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LONG TERM COMPARITIVE ANALYSIS OF SINGLE VENTRICLE PHYSIOLOGY PATIENTS FOLLOWING BIDIRECTIONAL GLENN SHUNT WITH OR WITHOUT ANTEGRADE PULMONARY BLOOD FLOW

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Abstract

The bidirectional Glenn procedure (BDG) is used in the staged surgical management of patients with a functional single ventricle. Controversy exists regarding whether accessory pulmonary blood flow (APBF) should be left at the time of BDG to augment systemic saturation or be eliminated to reduce volume load of the ventricle. The present study is a retrospective review of patients undergoing BDG that was conducted to assess the influence of APBF on survival rates.The data available on the use of Bidirectional Glenn with APBF is unclear , the advantages are higher oxygen saturation, lower mortality, favourable effects on cardiac functions, prevention of arteriovenous fistulas and a better growth of pulmonary arteries if compared with BDG with pulmonary artery interruption. patients who have undergone BDG with APBF for univentricular heart repair maintain circulation of hepatic factors into pulmonary arterial system to prevent pulmonary arteriovenous malformations and presence of APBF would result in raised pulmonary artery pressure volume load on the single ventricle

Keywords: Bidirectional, Shunt

INTRODUCTION

Bidirectional Glenn is a palliative procedure and a basic step towards total cavopulmonary connection. Preserving confluence of pulmonary arteries, obtaining bilateral pulmonary blood flow and leaving options open for any subsequent procedures. The bidirectional Glenn shunt is the second in a series of three staged surgeries to reconstruct a single ventricle heart, in situations where corrective bi-ventricular surgery or cardiac transplantation are not feasible. The bidirectional Glenn shunt procedure involves rerouting circulation such that the superior vena cava (SVC) drains into the right pulmonary artery. This results in deoxygenated blood returning from the head and upper body directly routed to the pulmonary arteries for oxygenation by the lungs, to some extent reducing the ventricular workload. Since the blood passing from the SVC into the pulmonary arterial system flows bidirectionally to both right and left lungs, it is called a bi-directional Glenn procedure.

The Glenn procedure was introduced in 1958 by William Glenn and modifications to the procedure were published by Dr. Azzolina in 1973. The original description by Dr. Glenn

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allowed communication only between the right pulmonary artery and the SVC, whereas the modified technique had the SVC connecting at or before the bifurcation between the right and left pulmonary arteries.

The bidirectional Glenn procedure (BDG) is used in the staged surgical management of patients with a functional single ventricle. Controversy exists regarding whether accessory pulmonary blood flow (APBF) should be left at the time of BDG to augment systemic saturation or be eliminated to reduce volume load of the ventricle. The present study is a retrospective review of patients undergoing BDG that was conducted to assess the influence of APBF on survival rates.

The data available on the use of Bidirectional Glenn with APBF is unclear , the advantages are higher oxygen saturation, lower mortality, favourable effects on cardiac functions, prevention of arteriovenous fistulas and a better growth of pulmonary arteries if compared with BDG with pulmonary artery interruption. patients who have undergone BDG with APBF for univentricular heart repair maintain circulation of hepatic factors into pulmonary arterial system to prevent pulmonary arteriovenous malformations and presence of APBF would result in raised pulmonary artery pressure volume load on the single ventricle . Hence weighing the advantages over disadvantages of both the procedures ie with antegrade flow and the other with pulmonary artery interruption in BDG single ventricle .we believe the need of review of literature with the above retrospective studies.

Methodology

Hypothesis:

We hypothesise that patients with antegrade flow have higher saturations ,comparatively bigger PA sizes ,assesement of CATH pressures pre fonatn may be same.

