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Bilateral Versus Unilateral Bidirectional Glenn Shunts: Saudi Experience

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Key Words: Glenn Cavopulmonary anastomosis.

Abstract

Introduction: Bidirectional Cavopulmonary Anastomoses (BDG a.k.a. Glenn shunts) as a first or second palliative stage procedure before embarking on a total cavopulmonary connection ("Fontan type" procedure) has been used to normalise volume loading of the single ventricle at an early age. The presence of bilateral Superior Vena Cava (SVC) may represent a technical challenge in the performance of the bilateral Cavopulmonary Anastomoses (b-CPA) connections.¹⁻²

Methods: The available chart and files data were reviewed and analyzed in 160 cases of unilateral bidirectional glenn (u-CPA) and 44 children undergoing bilateral cavopulmonary anastomoses (b-CPA). Uivariate analysis of the risk factors of mortality, morbidity and ICU and hospital stay was done.

Results: One hundred sixty patients had U-CPA (Group 1) and 44 Patients had b-CPA (Group 2) in King Faisal Specialist Hospital and Research Centre (KFSH&RC), Jeddah, Kingdom of Saudi Arabia, in more than 8 years. They were 127 males and 77 females. Interrupted IVC was present in 14 patients. Hypo RV was evident in 128 cases. The oxygen saturation increased from 77.4% to 82.9% and from 73.8% to 84.7% in Groups 1 and 2 and the pulmonary artery pressure dropped from 24.6 mmHg to 15.8 mmHg and from 27 mmHg to 16.6 mmHg in both groups. Mechanical ventilation, inotropes, nitric oxide and oscillator high frequency jet ventilation were needed more in Group 2. Hospital stay was longer in Group 2. Six cases died in Group 1, and three died in Group 2.

Conclusions: Bidirectional Glenn shunts can be done with acceptable morbidity and mortality in some cases of pulmonary hypertension. Bilateral CPA had a more tough postoperative course than unilateral CPA. The mortality risk factors in bidirectional Glenn shunts are bilateral SVCs, Status post

Damus Kaye Stansel (S/P DKS), S/P Norwood and high Pulmonary Artery Pressure (PAP).

Introduction

The bidirectional cavopulmonary anastomoses (CPAs) as a first or second stage procedure before embarking on a total cavopulmonary connection ("Fontan-type" procedure) has been used to normalise volume loading of the single ventricle at an early age. The presence of bilateral SVCs may represent a technical challenge in the performance of the BCPAs connections. In addition, bilateral BCPS have been also found associated with a higher operative mortality, an increased risk of thrombus formation, and a lower conversion rate to the "Fontan-type" circulation, if compared to a standard unilateral CPAs.¹⁻²

Application of the concepts for the Fontan circulation have evolved whereby nearly all patients with single ventricle physiology, including those with abnormalities of the systemic venous return are palliated on what has become known as the "Fontan tract." Although the initial results for the extension of the indications for the Fontan procedure were not always favorable, more recent studies have shown improvement.^{1,3}

Patients and Methods

This is a retrospective study including 204 cases of Glenn shunts operated upon in KFSH&RC; Jeddah, KSA, between 2000 and 2009. Preoperative evaluation included history taking, patient examination, routine laboratory and chest x-ray, ECHO diagnosis for all cases and cardiac catheterization for catheterization for higher risk cases, especially those with suspected or proved pulmonary hypertension. Detailed Consent from the family is taken. All our BDG were done through median sternotomy and with standard cardiopulmonary Bypass (CPB). No cardioplegia was given. Previous systemic to pulmonary artery shunts were disrupted by division, ligation or clipping. The azygos vein was clipped or ligated unless interrupted IVC or Glenn pressure markedly increased. The previous pulmonary artery band was tightened

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or main pulmonary artery was amputated from the confluence. Pulmonary Artery Banding was done as an associated procedure in forty cases. Pulmonary artery augmentation was done in twenty-four cases. The left SVC was occluded while the pressure in the cephalad side was monitored. If the pressure is more than 20 mmHg, this SVC was cannulated for CPB and a BBDG shunt was performed. Pressure monitoring was needed in the first few cases of our practice; however, we currently perform routinely BBDG in single ventricle with bilateral SVCs. ICU care included: monitoring, inotropes, mechanical ventilation, Nitric oxide and oscillator when needed, Head of the bed up to 45 degrees with furosemide, catopril, aspirin, dipyridamol, sildenafil with or without anticoagulation. Follow-up in the pediatric cardiology clinic was done. The median follow-up was 24 months with a range between 2-96 months.

