Original Article

The Glenn procedure: Clinical outcomes in patients with congenital heart disease in pakistan

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ABSTRACT

Objectives: Congenital heart defects (CHDs) affect more than 40,000 children annually in Pakistan. Approximately 80% of patients require at least one surgical intervention to achieve a complete or palliative cardiac repair. The Glenn shunt, a palliative procedure is established between superior vena cava (SVC) and the right pulmonary artery to provide an anastomosis offering minimal risk to patients with univentricular heart disease. The aim of this study was to assess the clinical outcomes of the Glenn shunt procedure in patients with complex congenital heart diseases in a developing country like Pakistan.

Materials and Methods: A retrospective chart review was conducted on patients who underwent a bidirectional Glenn shunt procedure from July 2006 to June 2017. Data were collected on a structured questionnaire and analyses performed on SPSS version 22. Frequencies and percentages were computed for categorical variables while mean and standard deviation for continuous variables where appropriate.

Results: A total of 79 patients underwent the Glenn shunt procedures. The median age was 1.9 years and 54.5% were male. Tricuspid atresia was the primary diagnosis in 30.4% of the patients. Common morbidities included arrhythmias (6.3%), pleural effusion (8.9%), wound infection (3.8%), pneumonia (2.5%), and seizures (3.8%); reopening was required in 2.5% of the patients and 8.8% were readmitted within 30 days of index operation. There were three (3.8%) deaths in total.

Conclusions: Bidirectional Glenn shunt procedure can be performed safely in patients with ideal characteristics as the first stage palliation and has favorable results with acceptable rate of complications.

Keywords: Bidirectional Glenn shunt, cavopulmonary anastomosis, congenital heart disease, Fontan completion, Glenn shunt, single ventricle

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INTRODUCTION

Congenital heart defects (CHD) are one of the major causes of birth defects associated with significant morbidity and mortality.[1] The incidence of congenital heart defects ranges from 4 to 50 per 1000 live births[2] and nearly 1.5% of infants are born with single ventricle physiology. In general, CHD affects about 40,000 births per year in the United States.[3] The survival depends upon the severity, diagnosis, and treatment of the diseased condition.

The single ventricle is a nonspecific term covering the heart defects that have a similar problem of possessing only one functional ventricle; the affected chamber may be small or underdeveloped. These defects can be treated by a series of palliative operations; the different stages

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include Norwood procedure, modified Blalock-Taussig shunt (MBTS), BT shunt/PA banding, bidirectional Glenn shunt (BDGS), and then Fontan completion. BDGS remains the intermediate palliative procedure and is known to reduce mortality in comparison to those patients who were originally advanced directly to a Fontan procedure. Better outcomes have resulted in Glenn shunt being made as an essential component for the management of patients with single ventricle physiology.

In Pakistan, the actual incidence of CHD cannot be estimated due to the lack of a dedicated registry or database. About 40,000 children in Pakistan are born with a CHD annually and 11% of these children die during the first month of birth.[4] Nonetheless, the data available on the consequences of Glenn shunt procedure, performed in Pakistan, is scarce, as only a few centers dealing with CHD perform BDGN, that too without database. In the present study, we aimed to determine the clinical outcomes of patients with complex CHD (single ventricle physiology), who have undergone Glenn shunt procedure at our institution.

MATERIALS AND METHODS

A retrospective chart review was conducted for all the patients with complex CHD, who underwent Glenn shunt for single ventricle physiology from July 2006 to June 2017. The exemption was obtained from the institutional ethical review committee. The data was collected on a structured questionnaire including demographics, perioperative characteristics of patients, and clinical outcomes until the last follow-up available in files and/or database registry. Incomplete or missing records were excluded.

