

Utility of the Bidirectional Glenn Shunt

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Abstract

Background: Congenital heart disease (CHD) is a significant cause of childhood morbidity and mortality worldwide. Bi-directional Glenn Shunts (BDGS) form part of the surgical strategy used to treat congenital heart disease (CHD); no data exists on BDGS usage in the study locality. **Methods:** A 7-year retrospective, descriptive study was carried out at the Kenyatta National Hospital (KNH) in Nairobi, Kenya between January 1st 2006 and December the 31st 2012. **Results:** Eleven (11) BDGS's were performed in 11 patients; 63.6% had tricuspid atresia, 27.3% had double outlet right ventricle and 9.1% had pulmonary atresia with

intact ventricular septum. **Conclusion:** Further studies are warranted to identify factors contributing to the late performance of BDGS, poor post-operative follow-up and failure to perform FC's.

Key words: Cavopulmonary, Glenn, Shunt, Bidirectional
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Introduction

Congenital heart disease is the most common congenital abnormality; its prevalence is estimated to be 1:1000 live births (1). As a result of contemporary interventions, the majority of patients can survive into adulthood (2); the median age at death was 2 years in 1993 and 23 years in the current era (3). From a surgical perspective, CHD may be divided into 3 categories:

1. Patients in whom the lesion can be anatomically corrected surgically and who will not require further surgery
2. Patients in whom the lesion can be anatomically corrected surgically but will require further surgical intervention(s)
3. Patients in whom the lesion can only be palliated surgically.

The palliative surgical solutions offered in earlier era's were not as efficient and durable as the current solutions. Presently, to survive into adulthood, most of the patients in the 3rd category will require that a single ventricle do the work of two. The most significant surgical innovations in this regard were the introduction of the "Classical" Glenn Shunt in 1958 (4) and the "Classical" Fontan connection (CFC) in 1971(5). Prior to these two innovations, a systemic arterial-to-pulmonary artery shunt such as a Blalock-Taussig Shunt (6) was created. It was symptomatically effective but caused significant volume loading of the systemic ventricle with eventual heart failure. The "Classical" Glenn Shunt (CGS)

involves the connection of the superior vena cava (SVC) to the right pulmonary artery alone (4). A subsequent modification (the Bi-directional Glenn shunt or BDGS; figure 1) connects the SVC to both the right and left pulmonary arteries (7). Over time the quality of palliation provided by the CGS and BDGS deteriorates as the relative volume of venous return contributed by the SVC decreases with age. The CFC improves arterial oxygenation by re-routing the inferior vena cava (IVC) to the left pulmonary artery in a patient with a prior CGS.

In contemporary surgery, it is important to allow the child to grow sufficiently before performing the Fontan connection (FC) as this enables an optimal sized prosthetic conduit to be inserted (between the IVC and the right pulmonary artery) (Figure 2) for adequate blood flow as an adult(8). This staging involves performing a BDGS first and completing the FC several years later. Contemporary practice favours performing a BDGS at 3 to 4 months of age and the creation of the FC at 3 to 4 years of age. This study aimed to determine the indications for and surgical outcomes of BDGS in the study locality.

Methods

This seven year retrospective, descriptive study was carried out at the Kenyatta National Hospital (KNH) in Nairobi, Kenya between January 1st 2006 and December the 31st 2012. This was a file-based study; patient case notes were examined and the relevant data was entered into a data sheet.

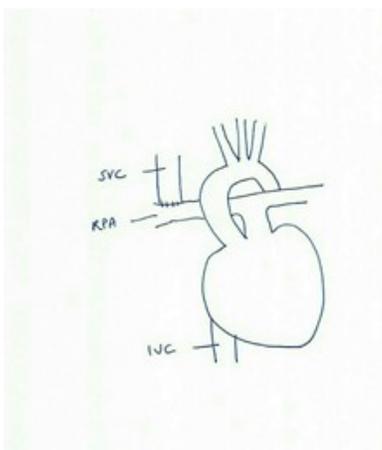


Figure 1: Bidirectional Glenn Shunt: SVC = superior vena cava; RPA = right pulmonary artery; IVC = inferior vena cava

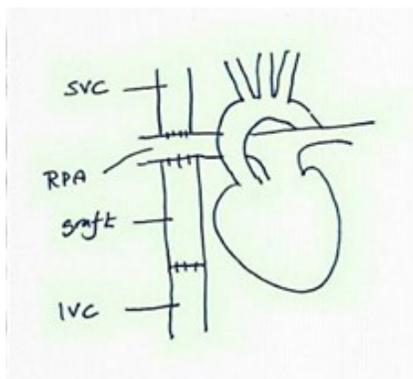


Figure 2: The Fontan Connection: SVC = superior vena cava; RPA = right pulmonary artery; IVC = inferior vena cava

