December 1991, documented 100 MBTS where polytetrafluoroethylene grafts were used in 86 patients with complex cyanotic congenital cardiac malformations. Patient ages ranged from 15 days to 22 years. Thirty-seven were operated on in infancy and one in the neonatal period. Shunt diameters varied from 3-8 millimetres. The mean follow-up period was 13.5 months (range, one to 50 months). There was one hospital mortality and one morbidity [23].

A univariate analysis from the China study showed that the age of the patient and graft diameter were risk factors for shunt blockage. Neonates were found to be at high risk of shunt blockage. Nevertheless, despite technical, perioperative care and surgical challenges, MBTS continue to be used in the neonatal group [23].

The only study that documented BTS surgery in South Africa was undertaken between 01 January 1994 and 31 December 2013 at the Charlotte Maxeke Johannesburg Academic hospital (CMJAH). The objective of the study was to describe the outcomes of children diagnosed with tetralogy of Fallot who underwent surgical repair with an emphasis on post-operative pulmonary regurgitation. The median age of corrective surgery was 39.5 months. Fifty-

four (75%) of the patients with tetralogy of Fallot had surgery, of whom 50 (92.7%) had complete repair. Of these patients who had surgery, two underwent a prior BTS and 4 (7.4%) had a BTS as the only operation. The BTS surgery was done at an older age in this study, with a median age of 3.2 months (p-value=0.973). Reasons for delayed surgery included lack of resources, poor health care worker recognition of the cardiac lesions and delayed referral from peripheral hospitals to the tertiary centre [24,25].

Recommendations from the above-mentioned studies from Libya and South Africa were to prioritise awareness campaigns for congenital cardiac conditions, to encourage the use of echocardiography to assist in their diagnosis. In addition, the Libyan study states that the need for palliation is even greater in developing countries because of lack or limitation of resources, late presentation, and late referral of patients [12]. The study also recommended that a classic BTS should be considered for patients who are deemed noncompliant and of poor social background [12].

The South African study, despite the small sample, showed that the operative mortality in patients undergoing BTS compared favourably to other first world centres [1,18,24,26].

In conclusion, studies have shown that the patency of the BTS shunt is largely dependent on the diameter chosen [6,8,9,10]. Experimentation has shown that 3.5 millimetres is the optimal diameter for a term neonate weighing more than

3.0 kilograms [9]. There is a risk of thrombosis, acute obstruction in the post shunt period and the child outgrowing the shunt if a smaller diameter is used.

A diameter more than 4 millimetres in a term infant may result in excessive pulmonary blood flow leading to increased pulmonary venous return to the left atrium and left ventricle which can cause left heart overload, heart failure and prolongation of the intensive care unit (ICU) stay [12,19,22].

In summary, the BTS is used in both developing and developed countries as a palliative procedure for patients with reduced pulmonary blood flow conditions. [12,17,24]. The indications for and types of aortopulmonary shunts, however, have changed over time [1]. We sought to review our institutional experience, in a country that has evolved from low-middle to upper-middle income, with regards to aortopulmonary shunts over the last 27 years.

2.0 MATERIALS AND METHODS

A retrospective clinical audit of patient files who underwent a palliative aortopulmonary shunt between 01 January 1980 to 30 December 2016 was undertaken. The Paediatric Cardiology computerized database at the Chris Hani Baragwanath Academic Hospital (CHBAH) which is the referral institution and the Paediatric Cardiac Surgical database at the Charlotte Maxeke Academic Hospital (CMJAH) which is the surgical institution were sourced for patient information.

Permission was obtained from the Human Research Ethics Committee (Medical) of the University of the Witwatersrand (Ethics committee number: M170409) and CMBAH Medical Advisory Committee to conduct the study. Permission to use the databases was provided by Professor Antoinette Cilliers and Dr Kathy Vanderdonck respectively. Missing information was obtained from the cardiac clinic patient files and surgical reports. Informed consent was not required because of the retrospective nature of the study.

The study period was divided into 3 stages between from 1980 to 2016. And for descriptive purposes as follows; 1980-1991 refers to Period 1, 1992-2003 refers to Period 2 and 2004-2016 refers to Period 3 of the study.

