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Is Interrupted Inferior Vena Cava a Risk Factor in Cases of Bilateral Bidirectional Glenn?

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ABSTRACT

Introduction: Bilateral superior vena cava–to–pulmonary artery anastomoses are technically challenging. Bilateral superior vena cavae (SVCs) have been thought to be a risk factor for poor outcome in children needing single-ventricle palliation.

Methods: The files of forty children who underwent bilateral cavopulmonary anastomoses (CPAs) were reviewed.

Results: Forty patients (31 male, 9 female) had bilateral bidirectional Glenn shunts in King Faisal Specialist Hospital and Research Center, Jeddah, in 7 years. Interrupted inferior vena cava (IIVC) was present in 8 patients. All IIVC cases featured a hypoplastic right ventricle. Twenty-four patients had a hypoplastic right ventricular morphology, and 16 patients had a hypoplastic left ventricular morphology.

Conclusions: In single-ventricle anatomy, cases of a bilateral SVC are more often associated with an IIVC than a single SVC. Patients who undergo bilateral CPAs with an IIVC have a difficult early postoperative course. We should look for IIVC and either exclude or prove IIVC in cases of bilateral SVCs. Postoperative anticoagulation therapy in children with bilateral CPAs is important but should be investigated further.

INTRODUCTION

Systemic venous anomalies are no longer considered major risk factors for the Fontan operation; however, the presence of a bilateral superior vena cava (SVC) still poses an increased challenge to the accomplishment of safe and reliable cavopulmonary anastomoses (CPAs) [McElhinney 1997].

The presence of a bilateral SVC, however, may pose a technical challenge to the performance of a cavopulmonary connection. It also produces a different flow pattern in the central pulmonary arteries compared with a standard bidirectional cavopulmonary connection. Most importantly, a bilateral SVC produces flow stagnation at the level of the pulmonary arterial confluence, leading to increased risk of thrombus formation and unfavorable growth of the central pulmonary arteries [Iyer 2000].

Application of the concepts for the Fontan circulation has evolved whereby nearly all patients with a single-ventricle physiology, including those with abnormalities of the systemic venous return, are given palliative treatment with what has become known as the "Fontan tract," although the initial results for the extension of the indications for the Fontan procedure have not always been favorable [Choussat 1978].

Performing a cavopulmonary connection as a stage of the Fontan procedure has been used as a method to normalize volume loading of the single ventricle at an earlier age. Normalization of volume loading is thought to reduce the risk for a Fontan procedure, particularly in children with high-risk complex lesions [Pridjian 1993].

Factors that have historically been associated with increased operative risk include abnormalities of the systemic venous connection (eg, an interrupted inferior vena cava [IIVC] or a bilateral SVC), a partial or total anomalous pulmonary venous connection (APVC), a common atrioventricular valve that is incompetent, and a morphologic right ventricle supporting the systemic circulation. In addition, the presence of sinus-node and conduction-system abnormalities, which are found in virtually all patients with heterotaxy syndrome, may increase the risk of early or late postoperative arrhythmia [Kim 2006].

Few studies have reported on outcomes in children who undergo bilateral bidirectional CPA, and far fewer, if any, have reported on outcomes in children with IIVC who undergo bilateral bidirectional CPA. We first describe patient characteristics, management, and outcomes related to bilateral CPAs and then compare the outcomes of the bilateral bidirectional Glenn (BBDG) shunt in patients with a normal IVC and those with an IIVC.

PATIENTS AND METHODS

The files of all patients who underwent bilateral CPA anastomoses between December 2000 and October 2007 at King Faisal Specialist Hospital and Research Center, Jeddah, were reviewed. The patients were divided into 2 groups according to the presence or absence of IIVC. Data were collected by reviewing hospital and clinical records. All survivors

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Table 1. Data of all Patients with Bilateral Superior Vena Cava–to–Pulmonary Artery Anastomosis*

Patient Characteristic	
Hypoplastic ventricular morphology, n	
Hypoplastic RV	24
Hypoplastic LV	16
Sex, n	
Male	31
Female	9
Age, mo	19.3
Interrupted IVC, n	8 (hypoplastic right ventricle)
Normal IVC, n	32
Preoperative PAP, mm Hg	27
Postoperative PAP, mm Hg	16
ICU stay, d	2.8
Hospital stay, d	10
Chylothorax, n	4
Arrhythmias, n	5
Reoperation, n	4
Follow-up mortality, n	3 (1, interrupted IVC; 2, post-Fontan)

*Data for age, pulmonary artery pressure (PAP), intensive care unit (ICU) stay, and hospital stay are presented as the mean. RV indicates right ventricle; LV, left ventricle; IVC, inferior vena cava.

were followed up (mean follow-up, 20 months; range, 3-72 months). All statistical analyses were performed with SAS version 6.12 software (SAS Institute, Cary, NC, USA) with the default settings. The Pearson chi-square test was used, and a *P* value <.05 was considered statistically significant. All bilateral bidirectional cavopulmonary shunts were performed via a median sternotomy and with standard cardiopulmonary bypass techniques. No cardioplegia was given. Previous systemic–to–pulmonary artery shunts were disrupted by division, ligation, or clipping. The left SVC was occluded while the pressure in the cephalad side in that SVC was monitored. If the pressure was >20 mm Hg, the left SVC was cannulated for cardiopulmonary bypass, and the procedure was completed with the creation of bilateral end-to-side CPAs. Pressure monitoring was necessary in the first few cases; however, we currently routinely perform the BBDG shunt without pressure monitoring.