Surgical technique

Most of the surgeries were performed under normothermic conditions, however, in certain cases, the patients were cooled down to desirable temperature to perform additional procedures with subsequent rewarming. Cardiopulmonary bypass (CPB) was established in all the procedures except in selected cases of bilateral SVCs where bilateral Glenn shunts were done without CPB. Glenn's procedure [Figure 1] was performed on the side with a well-developed pulmonary artery. After completion of the anastomosis, adequate function of the shunt was ensured by directly measuring mean Glenn pressure in all the patients. Postoperatively, the patients were stabilized in the cardiac intensive care unit and then placed on a step-down therapy, as their condition improved.

Statistical analyses were performed on SPSS version 22. Frequencies and percentages were computed for categorical variables and mean and standard deviation or median and range for continuous variables where appropriate. A $P < 0.05$ was considered statistically significant.

RESULTS

A total of 79 cases were analyzed. The median age of the patients was 1.9 years, (range 0.1–29.5) and 43 (54.5%) were males [Table 1]. The median weight was 10.0 kg (range 4.9–73.0). The majority of the patients, $n = 62$ (78.4%) presented with cyanosis or shortness of breath, and 11.4% had shortness of breath only. As shown in Table 2, tricuspid atresia was the primary diagnosis in 24 (30.4%) followed by

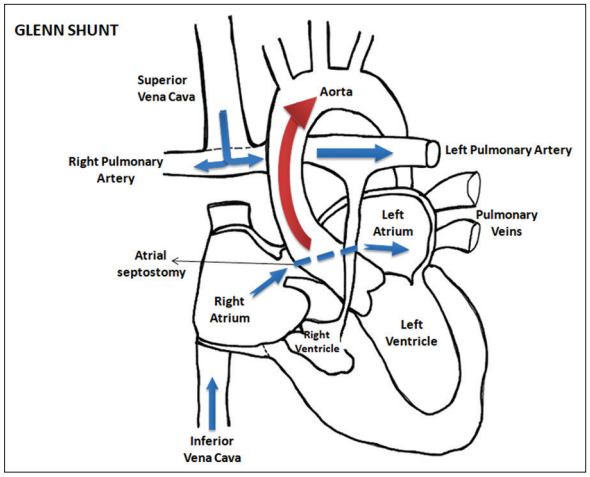

Figure 1: Diagrammatic representation of a Cavopulmonary Shunt

Table 1: Preoperative characteristics of patients, *n*=79

Demographics	*n* (%)
Gender	
Male	43 (54.4)
Female	36 (45.6)
Age (years)	
Mean±SD	4.5±6.1
Median (range)	1.9 (0.1-29.5)
Weight (kg)	
Mean±SD	14.4±12.1
Median (range)	10.0 (4.9-73.0)
Initial presenting symptoms	
Cyanosis	26 (32.9)
Cyanosis and shortness of breath	36 (45.6)
Shortness of breath	9 (11.4)
Preoperative oxygen saturation (%)	
Mean±SD	63.4±11.2

Table 2: Congenital anomalies

Anomaly	*n* (%)
Tricuspid atresia	24 (30.4)
Hypoplastic left heart/ventricle	19 (23.9)
d-Transposition of great vessels	16 (20.3)
Complete AVSD*	12 (15.2)
cc-TGA	7 (8.9)
Right atrial isomerism	1 (1.3)

*AVSD: Atrioventricular Septal Defect; cc-TGA: Congenitally Corrected Transposition of the Great Arteries

hypoplastic left heart syndrome or hypoplastic left ventricle (LV) in 19 (23.9%). Twenty-two (28%) patients had the prior palliative procedure and 14 (17.7%) underwent prior Blalock-Taussig procedure [Table 3].