DEFINITIONS

Early mortality: day one to 6 months' post operation

Late mortality: more than 6 months' post operation

Complex cardiac patients: double outlet right ventricle, Ebstein anomaly, single ventricle with pulmonary stenosis and pulmonary atresia

Other cardiac diagnosis: hypoplastic right ventricle, dextrocardia and any other congenital heart defect.

Follow-up: Patients seen at 10 years after initial cardiac surgery

2.1 STATISTICAL METHODS

Data extracted from the databases, patient files and surgical reports included demographic data consisting of age at first visit, sex, diagnosis and the province of referral. The second category of data collected were details of the surgery including age at surgery, type of Blalock-Taussig shunt, size and type of conduit used for the shunt, and length of ICU stay. The third category of data included post-operative complications, mortality, and patency of the shunt.

Counts and proportions are reported for categorical variables and a Fischer's Exact test used to compare categorical variables between groups. Medians and IQR are reported for quantitative variable and the Wilcoxon Rank Sum test used to compare these variables between groups.

Microsoft Excel was used to collect and store the data (Microsoft Office 2016) and the statistical package R (v3.5) was used for all analyses.

3.0 RESULTS

3.1 Primary objectives:

3.1.1 Demographics

There were 2145 total surgeries that were referred from CHBAH to the Paediatric Cardiac Surgical unit at CMJAH over the 37-year period of which 177 (8.3%) were surgical aortopulmonary shunts as shown in Table 1. Of the 177 patients, 165 (93.2%) patient files were available. Notably, the number of BTS procedure in increased in the last 2 periods of the study, although there was a decreased in the percentage of BTS procedure performed when comparing to total number of surgeries performed. Most patients were from the Gauteng Province (71.1%). Fifty-four percent of the patients included in the study were male.

3.1.2 Clinical

Fifty-seven (32.2%) patients presented with oxygen saturations < 75%, 53 (29.9%) patients presented with saturations between 75-90% and 2 (1.1%) presented with saturations >90%. Fourteen patients (7.9%) were intubated on arrival. One hundred and thirteen (63.8%) patients were cyanosed and 45 (25.4%) of those had digital clubbing. There were 10 (5.6%) patients with subtle dysmorphic features but only 4 had confirmed syndromes of whom two patients had Trisomy 21, one had Noonan syndrome and one had DiGeorge syndrome.

3.1.3 Echocardiographic data

In Period 1, 321 surgeries were referred from CHBAH and 35 (10.9%) were aortopulmonary shunts. Most patients had pulmonary atresia with VSD (22.8%) and tetralogy of Fallot (20.0%) and complex cardiac lesions (20.0%). The main

complications were pleural effusions (5.7%), convulsions (5.7%), over-shunting (5.7%) and sepsis (2.9%). Six patients died, with early mortality calculated to be 17.1% and the late mortality being 17.0%. Time from diagnosis to surgery was an average of 12 months.

In Period 2, 902 surgeries were referred from CHBAH and 79 (8.8%) were aortopulmonary shunts. Most patients had tricuspid atresia (30.4%), tetralogy of Fallot (27.8%) and pulmonary atresia with VSD (25.3%). The main complications were sepsis (3.8%), phrenic nerve palsy (2.5%) and over-shunting (2.5%). Twenty-one patients died with an early mortality of 26.6% and a late mortality of 11.4%. Time from diagnosis to surgery was an average of 4 months.

In Period 3, a total of 922 surgeries were referred from CHBAH and 63 of these (6.8%) were aortopulmonary shunts. Most patients were diagnosed to have tricuspid atresia (27.4%), pulmonary atresia with VSD (24.2%) and complex cardiac lesions (22.6%). The most common complications were sepsis (7.9%), convulsions (4.8%) and over-shunting (1.6%). Sixteen patients died with an early mortality of 25.4% and a late mortality of 0%. Time from diagnosis to surgery was average of 7.5 months.