RESULTS

A hypoplastic right ventricle was present in 24 cases, and a hypoplastic left ventricle was present in 16 cases. Thirty-one of the patients were male. The mean age was 19.3 months. The IVC was interrupted in 8 cases and normal in 32 cases. The mean preoperative pulmonary artery pressure (PAP) was 27 mm Hg, and the mean postoperative PAP decreased significantly postoperatively to 16 mm Hg. The mean (±SD) preoperative oxygen saturation was 75% ± 5.1% in the IIVC group and 72.6% ± 4.7% in the normal IVC group. The postoperative oxygen saturation was 81.5% ± 5.2% in the IIVC group and 82.2% ± 4.4% in the normal IVC group. The IIVC group had significantly higher rates of arrhythmias, reoperation, and chylothorax, and had a greater need for mechanical ventilation, nitric oxide, and oscillator ventilation. The mean intensive care unit (ICU) stay was 2.8 days, and the mean hospital stay was 10 days, with nonsignificantly longer ICU and hospital stays in the IIVC group. There was a significantly higher rate of reoperation in the IIVC group (*P* = .031; Tables 1 and 2).

COMMENTS AND DISCUSSION

Right isomerism (asplenia syndrome) is suspected when a patient has a single-ventricle anatomy and one or more of the following types of cardiac defects: (1) juxtaposition of the aorta and the IVC, (2) an unroofed coronary sinus, (3) a total APVC to a systemic vein, (4) a common atrioventricular canal, (5) a double-outlet right ventricle or transposition of the great vessels with a bilateral or subaortic conus, and (6) pulmonary valve stenosis or atresia. Left isomerism (polysplenia syndrome) is suspected when a patient has a single-ventricle anatomy and one or more of the following types of cardiac defect: (1) an interrupted IVC with azygos vein continuation, (2) total or partial APVC to the right atrium, (3) a complete or partial atrioventricular canal, and (4) normally related great vessels or a double-outlet right ventricle without subaortic conus [Kim 2006].

Although we believe in this classification, we did not follow it. This approach is simpler, more practical, and more concise. It was not commented on in our echocardiography reports.

A BBDG shunt has been performed for 40 patients since our center opened 7 years ago. We have performed >200 cases of the unilateral bidirectional Glenn (UBDG) shunt. Thus, the ratio of BBDG to UBDG operations is approximately 20%. This percentage is higher than the percentages reported by Iyer et al [2000], Chowdhury et al [2001], and Kim et al [2006], who reported incidences of 14.2% (39/274), 16% (8/50), and 3.2% (10/311), respectively.

The mean patient age was 19.3 months in our series (range, 3-60 months); the median age was 28 months. There were 14 patients in the first year of age, 12 in the second year, 6 in the third year, 4 in the fourth year, 2 in the fifth year, and 2 in the sixth year. The median age in the series of Iyer et al [2000], series was 11 months (range, 3-200 months).

IIVC was involved in 20% of the cases of BBDG shunt. Bilateral SVCs with single-ventricle anatomy were associated with IIVC in 20% of the cases, and single SVCs with single-ventricle anatomy were associated with IIVC in 1.5% of the cases. This prevalence is very interesting and surprising and must be taken seriously, because it is very high, an important observation, because a BBDG shunt will be sufficient without requiring a further Fontan procedure and because the presence of an IIVC will modify the steps and details of the operation. The azygos vein will be kept without clipping or interruption (Kawashima operation). We need to

anticoagulate such patients with both aspirin for life and Coumadin for 3 months. Bilateral SVCs were more often associated with IIVC than single SVCs (20% versus 1.5%). This finding is different from that of Kim et al [2006], who found IIVC to be more frequently associated with single SVC. The mean preoperative PAP was 27 mm Hg (range, 14-58 mm Hg), and the mean postoperative PAP was 16 mm Hg (range, 7-23 mm Hg). This preoperative PAP is different from the PAPs found in the series of Marvoudis et al [1999].