All patients who underwent a BDGS procedure at KNH during the study period were included. Variables collected were: type of cardiac lesion, age at surgery, use of postoperative anticoagulation, length of intensive care unit stay (LOICUS), operative mortality, length of hospital stay (LOHS), duration of post-operative follow-up and pre- and post-operative arterial oxygen saturations. Microsoft Excel (2010) was used for data analysis. The study was carried out after approval by the Kenyatta National

Results

A total of 11 BDGS were performed on 11 patients. All surgeries were performed by the resident team. The operative mortality was 9.1% (1/11) and the post-operative follow-up rate was 30% (3/10). Ten out of 11 (90.9%) BDGS were performed under cardiopulmonary bypass. One patient presented with a patent ductus arteriosus (PDA) which was not ligated at surgery. Post-operative heparin was used in 1 patient (9.1%) and no patients went home on aspirin. The single post-operative mortality was in a 1.5 month old male with

tricuspid atresia. No FC procedure was performed. The patient demographics are summarised in table 1. There were three types of lesions: 63.6% were tricuspid atresia (TA), 27.3% were double outlet right ventricle (DORV) and 9.1% were pulmonary atresia with intact ventricular septum (PA-IVS).

Table 1: Demographic details of patients

Parameter	Median Value
Age	36 months
Weight	11.95 kg
Pre-Op Arterial Oxygen Saturation	80%
Post-Op Arterial Oxygen Saturation	91%
LOICUS	2 days
LOHS	7 days

LOICUS= intensive care length of stay;

LOHS= hospital length of stay.

Discussion

When CHD cannot be anatomically corrected; the majority of patients can be offered a robust palliative surgical alternative (one of several variants of the CFC). The final result of these variants is that the SVC and IVC venous flow is redirected to the lungs completely bypassing the right side of the heart. This is achievable because the pulmonary arterial system is a low-resistance and low-pressure system. Neonates, however, have a high resistance and a high-pressure pulmonary arterial system; this falls to adult levels by the 3rd month of life (9). For this reason a systemic arterial-to-pulmonary artery shunt is performed in patients younger than 3 months of age as the CGS, BSGS or FC will not be tolerated (10).

When the palliative pathway is chosen, the goal is to create a FC. Typically, the ideal FC is achieved by the age of 3 years through 2 or 3 operations. In our study, the median age at BDGS creation was 36 months. This is significant for 2 reasons: firstly, this is far older than the recommended 3 months of age; secondly, a BDGS was created when ideally a FC was indicated. Previous work done in the study locality (11) demonstrated that the mean age at referral to a paediatric cardiologist was 16.9 months and that the mean age at confirmation of CHD by echocardiography was 18.6 months. The study also showed that the mean duration from confirmatory echocardiogram to the 1st operation for CHD was 8 months. This work highlights significant delays in both diagnosis and surgery; this may have contributed to the late BDGS in our study. A BDGS was performed as opposed to a FC even when patients presented late as evidence suggests that the outcome for FC's is better when performed in a staged manner (12).

It is striking that no FC's were performed. There is long-term (20-30 years) outcome data on CGS (with or without

concomitant systemic arterial-to-pulmonary artery shunts) that show results comparable to some FC long-term outcome data in terms of survival (13). It is conceivable that these data may have influenced surgical decision making in our resource-constrained environment favouring BDGS's over FC's. Our 30% follow-up rate is very low. There is anecdotal evidence that patients will not seek medical attention in our region as long as they are feeling reasonably well. BDGS's can provide a reasonable quality of life for several years. This too may have contributed to failure to perform FC's. Recent long-term FC outcome data is available that shows that the contemporary FC offers the best long-term palliation and that the dreaded protein-losing enteropathy occurs in only about 9% of patients (14).

Our surgical results and the spectrum of lesions palliated are similar to what has been reported elsewhere (10). There is only one public hospital that regularly performs cardiac surgery in Kenya and it accounts for about half of all cardiac surgeries performed in the country. Six cardiac surgeons work at this institution. It is estimated that over 12000 children are born in Kenya each year that require cardiac surgery on cardiopulmonary bypass; less than 1% receive surgery(11). Anecdotal evidence suggest that this is primarily because of inadequate mobilisation of resources to deliver this service; the quality of our BDGS service may be a reflection this.

Conclusion

Current evidence suggests that the FC should be offered where indicated to our patients, however, established protocols for optimal palliative surgery for CHD have not been employed in our patients. Further studies are warranted to identify factors contributing to the late performance of the BDGS and failure to perform the FC in our patients.

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