Results

One hundred sixty patients had U-CPA (Group 1) and forty-four patients had b-CPA (Group 2) in King Faisal Specialist Hospital and Research Centre (KFSH&RC), Jeddah, in more than 8 years. They were 127 (94 in Group 1 plus 33 in Group 2) males and 77 (66 in Group 1 plus 11 in Group 2) females. BTS was previously done for 104 cases (80+24) and Pulmonary Artery Banding was done for 36 (29 and 7). Interrupted IVC was present in 14 (4 in Group 1 plus 10 in Group 2) patients. Hypo RV was evident in 128 (102+26) cases. The preoperative oxygen saturation increased from 77.4% to 82.9% postoperatively, and from 73.8% to 84.7% in Groups 1 and 2 postoperatively. Indexed pulmonary vascular resistance was <3.5 Units in 125 (95+30), > 3.5 units in 21 cases (15+6), and not assessed in 58 cases (50+8). The pulmonary artery pressure dropped from 24.6 mmHg to 15.8mmHg, and from 27 to 16.6 mmHg in both groups. CPB time was longer in Group 2 (69 min vs. 48min). Pulmonary artery banding was done concomitantly in 31 cases (25+6) and pulmonary artery augmentation in 22 cases (15+7). Mechanical ventilation, inotropes, nitric oxide and oscillator were needed more in Group 2 (0.8 days -5 hours -3 h.-1h. vs. 1 day -6h -5h -3h -1h) respectively in (Group 1 and 2). The hospital stay was longer in Group 2. Six cases died in Group 1, and three died in Group 2. ICU and hospital stays were longer in Group 2 (2.1 and 7 days vs. 2.6 and 10 days). Chylothorax was detected in 21 and 8 cases in both groups. Significant arrhythmias were detected and investigated in 30 and 6 cases. Re-intervention was needed in 13 cases (10+3) in the form of thrombosis of the Glenn shunt in 4 cases (3+1), bleeding in 6 cases (4+2) and thoracic duct ligation in three cases all in Group 1. Incorporation of the hepatic veins was needed in three cases (one in Group 1, and two in Group 2), and Fontan was performed in 42 cases (38+4). The data was statistically analysed using the Mann-Whitney test for continuous variables and chi-square test for categorical variables. Regression model was used for the potential risk factors. The continuous variables were: Age, Weight and CPB time, while the non-continuous variables included: Ventricular morphology dominance, bilaterality of SVCs with or without interrupted IVC, previous surgeries, use of CPB or not, and associated procedures (Table 1).

Discussion

We are presenting a reasonably large series in a tertiary care centre in Jeddah over a span of more than 8 years. It includes 204 cases of Glenn anastomoses. More than 25% of them are b-CPAs. This ratio is higher than those of Iyer et. al., 2000, Chowdhury et. al., 2001 and Kim et. et. al., 2006, but less than those of Tanoue Y. et. al., 2007, who reported a ratio of 14.2%, 16%, 3.2% and 27.9%. This may be due to the random nature of the study with different socioeconomic and ethnic status.^{1, 4, 5, 6} Male/Female was 1.64/1. The mean age was 17.9 months and 19.3 months, which is relatively old, as we are a tertiary centre managing cases from all

over our region with some delay and sub-optimal general and cardiac conditions. As per Mitchell ME et. al., 2006, and Tanoue Y. et. al., 2007, BDG should be performed between three to six months to achieve maximum benefits.⁷

However, BDG can be done up to two years old.⁶ Our youngest case was 1 month, and the oldest was 13 years. BTS was previously done in 104 (51%), and previous PAB was done in 17.6% of cases, so about 68.6% received a first step palliation in the form of BTS and/or PAB. We followed a staged strategy in most

Table 1. Showing the Demographic, Radiological, Operative, Clinical, Therapeutic and Prognostic Data of Both Groups.

	Unilateral Glenn	Bilateral Glenn	
Morphology No. Hypo RV Hypo LV	160 102 58	44 26 18	
Sex Male Female	94 66	33 11	
Age (months) Weight (Kgm)	17.9 10.5	19.3 10.9	
S/P PAB BTS DKS Norwood LV diverticulum excision	29 80 2 1 1	7 24 2 1 0	
IIVC	4	10	P<0.05
Preop. Saturation Postop. Saturation Preop. PAP Postop. PAP PVRI < 3.5 >3.5 NA	77.4 82.9 24.6 15.8 95 (53.1%) 15 (9.3%) 50 (37.6%)	73.8 84.7 27 16.6 30 (68%) 6 (13.6%) 8 (18.4%) P<0.05 P<0.05 P<0.05	
Bypass Time Associated PAB Pulm. A. Augment. ICU Stay Mechanical ventilation Inotropes Nitric Oxide Oscillator	48 25 15 2.1 d 0.8 d 5 h. 3 h 1 h	69 6 7 2.6 d 1 d 6 h. 5 h 3 h	P<0.05 P<0.05
Hospital Stay	7 d	10 d	P<0.05
Chylothorax	21 (13.1%)	8 (18.1%)	P<0.05
Arrhythmias	30	6	
Reoperation Thrombosis Bleeding Thoracic duct ligation	3 4 3	1 2 0	P<0.05 P<0.05
Mortality	6	3	P<0.05
Next Surgery Fontan Incorporation	39 38 1	6 4 2	