All surgeries were performed using the CPB, with an average CPB time of 81.8 (±49.7) min. The mean preoperative Glenn shunt pressure was 15.3 ± 3.1 (mmHg) with subsequent Glenn pressures of 16.4 (±3.7) mmHg postoperatively in the CICU at 24 h. The mean preoperative peripheral oxygen saturation increased from 63.4 ± 11.2 (%) to 85.4 ± 7.0 (%) intraoperatively and 83.7 ± 5.4 (%) postoperatively. The prime concomitant procedures performed were atrial septectomy in 30 (37.9%) patients, PDA ligation in 19 (24.1%), and bilateral BDGS was performed in 15 (19%) patients having bilateral SVCs [Table 4]. The mean cross-clamp time with ventricular fibrillatory arrest (for atrial septectomy) was 3.9 ± 4.5 min and median inotropic score were 8.0 (2.0–87.0). Perioperative data is given in Table 5.

Table 6 summarizes the postoperative variables of the participants. The median time for mechanical ventilation, ICU stay, and length of hospital stay was 10.0 (1.0–470) h, 43.0 (5.0–474.0) h, and 8 (3–50) days, respectively. The reason for prolonged stay was found to be reintubation and prolonged ventilator support due to respiratory failure after the procedure. As seen in Table 7, overall 30 days morbidity was 38.3% which included readmission 07 (8.8%), reopening of sternum 02 (2.5%), pleural effusion 07 (8.9%), wound infection 03 (3.8%) including all those patients who had culture-proven wound infection, arrhythmias 05 (6.3%), seizures 03 (3.8%), pneumonia 02 (2.5%), and gastroenteritis 01 (1.3%). Out of the entire sample, 6 (7.6%) of the patients were referred for staged completion of the Fontan procedure while 16 (20.3%) waiting for scheduled Fontan in near future, the rest either were, lost to follow-up or deemed unsuitable for Fontan completion. We achieved an 80% follow-up rate with a median follow-up duration of 10 months (range 1–72 months). Most of the patients were doing well up to their respective follow-up time with no major sequela, showing a well-functioning Glenn shunt without any stenosis of superior vena cava-pulmonary artery anastomosis on echocardiography. Overall mortality was three (3.8%). Among them, one child died due to shunt failure, requiring redo sternotomy within 3 days of surgery while two died of sepsis.

DISCUSSION

CHDs are the most frequent lethal cardiac malformations, affecting about 1% of newborns and causing significant

Table 3: Prior palliation procedures, *n*=22

Procedure	*n*	%
Blalock-Taussig shunt	14	17.7
Blalock-Taussig shunt + PDA Ligation	3	3.8
Balloon atrial septoplasty + Blalock-Taussig shunt	1	1.3
Balloon atrial septoplasty + Pulmonary artery banding	1	1.3
Balloon atrial septostomy	1	1.3
PDA Ligation + Pulmonary artery banding	1	1.3
Pulmonary artery banding	1	1.3

PDA: Patent Ductus Arteriosus

Table 4: Concomitant procedures, *n*=55

Procedure	*n*	%
Total patients undergoing concomitant procedures	55	69.6
Atrial septectomy	15	18.9
Atrial septectomy + Bilateral bidirectional GS	4	5.1
Atrial septectomy + Bilateral bidirectional GS + PDA ligation	1	1.3
Atrial septectomy + PDA ligation	7	8.9
Atrial septectomy + PDA ligation + Pulmonary artery patching	2	2.5
Bilateral bidirectional GS	15	19
Bilateral bidirectional GS + Pulmonary artery patching	2	2.5
Pulmonary artery patching + banding + valvectomy	4	5.1
Other procedures	12	15.2

GS: Glenn Shunt; PDA: Patent Ductus Arteriosus

Table 5: Perioperative data

	Mean±SD
Perioperative oxygen saturation (%)	85.4±7.0
Perioperative Glenn pressures (mmHg)	15.3±3.1
Cross clamp time (min)	3.9±14.5
CPB time (min)	81.8±49.7
Circulatory arrest time (min)	2.6±8.8
Inotrope score Median (range)	8.0 (2.0-87.0)