3.2 Secondary objectives:

3.2.1 Trends in aortopulmonary shunts designs

Table 1 demonstrate the numbers and trends of surgical aortopulmonary shunts performed over the 3 periods of the study. As shown by p-value a and p-value b there was no statistical difference in terms of the trend of type of shunt done the most across three periods. The number of modified BTS undertaken was 157/177 (87.6%). Eighty-five (54.1%) were done through a left thoracotomy, 45 (28.7%) were done through a right thoracotomy and 9 (5.7%)

were done through a sternotomy. The type of approach for the remaining 18 (11.5%) modified BTS were not documented.

The remaining shunts 20/177 included 12 (6.8%) central shunts, 5 (2.8%) unknown BTS and 3 (1.7%) classic BTS.

The central shunts were mainly done in cases where the branch pulmonary arteries were very small. There were no documented cases of Waterston or Potts shunts.

Eighteen patients had a 2nd aortopulmonary shunt procedure in this study. There were 8 patients in period 1; followed by 8 patients in period 2 and 2 patients in period 3. All eighteen patients had modified BTS. There were no complications documented in these patients.

3.2.2 Assessment of the patency of shunts

There were no blocked shunts documented in the entire study.

3.2.3 A progression to definitive surgery and follow up

A total of 37/177 patients underwent corrective surgery after receiving their aortopulmonary shunt. The Glenn procedure was the most common corrective operation performed (Table 3). Many patients were lost to follow up (34.4%) and not seen 10 years' post BTS and this may be the reason for so few patients having corrective surgery.

3.2.4 Outcomes in terms of mortality and morbidity

Median postoperative length of hospital stay was an average of 10 days (p-value 0.124) for the study period (Table 2). The overall mortality for all BTS patients was 35.4%, with tricuspid atresia being the most common diagnosis in patients who died (35.3%) compared to other conditions as shown in table 4.

Table 1: Demographic and clinical characteristics of aortopulmonary shunts

Variable	Total	Period 1	Period 2	*p-value a	Period 3	**p-value b
N (%)	177(100)	35 (19.8)	79 (44.6)		63 (35.6)	
Median (IQR) age at presentation (months)	3.3 (0.5-12.4)	3.5 (1.4-8.7)	2.7 (0.3-13.1)	0,592	3.8 (1-11.7)	0,937
Mean (SD) age at presentation (months)	14.5 (27.1)	14.9 (30)	14.1 (26.8)	0,894	15 (26.3)	0,989
Male n (%)	99 (55.9)	22 (62.9)	43 (54.4)	0,527	34 (54.0)	0,523
Diagnosis n (%)				0,328		0,548
Tricuspid atresia	47 (26.6)	6 (17.1)	24 (30.4)		17 (27.4)	
Pulmonary atresia with VSD	43 (24.1)	8 (22.8)	20 (25.3)		15 (24.2)	
Tetralogy of Fallot	40 (22.5)	7 (20.0)	22 (27.8)		11 (17.7)	
Complex cardiac lesion	29(16.4)	7 (20.0)	8 (10.1)		14 (22.6)	
Transposition of great vessels	5 2.8	2 (5.7)	2 (2.5)		1 (1.6)	
Other	7 (3.9)	0 (0.0)	3 (3.8)		4 (6.5)	
Unknown	6 (3.3)	5 (14.3)	0 (30.4)		1 (27.4)	
Province n (%)				0,858		0,093
Gauteng	126 (71.1)	26 (74.3)	58 (73.4)		42 (66.7)	
Free State	12 (6.8)	4 (11.4)	5 (6.3)		3 (4.8)	
North West	9 (5.1)	0 (0.0)	2 (2.5)		7 (11.1)	
Limpopo	7 (4.0)	2 (5.7)	3 (3.8)		2 (3.2)	
Mpumalanga	6 (3.4)	0 (0.0)	2 (2.5)		4 (6.3)	
KwaZulu-Natal	4 (2.3)	1 (2.9)	2 (2.5)		1 (1.6)	
Unknown	13 (7.3)	2 (5.7)	7 (8.9)		4 (6.3)	
Type of Shunt n (%)						0,179
Central	12 (6.8)	3 (8.6)	7 (8.9)		2 (3.2)	
Classic	3 (1.7)	2 (5.7)	0 (0.0)		1 (1.6)	
Modified	157 (87.6)	28 (80.0)	69 (87.3)		60 (95.2)	
Unknown	5 (2.8)	2 (5,7)	3 (3.8)	0,148	0 (0)	
Median (IQR) length of stay in days (ICU stay)	10 (5-13)	11 (10-12)	10 (5-13)	0,615	5.5 (3.8-11)	0,124
Mean (SD) length of stay in days (ICU stay)	11.1 (9.4)	11.2 (1.3)	12.6 (11.1)	0,535	7.9 (6.8)	0,135
Early mortality n (%)				0,621		0,794
Alive	127 (71.8)	23 (65.7)	57 (72.1)		47 (74.6)	
Died	43 (24.2)	6 (17.1)	21 (26.6)		16 (25.4)	
Unknown	7 (3.9)	6 (17.1)	1 (1.3)		0 (74.6)	
Late mortality n (%)				0,741		<0.001
Alive	73 (41.2)	7 (20.0)	15 (18.9)		51 (80.9)	
Died	15 (8.46)	6 (17.0)	9 (11.4)		0 (0.0)	
Lost to follow	61 (34.4)	18 (51.4)	42 (53.2)		1 (1.6)	
Unknown	28 (15.8)	4 (22.6)	13 (16.4)		11 (17.5)	
Median (IQR) age at operation (months)	5 (1.5-21.1)	14.3 (3.8-34.4)	2.6 (0.8-6.9)	<0.001	7.9 (1.9-25)	0,201
Median (IQR) time between diagnosis and operation (months)	0.5 (0.1-2.1)	1.3 (0.3-12.9)	0.3 (0.1-0.6)	0,001	0.8 (0.1-3.4)	0,305