cases, as the quality of life in patients after Blalock-Taussig shunt operation may be better than that in patients after BDG, as BDG is not used as the final operation for patients who cannot be completed by the Fontan circulation.⁶ The median follow-up in our series was 24 months with a range between 2-96 months. Fontan was done for 38 and 4 cases in both groups (23.7% and 9%) respectively. Incorporation of the hepatic veins was done in three Kawashima cases. The aetiology of desaturation after Glenn is usually due to development of venous collaterals from the SVC to the IVC and intra-pulmonary arteriovenous shunting and reduction of the SVC/IVC flow ratio with age.⁸

We did believe in the staged palliation of single ventricle, especially early in our centre experience. However, we started to be a little more aggressive towards primary Glenn, so 31.4% received primary Glenn without previous palliation. The indication of BDG should be decided according to the surgical strategy aiming at the Fontan operation.⁶ Concerns about the performance of BCPS in young infants may have to do with the history of unfavorable results after performing a classic Glenn shunt in infants along with uncertainty regarding the reactivity of the pulmonary vasculature in this group of patients. Between 1990 and 1995, over one-third (36%) of all BCPS procedures performed at the University of California at San Francisco were in infants <6 months of age. Early results in this cohort of young infants have been similar to those in all patients undergoing BCPS during the same period, with mortality rates of 4.8% and 5.1% and BCPS failure rates of 11.9% and 8.5%, respectively. The only independent risk factor for early death and BCPS failure was age, <1 month; and the strongest risk factor for early or late BCPS failure (death or take-down) was age <2 months.⁹

BDG procedure offers several potential advantages over the use of systemic to pulmonary shunts. In particular, it avoids pulmonary arterial distortion and also the additional volume load on the systemic ventricle created by the use of systemic to pulmonary shunts. Both these complications are important risk factors for subsequent Fontan repair. In addition, bidirectional cavopulmonary anastomosis is a more logical strategy as it represents an evolution of the patients' anatomy towards the final Fontan circulation.¹⁰

In all cases, we abolished the retrograde accessory pulmonary flow coming through the BT shunt as the goal is to try to achieve an effective/total pulmonary blood flow ratio of about 1. The more the effective and the total pulmonary blood flows coincide, the better the palliation. All efforts were made to obtain the maximum oxygen saturation with the minimum ventricular overload. The advantage of the BCPA over the Blalock-Taussig shunt is to allow an increase of

effective pulmonary blood flow without an increase in total pulmonary blood flow and cardiac work. BCPA with Antegrade Pulmonary Blood Flow is a better step towards Fontan than BCPA with an additional pulmonary blood flow through a Blalock-Taussig shunt.¹¹

There was a significantly higher percentage of patients with Pulmonary Vascular Resistance Index (PVRI) >3.5 in Group 2 (13.6% vs. 9.3%), which may be a well-known high risk factor although there was no significant difference in the preoperative or postoperative PAP. This, in addition to longer CPB and heterogenous anatomical diagnosis, may explain the finding of longer Mechanical ventilation, inotropes, Nitric Oxide and Oscillator in Group 2 (0.8 days -5 hours -3 h. -1h. vs. 1 day -6h -5h -3h.) respectively in (Group 1 and 2), and the hospital stay was longer in Group 2, and that six cases died in Group 1, and three died in Group 2.

ICU and hospital stays were longer in Group 2 (2.1 and 7 days vs. 2.6- and 10 days). Using the univariate risk analysis, only the bilaterality of SVCS, interruption of IVC and PVRI >3.5 were significant risk factors for the longer ICU and hospital stay and chylothorax. As per Reddy VM et. al., 1997, elevated pulmonary vascular resistance did not appear to be a problem among any of the neonates and very young infants who fared badly after the operation, because they had normal pulmonary artery pressures and transpulmonary gradients throughout their postoperative courses. However, it has been pointed out that pulmonary artery pressures and gradients are not necessarily reliable indicators of resistance when pulmonary blood flow is low or when no good estimate of flow is available. Therefore, BCPS does not appear to be a viable option in neonates and should preferably be postponed beyond 2 months of age.⁹