Objectives:

1) To Assess advantages and disadvantages of antegrade pulmonary blood flow in post op Bidirectional Glenn patient with single ventricle
2)To compare pulmonary artery size prior to fontan completion
3) To Assess the Survival of patients following Bidirectional Glenn shunt

Inclusion criteria:

All patients with Single ventricle physiology who underwent Bidirectional Glenn Shunt during the study period(2006-2019)

Exclusion Criteria:

1) All patients who underwent Bidirectional Glenn Shunt with biventricle physiology
2)Patient in the Study period without CATH data (Pulmonary artery pressures, prior to Fontan) and not have undergone CT imaging
3) Patient's who lost to follow up

Bidirectional Glenn is a palliative procedure and a basic step towards total cavopulmonary connection. Preserving confluence of pulmonary arteries , obtaining bilateral pulmonary blood flow and leaving options open for any subsequent procedures. The data available on the use of Bidirectional Glenn with APBF is unclear , the advantages are higher oxygen saturation, lower mortality, favourable effects on cardiac functions, prevention of arteriovenous fistulas and a

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better growth of pulmonary arteries if compared with BDG with pulmonary artery interruption. However patients undergoing BDG for univentricular heart repair should have an antegrade pulmonary flow to maintain pulsality and circulation of hepatic factors into pulmonary arterial system to prevent pulmonary arteriovenous malformations. however in patient with pulmonary with increased pulmonary blood flow and raised pulmonary artery pressure needed pulmonary artery interruption to prevent development of SVC syndrome also the pulmonary artery interruption prevents the load on the single ventricle . Hence weighing the advantages over disadvantages of both the procedures ie with antegrade flow and the other with pulmonary artery interruption in BDG with single ventricle .we believe the need of review of literature with the above retrospective studies.

Results

STATISTICAL METHODS:

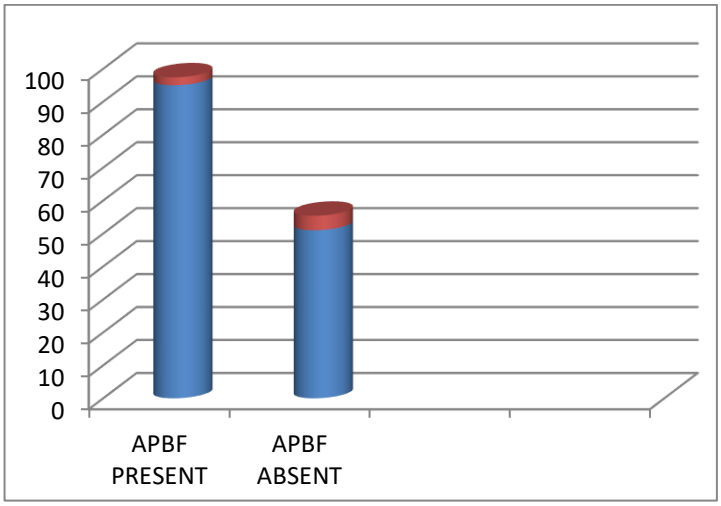
PA Pressures, Saturation, RPA and LPA Sizes were considered variables. Antegrade Pulmonary Blood Flow was considered as primary explanatory variable.

All Quantitative variables were checked for normal distribution within each category of explanatory variable by using visual inspection of histograms and normality Q-Q plots. Shapiro- wilk test was also conducted to assess normal distribution. Shapiro wilk test p value of >0.05 was considered as normal distribution. For normally distributed Quantitative parameters the mean values were compared between study groups using independent sample t-test (2 groups). The change in the quantitative parameters, before and after the intervention was assessed by paired t-test (In case of two time periods) P value < 0.05 was considered statistically significant. IBM SPSS version 22 was used for statistical analysis.(1)

1. IBM Corp. Released 2013. IBM SPSS Statistics for Windows, Version 22.0. Armonk, NY: IBM Corp.

Table 1: Descriptive analysis of Antegrade Pulmonary Blood Flow in the study population (N=144)

Antegrade Pulmonary Blood Flow	Frequency	Percentage
Present	93	64.58%
Absent	51	35.42%



There were a total of 263 patients who underwent BDG from 2006-2019 with univentricular physiology. Out of these 144 patients who underwent BDG underwent both CATH Study and

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CT imaging and met the requirement of the cohort and fulfilled the inclusion criteria(who were waiting for TCPC or already had undergone TCPC). Out of that 144 patients 93 patients were grouped into 1st subset ie they had antegrade pulmonary blood flow and 51 patients did not have antegrade pulmonary blood flow.