SD: ± Standard Deviation

Table 6: Postoperative data

Outcomes	Mean±SD or Median (minimum-maximum)
Glenn shunt pressure (mmHg)	16.4±3.7
Postoperative oxygen saturation (%)	83.7±5.4
Duration of ventilator support (h)	10.0 (1.0-470.0)
Duration of chest tube drainage (h)	25.0 (8.0-240.0)
Duration of ICU stay (h)	43.0 (5.0-474.0)
Duration of hospitalization (days)	10.3±8.7
Median (range)	8 (3-50)
Follow up (months)	10.0 (1.0-72.0)
Patients with Fontan completion/*n*(%)	6 (7.6)
Patients waiting for Fontan procedure to be done/*n*(%)	16 (20.3)

Table 7: Complications (within 30 days)

	n	%
Overall Morbidity		38.3
Reopen	02	2.5
Wound infection	03	3.8
Readmission	07	8.8
Pneumonia	02	2.5
Pleural effusion	07	8.9
Arrhythmias	05	6.3
Seizures	03	3.8
Gastroenteritis	01	1.3
Mortality (within 30 days)	03	3.8

morbidity and mortality in children.[5] In some parts of the world, owing to lack of designated registry, the actual incidence of congenital cyanotic heart diseases is uncertain, owing to the ones who are asymptomatic or who were missed to diagnose and thus presented very late. CHD with single ventricular physiology poses a great challenge to the pediatric cardiology and congenital cardiac surgeons and these children are managed with intermediated palliation with BDG to prepare them towards Fontan circulation.

Therefore, Glenn shunt provides one such anastomosis offering minimal risk, excellent palliation, good pulmonary artery growth increasing pulmonary blood flow in the absence of increased heart workload, and marked improvements in survival.[6] Hence, in infants with the univentricular type of CHD, the staged procedures towards completion Fontan have largely been adapted with better outcomes, when considered in ideal/appropriate candidates. The criteria being; an age above 6 months, pulmonary artery pressure <18 mmHg, a mean ventricular end-diastolic pressure <12 mmHg, and not more than mild atrioventricular regurgitation.[7] Most of our patients fell under these selection criteria.

Around 15 (19%) patients were having bilateral SVCs requiring bilateral bidirectional shunt including two patients with azygos continuation of IVC as well, where Kawashima procedure was performed. Execution of both the stages (Glenn shunt followed by Fontan completion) separately offers reduced mortality as compared to the patients who are directly destined to the second stage.[8] Nonetheless, continuously elevated Fontan pathway pressures with consequently raised systemic pressures lower the longevity of this procedure, leading to increased morbidity with atrial arrhythmias,[9] protein-losing enteropathy,[10] and pleural effusions,[11] which worsens with age. In the nonexistence of any pulmonary arteriovenous malformations, improved cardiac output is provided by the Glenn shunt, with decreased morbidity and mortality than the Fontan approach.[12] A huge fraction will still have late complications of systemic venous hypertension, some of whom, known as the high-risk patients, might turn out to be the candidates for heart transplant eventually.

Previously, most of the Glenn shunt procedures have been performed on children between the age group of 6 to 12 months, suggesting that the optimal age is between 3–9 months,[7] followed by Fontan completion a year later. Moreover, studies have suggested that older patients with CHD are affected with significant morbidity from complications of their disease, palliations, or repairs.[13] Similarly, our study patients were also older than those in series from more developed countries.[10] The relatively older age of our patients at the time of presentation and subsequently late diagnosis results from lack of established primary healthcare facilities and expertise, as well as facilities for in timely diagnosis and referral; reduced awareness regarding available palliative options, and poor financial status also played a huge role. However, in our study, no association was found between patient age at the time of surgery and length of intensive care unit or hospital stay, surgical morbidity or mortality, when compared with different age groups, morbidity, and mortality. Furthermore, there was less distortion of pulmonary arteries, the development of pulmonary hypertension, pulmonary arteriovenous malformations, and most importantly, arterial oxygen saturation was corrected. Overall, the oxygen saturation was reported to be around 80% in infants following BDGS procedure[14] which was corroborated in our study wherein the mean oxygen saturation was 83.7 ± 5.4 (%) at discharge.