*p-value a: compares Period 1 (1980-1991) to Period 2 (1992-2003), ** p-value b: compares Period 1 (1980-1991) to Period 3 (2004-2016)

Table 2: Mean age at operation and average time between diagnosis and operation

Period	Mean age operation	Average time between diagnosis and operation
1	48 months	12 months
2	16 months	4 months
3	24 months	7.5 months
Total	**24 months**	**6.2 months**

Table 3: Repair/Corrective Procedure

Period	1 N (%)	2 N (%)	3 N (%)	Total N (%)
Glenn	4 (33.3)	10 (52.6)	3 (50.0)	17 (45.9)
TOF repair	4 (33.3)	4 (21.1)	1 (16.6)	9 (24.3)
Rastelli	3 (25.0)	3 (15.8)	1 (16.6)	7 (18.9)
Fontan	0 (0)	2 (10.5)	1 (16.6)	3 (8.1)
Pulmonary valvotomy	1 (8.3)	0 (0.0)	0 (0.0)	1 (2.7)
Total	**12 (32.4)**	**19(51.4)**	**6 (16.2)**	**37 (100)**

Table 4. Deaths according to diagnosis

Diagnosis	No of deaths (%)
Tricuspid atresia	18 (35.3)
Pulmonary atresia with VSD	14 (27.5)
Tetralogy of Fallot	8 (15.7)
Transposition of great vessels	4 (7.8)
Double outlet right ventricle	2 (3.9)
Ebstein anomaly	2 (3.9)
Single ventricle with PS	2 (3.9)
Dextrocardia	1 (2.0)
Total	**51 (100)**

4.0 DISCUSSION

The study reviewed aortopulmonary shunts undertaken over a 37-year period at a large tertiary referral centre in South Africa. The four most common diagnoses across the entire study period were tricuspid atresia (26.6%), pulmonary atresia with VSD (24.3%), tetralogy of Fallot (22.5%) and complex cardiac lesions (16.4%), with no trend in the proportion of these diagnoses presenting across the study period. Although the number of aortopulmonary shunts were higher in period 2 [79 (8.8%)] and 3 [63 (6.8%)], there was a decrease in the percentages compared to the total surgeries over the study period. There was no statistical difference between period 1 versus 2 and 1 versus 3 as indicated by p-value a and b in table 1. Most patients receiving aortopulmonary shunts underwent the modified Blalock-Taussig shunts (BTS) design (91.3%). Patients in Period 1 had the most complications (28.6%) compared to 11.4% in period 2 and 19.1% in period 3. Sepsis as a complication increased over the study period from 2.9% in period 1 to 3.8% in period 2 and 7.9% in period 3.