Table 2: Comparison of the age at which the patients underwent BDG in the cohort

Parameter	Antegrade Pulmonary Blood Flow (Mean± SD)		P value
	Patients with APBF (N=93)	Patients without APBF (N=51)	
Age at BDG	3.19 ± 3.55	2.01 ± 2.14	0.032

Age: The clinical features, timing and type of presentation of single ventricle, depends on the associated
lesions and degree of outflow obstruction. The most frequent presenting symptom is
cyanosis since birth. Thus the Age at which the patients underwent BDG in Subsets of Patients without
antegrade flow as expected was earlier in comparison with the patients with Antegrade blood flow ,
with mean age at 2 years for the 2nd group and 3 years and two months for the 1st group. A patient in the
subset with no Antegrade pulmonary blood flow underwent BDG as early as 2nd months of life in contrary to
patients with antegrade blood flow the maximum age at BDG was 18 years.

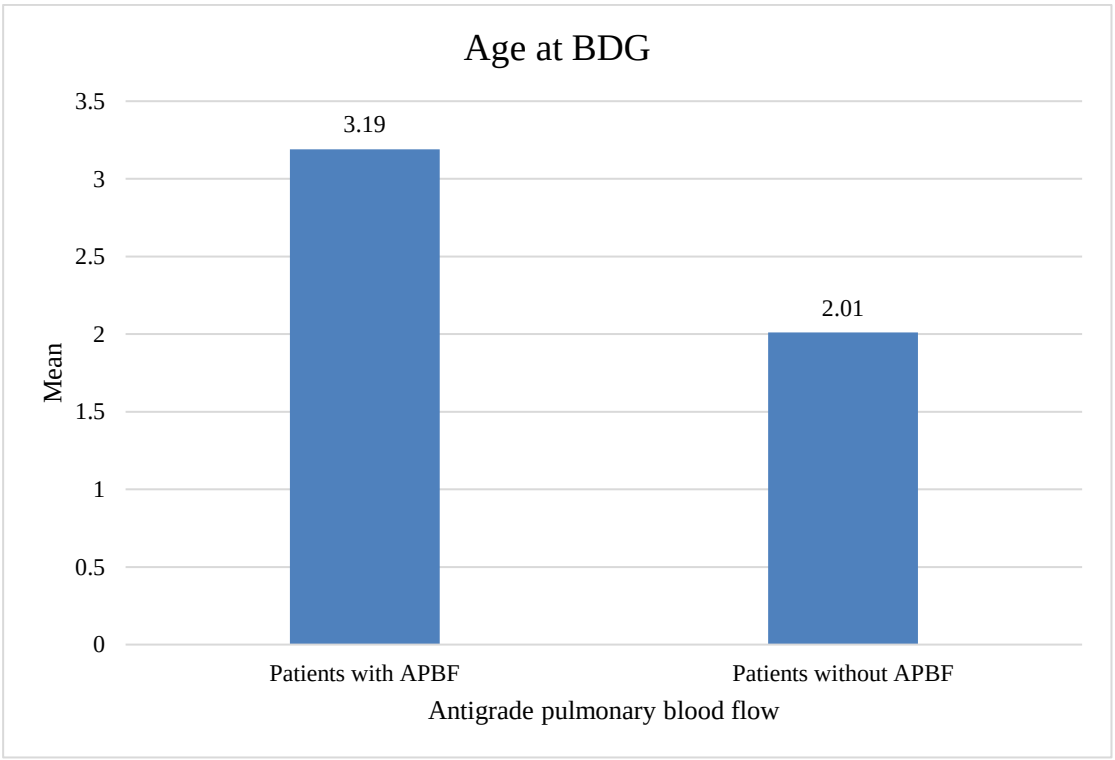

Figure 1: Bar chart of comparison of Age at BDG between antegrade pulmonary blood flow (N=144)

