Impact of Bilateral Superior Venae Cavae on Outcome of Staged Fontan Procedure

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Background. The presence of bilateral superior venae cavae may add complexity to the performance of a bidirectional Glenn procedure (BDG). Stagnation of blood flow between the two cavopulmonary anastomoses may increase the risk of thrombosis and impair central pulmonary artery growth.

Methods. Forty patients underwent BDG from January 2004 to April 2011. The cohort was divided into two groups: those receiving bilateral BDG (b-BDG, n = 13) and those receiving unilateral BDG (u-BDG, n = 27). Operative, angiographic, and follow-up data were analyzed retrospectively.

Results. None of the patients experienced thrombosis. There was no difference in actuarial survival rate (u-BDG vs b-BDG, 100% vs 92% at 5 years, $p = 0.15$). On follow-up angiography, no difference in central pulmonary artery index was noted (78.4 ± 45.5 vs 60.4 ± 32.1, $p = 0.24$). Central pulmonary artery stenosis was detected in 6 patients (4 with u-BDG and 2 with b-BDG), 4 of whom (2 from each group) underwent balloon pulmonary artery plasty before the Fontan procedure. There was no difference in freedom from reintervention for central pulmonary artery stenosis (93% vs 85% at 1 year, $p = 0.59$). The rate of Fontan completion was comparable between groups, with similar operative variables and satisfactory outcomes.

Conclusions. Bilateral BDG did not increase the risks of thrombosis and central pulmonary artery hypoplasia and can be performed safely without altering the outcome of the Fontan procedure.

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The bidirectional Glenn (BDG) procedure frequently precedes a staged Fontan procedure. The BDG will normalize volume loading of the single ventricle and reduce the risks associated with the Fontan procedure, particularly in high-risk patients [1]. However, the presence of bilateral superior venae cavae (SVCs) may compromise the outcome of BDG [2, 3]. Pulmonary blood flow supplied by bilateral SVCs competes at the level of the pulmonary artery (PA) confluence, leading to flow stagnation and increased risk of thrombus formation. Moreover, it may impair the growth of the central PA and necessitate PA reconstruction at the time of the Fontan procedure. There are several BDG modifications, including interposition of a prosthetic graft between both SVCs [4] and unifocalization of the two SVCs [5, 6]. Other reports, however, show that conventional bilateral BDG can be safely performed in patients with bilateral SVCs [1, 7]. The purpose of this study was to (1) determine the actual risk of bilateral BDG, focusing on central PA growth and reintervention, and (2) assess the impact of bilateral BDG on the outcome of a staged Fontan procedure.

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Patients and Methods

Patients

We retrospectively reviewed all patients who underwent BDG between January 2004 and April 2011 at our institution. During this period, 41 patients with a functional single ventricle underwent BDG. One patient, who had interruption and azygos continuation of the inferior vena cava, underwent a Kawashima procedure and was excluded; thus, 40 patients constituted the study group. The patients were divided into two groups: unilateral BDG (u-BDG) and bilateral BDG (b-BDG). Bilateral SVCs were present in 14 patients, and 13 underwent b-BDG. One patient, who had a large bridging vein with a normal-sized right SVC and a small left SVC, underwent u-BDG with ligation of the left SVC and was included in the u-BDG group. Data were collected by a retrospective review of medical records. The patient characteristics are shown in Table 1. The median follow-up time was 59 months (range, 4 to 114 months).

Surgical Procedures

All BDG procedures were performed with the patients under standard cardiopulmonary bypass. In patients with bilateral SVCs, both were routinely cannulated unless one or the other was too small to cannulate. Myocardial protection was performed with cold crystalloid cardioplegia solution containing 5% albumin combined with topical cooling when an intracardiac procedure was

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Table 1. Patient Characteristics

Characteristic	u-BDG (n = 27)	b-BDG (n = 13)	*p* Value
Age at BDG, mo	18.8 ± 11.8	11.1 ± 3.8	0.004
Weight at BDG, kg	8.5 ± 2.3	7.8 ± 1.3	0.350
Diagnosis			
TA	5	4	
PA/IVS	5	0	
Ebstein's anomaly	3	0	
DORV	2	0	
Unbalanced AVSD	1	1	
ccTGA	1	0	
PA/VSD	0	2	
Other single ventricles	10	6	
Heterotaxia	3	3	
Dominant ventricular morphology			
Left	17	6	
Right	10	7	

AVSD = atrioventricular septal defect; b-BDG = bilateral bidirectional Glenn procedure; BDG = bidirectional Glenn procedure; ccTGA = congenitally corrected transposition of the great arteries; DORV = double-outlet right ventricle; PA/IVS = pulmonary atresia with intact ventricular septum; PA/VSD = pulmonary atresia with ventricular septal defect; TA = tricuspid atresia; u-BDG = unilateral bidirectional Glenn procedure.