The frequency of congenital heart lesions needing BTS was very similar across the different study periods. The more common lesions included tricuspid atresia, pulmonary atresia with VSD and tetralogy of Fallot, with each group comprising 24-26% of the total number. In contrast, a study from Baltimore in the USA, showed TOF to be the main diagnosis in 72% of patients over 6 decades [1]. Similarly, reports from other developing countries such as China, Libya, Kenya, and Turkey showed TOF to be the commonest cyanotic condition [13,23,32,33]. The reason for this difference in our study compared to the literature could not be explained but could be related to genetic or ethnic differences in our study population.

The modified BTS has been the preferred surgical aortopulmonary shunt of choice with a frequency of (91.3%), which is higher compared to central shunts (7.0%) and classic shunts (1,7%). The earlier generation of shunts such as the Waterston and Potts shunts which are fraught with complications were not documented in the study cohort. The exclusive use of the MBTS and central shunts in our study, is in keeping with the trend in both developing and developed countries [1,3,12,23].

The total number of all surgeries referred over the study period from Baragwanath was 2145, of which 8.3% were aortopulmonary shunts and the frequency of these shunts increased in period 2 and decreased again in period 3. Period 1= 19.8%, period 2= 44.6% and period 3= 35.6%. This trend of shunts being performed in the modern era is similar to other institutions in the western world where with the advancement of surgical skills, palliative surgery has been replaced by complete repair of TOF and other cardiac lesions, which has yielded better outcome [1,6,8,12,27,28,29,30,31].

There was limited data regarding shunt diameter versus patient weight in this study. However, no blocked shunts were recorded, suggesting that adequate shunt sizes were selected for patients. A minority of the study patients were diagnosed to be “overshunted” suggesting a trend to oversizing shunts at our institution to avoid undersizing and possible blockage of shunts. The poor documentation of shunt sizes used in our study is a major limitation preventing a shunt size/patient weight ratio comparison to other studies [6,8,9,10,12,22].

The age for palliation has decreased over time suggesting that patients are presenting earlier, and that operative management of smaller patients has improved over the last 3 decades. Most of the patients were males which is in keeping with the trends in both the developing and developed countries [1,12,24,23].

The total mortality rate was 35.4% which is twice that of the Baltimore study which was reported to be 14%. [1]. The Libyan study showed an overall mortality of 18% over the 6-year study, which if compared to our institution was lower [24]. There was also a high mortality rate in other developing countries like Kenya (25%), Turkey (18.2%), and Pakistan (27.2%) [32,33,34]. Our early mortality was 25.3% higher than late mortality at 10.1% , similar to findings in the Pakistan study even though the main cause of early mortalities were different, with sepsis being the main cause in our study versus shunt thrombosis and occlusion in the Pakistan study [34]. However, there was a decrease in mortality in the last decade of our study showing an improvement in BTS outcomes over time. Where p-value b (<0.001) statistically significantly comparing late mortality period 1 to 3.

An analysis of the causes of morbidity and mortality showed sepsis to be the leading cause of complications, especially in the last period. Sepsis in the Libyan study, which is also regarded as a developing country, was the cause of late mortality in 50% of cases [11]. Our study showed a much lower incidence of sepsis, but sepsis as a complication increased over the study period from 2.9% in period 1 to 3.8% in period 2 and 7.9% in period 3. The reason for this may be similar to the Baltimore study that attributed this increase to the increased complexity of patients undergoing BTS [1].

In our study, a total of 37 patients had further procedures after the BTS. The Glenn procedure was the most done procedure after the initial surgery. The lack of follow-up surgery may be related to the large number of patients (34.4%) that were lost to follow up. The reasons for the poor follow-up of the study patients are possibly three-fold. Firstly, most of our patients are indigent and are subjected to difficult socio-economic conditions that hinder compliance with hospital follow-up visits. Secondly, some patients may have died while awaiting second next surgical procedure and thirdly, it is possible that patient symptoms improve after surgery and therefore do not return for follow up and do not go on to have further surgery [32].