DIAGNOSIS ACROSS THE COHORT

1. Tricuspid atresia with pulmonary stenosis/PAH-66

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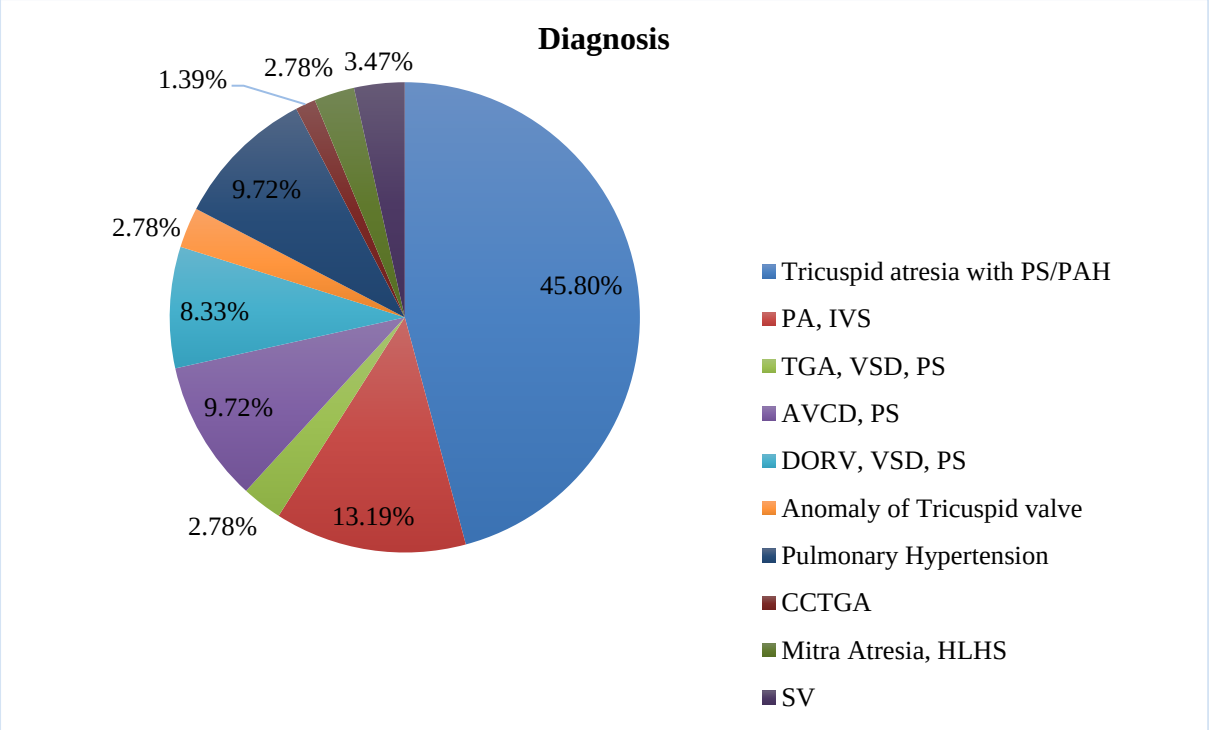
2. Pulmonary atresia with Intact Ventricular septum and Hypoplastic right heart variants PA, IVS Transposition of Great Arteries, with Ventricular septal defect, Pulmonary stenosisTGA, VSD, PS-19
3. Common Atrioventricular canal defects, with pulmonary stenosis/pulmonary hypertension AVCD, PS-14
4. 4)Double Outlet Right Ventricle with pulmonary stenosis/pulmonary hypertension DORV, VSD, PS-12
5. Ebsteins anomaly of Tricuspid Valve, with poor functional right ventricle-4
6. Double Inlet left ventricle with pulmonary stenosis/pulmonary hypertension-14
7. Congenitally Corrected Transposition of Great Arteries with Ventricular septal defect, Pulmonary stenosis CCTGA-2
8. Mitral Atresia and hypoplastic left heart syndrome -4
9. Large V.S.D Amounting to single ventricle-5

Descriptive analysis of Diagnosis in the study population (N=144)