Table 2. Procedures Performed Concomitantly With Bidirectional Glenn Procedure

Procedure	u-BDG (n = 27)	b-BDG (n = 13)
Atrial septectomy	13	7
PA plasty	11	2
No patch	2	2
Pericardial patch	7	0
PTFE patch	2	0
MPA division or ligation	9	5
PA banding	5	2
Atrioventricular valvuloplasty	2	2
Systemic-to-PA shunt	2[a]	1[b]
TAPVR repair	2	0
Other procedures	2[c]	2[d]
Additional PBF	11	4

[a] Central shunt. [b] Modified Blalock-Taussig shunt. [c] Includes one each right ventricular exclusion and implantation of a permanent pacemaker. [d] Includes one each Damus-Kaye-Stansel operation and semiclosure of the mitral valve.

b-BDG = bilateral bidirectional Glenn procedure; MPA = main pulmonary artery; PA = pulmonary artery; PBF = pulmonary blood flow; PTFE = polytetrafluoroethylene; TAPVR = total anomalous pulmonary venous return; u-BDG = unilateral bidirectional Glenn procedure.

necessary. The SVC was anastomosed end to side to the ipsilateral PA with a continuous 6-0 or 7-0 polydioxanone suture. In patients with bilateral SVCs, b-BDG anastomoses were created on the proximal portions of bilateral pulmonary arteries to form a U-shaped appearance on angiography. Previous systemic-to-PA shunts were either divided or ligated, and the SVCs were anastomosed to the shunt insertion site if they were sufficiently located on the proximal PA. Nineteen patients in the u-BDG group and 9 in the b-BDG group had antegrade pulmonary blood flow (PBF) from a ventricle through the main PA at the time of BDG that was left alone in 5 patients and disrupted in 16 patients, and PA banding was performed in 7 patients. Patent ductus arteriosus was either divided or ligated. Because of insufficient systemic oxygen saturation, 3 patients required a systemic-to-PA shunt. As a result, an additional source of PBF was left in place in 15 patients (11 with u-BDG and 4 with b-BDG). Concomitant procedures are shown in Table 2.

Postoperative Antithrombotic Therapy

Nine u-BDG patients (33%) and 6 b-BDG patients (46%) received unfractionated heparin during the early postoperative period. Two patients received heparin 400 U/kg/day by bolus intravenous injection, and the rest received heparin by continuous intravenous infusion at a dosage of 200 to 400 U/kg/day in an attempt to maintain an activated clotting time between 130 and 150 seconds. Aspirin 1 mg/kg/day was given to all patients but 1 (who had undergone one-and-a-half ventricular repair). Indication for warfarin therapy depended on the cardiologist's preference. Fifteen u-BDG patients (56%) and 6 b-BDG patients (46%) received low-dose warfarin titrated to maintain a target international normalized ratio of 1.5.

Angiographic Measurements

Pulmonary angiographic measurements were performed after BDG to assess the PA configuration. In the anteroposterior projection, the diameters of the right and left PAs were measured at the hilum, just proximal to the takeoffs of the first branches, and the diameter of the central PA was measured at the narrowest portion. The cross-sectional areas of the PA were calculated for these three segments, indexed to the patient's body surface area, and expressed as cross-sectional area index (CSAI) for each segment. Nakata's PA index was calculated as the sum of the right and left CSAIs. To assess PA growth, the percentage change (Δ%) of the PA diameter (PAD) was calculated as follows: ΔPAD% = (PAD after BDG − PAD before BDG) / PAD before BDG × 100. Central pulmonary artery stenosis (PS) was defined as segmental or diffuse narrowing with a minimum diameter less than 50% of the average of the right and left pulmonary arteries.

Data Analysis

Data are presented as frequencies, medians with ranges, and means with standard deviations. Differences between groups were analyzed by Student's *t* test, χ^2 test, and analysis of variance. Actuarial survival and freedom from reintervention of the PA were analyzed by Kaplan-Meier estimates and compared between groups by use of a log-rank test. A p value < 0.05 was considered significant.