There was no significant difference between the 2 groups, but the there was a significant reduction in the PAP in both groups. We fully agree and believe that patients who fall into high-risk groups should receive single-ventricle palliation. Our procedure is first to review all of the data in detail. Then, full consideration is given as to whether there are any surgically (or interventionally) correctable lesions, such as atrioventricular valve regurgitation that is amenable to repair, or isolated stenoses or hypoplasia within the central pulmonary arteries. Ventricular function, if impaired, may be optimized by altering medical management (with vasodilators and/or diuretic therapy). Evidence of increased pulmonary vascular resistance is treated with a course of pulmonary vasodilator therapy (sildenafil). The patient is then reassessed for suitability for a possible completion of a Fontan procedure. Therefore, many of the original commandments may not be strict requirements for a successful outcome. Some studies have reported that total cavopulmonary connection can be successfully performed on many patients younger than 4 years. Sinus rhythm can be maintained pharmacologically or with atrioventricular pacing. The venous drainage can be surgically corrected, and enlarged atria may undergo reduction atrioplasty. Discrete stenosis in otherwise normal central pulmonary arteries can now be managed by stenting or surgical enlargement and would not be considered a contraindication to total cavopulmonary connection if pressures are otherwise normal [Hosein 2007].

Table 2. Comparison of Bilateral Superior Vena Cava–to–Pulmonary Artery (SVCP) Anastomoses with Interrupted Inferior Vena Cava (IVC) and Bilateral SVCP Anastomoses with a Normal IVC*

Characteristic	Bilateral SVCP Anastomosis with Interrupted IVC	Bilateral SVCP Anastomosis with Normal IVC	*P*
Patients, n	8	32	
Sex, n			
Male	5	26	
Female	3	6	
Hypoplastic RV	8	16	.001
Hypoplastic LV	0	16	
Oxygen saturation, %†			
Preoperative	75.0 ± 5.1	72.6 ± 4.7	.139
Postoperative	81.5 ± 5.2	82.2 ± 4.4	.139
Pulmonary artery pressure, mm Hg			
Preoperative	28	26	
Postoperative	17	16	
Mechanical ventilation, d	1.6	1.0	.001
Nitric oxide need, d	1.1	0.5	.001
Oscillator need, d	0.8	0.2	.001
Bypass time, min	64	68	
ICU stay, d	3	2.4	
Hospital stay, d	11	9.3	
Reoperation, n	2	2	.031
Thrombosis and technical stenosis	1	0	
Bleeding	0	2	
Incorporation of the hepatic veins	1	0	
Significant arrhythmias, n	2	3	.031
Chylothorax, n	3	1	.003

*Data are presented as the mean for oxygen saturation, pulmonary artery pressure, mechanical ventilation, nitric oxide need, oscillator need, bypass time, intensive care unit (ICU) stay, and hospital stay. RV indicates right ventricle; LV, left ventricle.

†Data are presented as the mean ± SD.

There was a significant difference between the 2 groups with respect to the early postoperative course. It was more difficult in the IIVC group. Patients in the IIVC group required longer inotropic support (6 hours versus 2 hours), longer mechanical ventilation (1.6 days versus 1.0 days), longer nitric oxide need (1.1 day versus 0.5 days), longer oscillator (high-frequency jet ventilation) need (0.8 days versus 0.2 days), and longer ICU stays (3 days versus 2.4 days). Such patients also had a higher incidence of significant arrhythmias (25% versus 9.3%), a higher frequency of chylothorax (37.5% versus 3.1%), and longer hospital stays (11 days versus 9.3 days). There was also a significantly higher rate of reoperation in the IIVC group (20% versus 6.2%). All of the chylothorax patients were treated with conservative treatment: keeping the chest tubes in, nothing by mouth, a fat-free diet with octreotides with or without steroids. None of these patients required surgical management for chylothorax.

One patient in the IIVC group had a massive edema of the head and upper half of the body with desaturation. An echocardiography evaluation showed no flow through the shunt. An urgent exploration revealed stenosis of the left Glenn shunt with a thrombus in the segment between the 2 shunts. The thrombus was removed, and the shunt was revised. The patient did well postoperatively. Another patient with IIVC died 1 year after the BBDG shunt after trial incorporation of the hepatic veins because of pulmonary atrioventricular malformations. Significant bleeding was surgically controlled in 2 patients of the normal-IVC group. Two patients died in the ICU after the Fontan procedure because of irreversible heart failure.

Limitations of the Study

First, this study was retrospective in nature. Although we believe in the classification of right and left atrial isomerism, we did not follow it. The practice we have described will be simpler, more practical, and more concise. In addition, the number of cases is small, especially in the IIVC group.

CONCLUSIONS

The prevalence of IIVC with the BBDG shunt in this series is significant (20%), with higher rates of thrombosis and reoperation and with a more difficult early postoperative course. In cases of bilateral SVCs, evaluations should be conducted to exclude or prove IIVC. Postoperative anticoagulation treatment in children with bilateral CPAs is important but requires further investigation.

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