Tricuspid atresia with PS/PAH	66	45.8%
PA, IVS	19	13.19%
TGA, VSD, PS	4	2.78%
AVCD, PS	14	9.72%
DORV, VSD, PS	12	8.33%
Anomaly of Tricuspid valve	4	2.78%
DILV	14	9.72%
CCTGA	2	1.39%
Mitra Atresia, HLHS	4	2.78%
Large V.S.D mounting toSV	5	3.47%



Single ventricle as quoted is wide Spectrum of anatomic variations from Tricuspid atresia to HLHS, The single entity in an Indian setup is Tricuspid atresia, which is in contrast to the

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western world (most of their cases are HLHS) Triuspid atresia is the commonest anomaly palliated with BDG. Patients with a primary diagnosis of Tricuspid atresia constituted 45.8% in the Cohort . Other major lesions palliated with BDG included AVCD, DORV, SV and PA with IVS which constituted 41% of the cohort. Single ventricle including DILV was present in 10% of patients. A diagnosis of TGA, PA IVS and CCTGA were present in less than 10% of case.

Discussion

It's a larger Cohort study where patients underwent BDG having different spectra of diseases but physiologically was a single ventricle. The Glenn shunt is usually done after 3 months. Patients who have Single ventricle physiology has adequate source of pulmonary flow in patients with APBF. This Patients are usually protected with Higher PA pressure due to the Avascular bed but at the cost of saturation ,but usually the maintain adequate saturations . According to the Cohort we could state that ,patients with APBF usually did not require early palliation and BDG could be done on elective basis and the other subset without antegrade flow required early palliation before 6 months of age. In patients where pulmonary flow was unrestricted ,PA banding was done with BDG . Early BDG in the cohort were performed for patients having cyanotic spells. There were 19 patients (5%) who underwent BDG before 6 months of age, out of which 4(2.6% of total cohort) had the surgery within 4 months of age. According to the Cohort the majority diagnostic criteria belongs to Tricuspid atresia which eventually ended up into TCPC and the mortality rate is much lower may be attributed to the TA group . This data is quite contrasting to data from the western world, where the SV palliation is done for HLHS.The data available on the use of Bidirectional Glenn with APBF is unclear , the advantages are higher oxygen saturation, lower mortality, favourable effects on cardiac functions, prevention of arteriovenous fistulas and a better growth of pulmonary arteries if compared with BDG with pulmonary artery interruption. However patients undergoing BDG for univentricular heart repair should have an antegrade pulmonary flow to maintain pulsality and circulation of hepatic factors into pulmonary arterial system to prevent pulmonary arteriovenous malformations.however in patient with increased pulmonary blood flow and raised pulmonary artery pressure needed pulmonary artery interruption to prevent development of SVC syndrome. The pulmonary artery interruption also prevents the load on the single ventricle. BDG with APBF could be an destination therapy where the patients are not fit to undergo TCPC.

1)Age; The term Single ventricle is a spectrum of disease with wide anatomic variations from tricuspid

Atresia to HLHS syndrome .In our Cohort the age at which patients underwent BDG varied from as Early as 2 months to as late as 18 years of life. The extreme variation in the age can be attributed to Antegrade blood flow ,patients with absent APBF had severe cyanosis and presented earlier and also the Patient with unrestricted pulmonary blood flow presented with failure before 1 year of life and also the Advances in imaging with early antenatal diagnosis has been the major contributing factor in patients without symptoms. Our cohort study the median age group where the patients underwent BDG is around 3.2 years in patients with APBF in contrast to patients without APBF who needed early surgery with Median age of 2 years which can be attributed to presenting symptoms like cyanosis or cyanotic spells.