Management of hypoxia included an increase in the proportion of inspired oxygen thus enhancing inotropic support to improve the cardiac output thereby the flow to the lungs. Postsurgically, all the patients were heparinized for the first 24 h, followed by aspirin. Diuresis, chest tube drainage, and salt-restricted diet were included in our standard postoperative protocol. Patients who developed a neurological complication were investigated by a CT scan on the manifestation of the corresponding symptoms like seizures. At the time of discharge, the patients were assessed clinically for any neurologic dysfunction with all of them being discharged with no neurological abnormality.

Early studies have recorded significant morbidity and mortality in the early postoperative period among patients undergoing BDGS procedure in the form of pleural effusion, requiring intervention, or prolonged duration of hospital stay; this was the case in our study as well, where seven (8.9%) patients developed pleural effusion post-surgery which contributed to one of the major causes for prolonged hospital stay,[15] which was comparable to previous reports from successful Glenn surgeries.[16] This problem is likely to be associated with an increased left-to-right shunt. A comparable fraction of wound infection (4.8%) was reported by others on culture-proven reports.[16] Our study participants had comparable rather better rates of culture-proven wound infection of about 3.8%. In addition, only 1.2% of our sample developed pericardial effusion and similar results (0.7%) were observed by Kogon *et al.* following a BDGS procedure.[17] In the same study, the prevalence of pneumonia was 1.5%, comparable to our results of 2.5%. About 6% of the patients who suffered from arrhythmias were aggressively managed with

antiarrhythmic agents, and selective atrial cardioversion was required and pacing in case of nodal rhythm assisted in alleviating arrhythmias for maintaining cardiac output.

Only 7.6% of children were proceeded to Fontan completion compared to 69–76% of the cases reported previously.[18,19] Since only a few centers perform the procedure due to lack of resources and expertise, lack of awareness delayed presentation, shorter or lost to follow-up, as well financial constraints of the family, explains the decreased rates of patients proceeding to Fontan completion. This is further supported by the fact that the median follow-up period in our study was found to be only 10 months while both the above-mentioned studies had a mean follow-up period of 5.6[18] and 9.1[19] years, respectively. An efficient primary healthcare system with available financial assistance for the deprived may lessen the rates of delayed presentation with CCHD in the presence of a national registry and database with regular follow-up with improved documentation is recommended.

Previously, the overall mortality rate following a Glenn shunt procedure was reported to be 5–15%[20] with heterotaxy syndrome, systemic right ventricle, anomalous pulmonary venous drainage, abnormal pulmonary artery architecture and high pulmonary artery pressure being major factors influencing mortality. Our study had comparable results in the lower range (3.8%) of the previously reported mortality rates. One death resulted from early shunt failure, requiring redo sternotomy and revision of shunt but the patient died later due to coagulopathy, bleeding, and respiratory failure while the other two patients died of sepsis.

Limitations

The results of the study should be interpreted with some limitations: It is a single-center retrospective audit so the results should be generalized with caution. The deficient and/or missing data tend to affect the prevalence and related outcome estimates. The omission of the use of risk stratification for the classification of patient might have introduced a selection bias. In addition, psychological and developmental evaluation of the patients was not done, which might have indicated any influence on CNS and eventually on the outcomes. To overcome these drawbacks, detailed studies are needed to determine outcomes of the Glenn shunt procedure in future, on a larger sample, with a prospective study design involving multiple institutions.

CONCLUSIONS

BDGS procedure can be performed safely in patients with ideal characteristics, as the first or intermediate stage palliation and has favorable results with a low rate of complications in a developing country like Pakistan despite delayed presentation, conversion rate to Fontan completion can be better predicted. In the present era, a nationwide registry and database are the prime necessities for better outcomes.

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Conflicts of interest

There are no conflicts of interest.

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