In 2000, KFSH&RC; Jeddah, instituted a policy of offering BDG shunts to patients with pulmonary hypertension if their PVRI decreased to 3.5 WU/m² on 100% oxygen irrespective of mean PAP and PVR in room air and mean PAP on 100% oxygen. Current suggested criteria to not offer BDG shunting to those with PVRI >3 WU and/or mean PAP >18 to 20 mm Hg by some and >25 to 30 mm Hg by others work well with respect to good outcomes and low mortality. However, there are three reservations to these guidelines. First, in the absence of pulmonary stenosis, elevated mean PAP does not necessarily indicate the presence of obstructive pulmonary vascular disease and may reflect only the transmission of systemic pressure to pulmonary circulation and high pulmonary flow. Second, because PAP and PVR are flow dependent, it seems inappropriate to use a uniform level of PAP or PVR as a criterion for patients with "under circulated" or "over circulated" pulmonary circulation. The former reduces and the latter increases pulmonary flow, which affects PAP and PVR, and the current approach ignores this important variable. Third, it has not been well-studied, in the presence of unrestricted and increased pulmonary flow and elevated PAP, what degree of elevated PVR would lead to the failure of a BDG shunt.¹²

Chylothorax was detected in 29 cases (21+8). It was suspected when the amount of drainage has a milky nature and diagnosis is confirmed by the presence of chylomicrons in the drained fluid with lymphocytes more than 80% with fluid triglycerides/cholesterol ratio more than 1 or pleural fluid triglyceride level is >1.1 mmol/L. We think that it is due to high venous pressure (systemic and/or pulmonary), disruption of the minor lymphatic channels rather than thoracic duct injury. Our first line of treatment is a fat-free diet; monogen for at least 3-6 weeks with or without NPO, diuretics, captopril. For resistant cases, we may use a week course of steroids and or octreotides. We needed thoracic duct ligation in three cases; all were of the u CPAs group. So, although chylothorax incidence was significantly higher in Group 2 (18.1% vs. 13.1%), refractory chylothorax requiring thoracic duct ligation was only in Group 1. The higher incidence in Group 2 may be related to higher PAP and/or PVRI. However, we could not explain the refractory nature of three cases in Group 1. Chylothorax may be an indicator of additional pulmonary blood flow. Monogen is designed for infants and

“Bidirectional Glenn, whether unilateral or bilateral, can be done with acceptable morbidity and mortality. The mortality risk factors in bidirectional Glenn Shunts are bilateral SVCs, S/P DKS, S/P Norwood and high PAP. Using the univariate risk analysis, only the bilaterality of SVCS, interruption of IVC and PVRI >3.5 are the significant risk factors for the longer ICU and hospital stay and chylothorax.”

“Bidirectional Cavopulmonary Anastomoses (BDG a.k.a. Glenn shunts) as a first or second palliative stage procedure before embarking on a total cavopulmonary connection (“Fontan type” procedure) has been used to normalise volume loading of the single ventricle at an early age.^{1,2”}

children with lipid and lymphatic disorders. The osmolality of Monogen is substantially lower than Portagen and most other elemental or fat-free formulas, a difference that improves gastrointestinal tolerance of the feed. Monogen also has a substantially higher energy level than alternative feeds. This point is important because patients with chylothorax have high energy requirements due to increased metabolic demand from the combination of chyle loss and hypermetabolism associated with surgery.^{13,14}

Nine cases (4.4%) died during the follow-up - six cases from the Group 1 (3.75%) and three cases (6.8%) from Group 2. So, we can say that bidirectional glenn, whether unilateral or bilateral, can be done with acceptable morbidity and mortality. The median survival of those nine mortalities after the Glenn was 6 months. The accused causes of death were myocardial dysfunction, arrhythmias, tension pneumothorax and cerebrovascular strokes. The mortality risk factors in our study were bilateral SVCs, S/P DKS, S/P Norwood and high PAP. Kogon BE et. al., 2007 found longer CPB time and High CVP as the only significant mortality risk factors. They also found those two factors plus high transpulmonary gradient and right ventricular morphology and low body weight at the surgery time as the significant risk factors for the longer ICU and hospital stay.¹⁵

Conclusion

Bidirectional glenn, whether unilateral or bilateral, can be done with acceptable morbidity and mortality. The mortality risk factors in bidirectional Glenn Shunts are

bilateral SVCs, S/P DKS, S/P Norwood and high PAP. Using the univariate risk analysis, only the bilaterality of SVCS, interruption of IVC and PVRI >3.5 are the significant risk factors for the longer ICU and hospital stay and chylothorax.

Limitations of the Study

This is a retrospective study with all the defects of retrospective studies with lack of randomization and unequal group sample sizes.

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