A Retrospective study by Jae Suk Baek Et al in 2021 for the patients who underwent BDG from 2000-2015 as a interim palliation for single ventricle physiology with Exclusion of patients who underwent Kawashima or Norwood operation , with total of 279 patients

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underwent BDG and out of which 202 patients with pre existing antegrade blood flow were included ,out of which Antegrade flow was terminated in 110 patients (no antegrade flow) and maintained in 92 patients The impact of antegrade pulmonary blood flow at bidirectional Glenn shunt on long-term outcome was analyzed using inverse probability of treatment weighting.
The Conclusion was the median age group was 8 months and weight was 7.8 kgs. The prologation in the pleural drain or readmission for the same was higher in patients with APBF In the no antegrade pulmonary blood flow group, B-type natriuretic peptide level was decreased further until the Fontan operation . In Patients with no antegrade flow ,saturation of oxygen was usually low after bidirectional Glenn shunt, although it was increased further until Fontan operation ($P < .001$), despite still lower oxygen saturation before Fontan operation compared with antegrade pulmonary blood flow group ($P < .001$). The McGoon ratio was decreased in both groups without intergroup difference, although the McGoon ratio before Fontan operation was higher in the antegrade pulmonary blood flow group (2.3 ± 0.4 vs 2.1 ± 0.4, $P = .008$). Overall transplant-free survival was worse in the antegrade pulmonary blood flow group (hazard ratio, 2.37; confidence interval, 1.089-5.152; $P = .030$).
Hence concluded Maintaining antegrade pulmonary blood flow at bidirectional Glenn shunt was beneficial for higher oxygen saturation and increase in the pulmonary artery size before Fontan operation.
However, it did not favour overall transplant-free survival with a sustained higher risk of death or transplant until the elimination of antegrade pulmonary blood flow.
2)Saturations: The varied anatomical spectrum from a right sided lesion like the commonest tricuspid to the rarely presenting HLHS in our cohort study (i.e Tricuspid atresia patients occupied 45% of the cohort and only two patients included HLHS of the total 144),the Saturations were better in patient with antegrade flow(both pre op and post op) and they presenting to the hospital at later stages in life (few were diagnosed accidentally ,ie during evaluation for a murmur),the rational being patients with antegrade flow had saturations which were just adequate to mask the effect of cyanosis and patients without antegrade flow often presented earlier with appreciable cyanosis or history of cyanotic spells at earlier part of life .our cohort is in accordance with other studies like Michele A. Frommelt and others in 1995,where they retrospectively studied 43 patients who underwent BDG in Children's Hospital of Wisconsin with Anatomic diagnoses included double inlet left ventricle (14 patients), tricuspid atresia (8 patients), pulmonary atresia with intact septum (6 patients), single right ventricle (5 patients), hypoplastic left heart (3 patients), unbalanced atrioventricular septal defect (3 patients), and other complex lesions (4 patients). they then divided the patients into two groups for analysis. Group 1 had only the cavopulmonary shunt as a source of pulmonary flow (22 patients) and the other group had an APBF as the additional source (21 patients). Patient age at the time of BDGranged from 6 months to 12 years, with group 1 patients being younger (31 versus 45 months, P=.05). Group 2 patients had higher postoperative central venous pressures (17.8 versus 14.1 mm Hg, P<.001) and oxygen saturations (86% versus 81%, P<.001) than did group 1 patients. There was no statistical difference between groups in the number of chest tube days or hospital days. There was 1 early death in group 1 related to severe ventricular dysfunction and 1 late death in group 2 related to sepsis. Five patients in group 2 were readmitted to the hospital for drainage of a large chylothorax compared with
none in group 1.Hence the study concluded that with an additional source of pulmonary blood flow after bidirectional glenn shunt have higher postoperative central venous pressures, have higher oxygen saturations, and are at risk for the

late development of a chylothorax

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Conclusions

The Average age where patients underwent BDG were late which can be attributed to late diagnosis. Tricuspid atresia is the commonest anomaly palliated with BDG and a higher proportion of patients with TA complete Fontan procedure. Tricuspid atresia has the lowest mortality among all patient groups. Bidirectional Glenn surgery has overall low mortality rate and good long term survival. It can provide effective long term palliation prior to Fontan completion. Palliation with SV of LV morphology and preserved antegrade pulmonary blood flow are significant predictors of survival.Bidirectional Glenn surgery with APBF had better Saturations ,PA Sizes and the need for surgery in Later stages of life but with same hospital stay and cath pressures .

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