STUDY LIMITATIONS

As this was a retrospective study, there were limitations. The record keeping, particularly during the initial 12 years of the study period was not optimal. Many of the records retrieved for the study had missing data especially shunt size, type of conduit used, age and weight of patients. It is also possible that there was under reporting of the number of BTS done, complications and deaths. Nevertheless, enough data was obtained to make comparisons with other institutions

5.0 RECOMMENDATIONS

This study has highlighted that although the aortopulmonary shunt is essential in the management of patients with cyanotic heart disease associated with reduced pulmonary blood flow, there should be a move to corrective surgery in neonates and small children, if possible. In addition, there is a need for improvement in postoperative care, especially regarding infection control practices. Lastly, record keeping needs to be improved particularly regarding shunt sizes and patients' follow-ups.

5.0 CONCLUSION

This was a descriptive study on the use, complications, and outcomes of aortopulmonary shunts for paediatric cardiac conditions in a resource constrained institution. The commonest indication for aortopulmonary shunts was tricuspid atresia, pulmonary atresia with VSD, tetralogy of Fallot and complex cardiac lesions. The frequency of aortopulmonary shunts compared to total surgeries has decreased over the study period, which may imply a trend towards corrective surgery for these cardiac lesions, although not statically significant. The modified BTS is the most frequently performed aortopulmonary shunt used for palliative surgery in our setting, similar to the trend in developed countries.

The morbidity and mortality in this study was higher compared to developed countries, with sepsis being the most common complication with an increased trend over the study period. Infection control practises will have to be carefully inspected and managed in our hospitals to prevent or decrease the prevalence of sepsis-related complications or other co-morbidities in the paediatric cardiac patients undergoing palliative surgical procedures such as BTS.

Conflict of interest: none declared

7.0 REFERENCES

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8.0 APPENDIX

Ethics Certificate

UNIVERSITY OF THE WITWATERSRAND, JOHANNESBURG

R14/49 Dr Nontobeko Charity Dladla-Mukansi

HUMAN RESEARCH ETHICS COMMITTEE (MEDICAL)

CLEARANCE CERTIFICATE NO. M170409

NAME: (Principal Investigator)	Dr Nontobeko Charity Dladla-Mukansi
DEPARTMENT:	Paediatrics Chris Hani Baragwanath Academic Hospital
PROJECT TITLE:	Surgical aortopulmonary shunt: A thirty-seven year experience in South African tertiary institution
DATE CONSIDERED:	05/05/2017
DECISION:	Approved unconditionally
CONDITIONS:	Title change (01/11/2019)
SUPERVISOR:	Prof A Cilliers, Dr K Vanderdonk and Dr V Mammen
APPROVED BY:	Dr C Penny, Chairperson, HREC (Medical)
DATE OF APPROVAL:	31/05/2017

This clearance certificate is valid for 5 years from date of approval. Extension may be applied for.

DECLARATION OF INVESTIGATORS

To be completed in duplicate and **ONE COPY** returned to the Research Office Secretary in Room 301, Third floor, Faculty of Health Sciences, Phillip Tobias Building, 29 Princess of Wales Terrace, Parktown, 2193, University of the Witwatersrand. I/we fully understand the conditions under which I am/we are authorized to carry out the above-mentioned research and I/we undertake to ensure compliance with these conditions. Should any departure be contemplated, from the research protocol as approved, I/we undertake to resubmit the application to the Committee. **I agree to submit a yearly progress report**. The date for annual re-certification will be one year after the date of convened meeting where the study was initially reviewed. In this case, the study was initially reviewed in July and will therefore be due in the month of July each year. Unreported changes to the application November invalidate the clearance given by the HREC (Medical).

Principal Investigator Signature _____ Date _____

PLEASE QUOTE THE PROTOCOL NUMBER IN ALL ENQUIRIES