Systemic Venous Collateral Development after the Bidirectional Cavopulmonary Anastomosis

Prevalence and Predictors

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Objectives. To determine the prevalence of systemic venous collaterals after the bidirectional cavopulmonary anastomosis and the factors associated with their development.

Background. Systemic venous collaterals have been found after cavopulmonary anastomosis.

Methods. Cardiac catheterization was performed in 103 patients before and after a bidirectional cavopulmonary anastomosis.

Results. After surgery, 51 venous collaterals were identified in 32 patients (31%). Collateral development was associated with an abnormal superior vena caval connection (56% incidence vs. 26% with a single right superior vena cava, \( p = 0.01 \)) and postoperative factors including pulmonary artery distortion (53% incidence vs. 22% without distortion, \( p = 0.002 \)); increased superior vena caval mean pressure (14 ± 5 mm Hg versus 11 ± 4 mm Hg with no collaterals, \( p = 0.0002 \)); increased pulmonary artery mean pressure (13 ± 4 mm Hg vs. 11 ± 4 mm Hg with no collaterals, \( p = 0.02 \)); lower right atrial mean pressure (5 ± 3 mm Hg vs. 6 ± 3 mm Hg with no collaterals, \( p = 0.04 \)); and increased mean gradient between superior vena cava and right atrium (8 ± 3 mm Hg vs. 5 ± 4 mm Hg with no collaterals, \( p = 0.0002 \)). Using multiple logistic regression, only this last factor was independently associated with collateral development with an odds ratio per 1 mm Hg of 1.33 (95% CI 1.12–1.58, \( p = 0.001 \)) for their presence.

Conclusions. Systemic venous collaterals occur frequently after a bidirectional cavopulmonary anastomosis and are found postoperatively when a significant pressure gradient occurs between cava and right atrium.

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The bidirectional superior cavopulmonary anastomosis (BCPA; also known as the bidirectional Glenn), while originally introduced as interim palliation for the high-risk Fontan candidate (1), is also employed as definitive palliation (2) or as a component of a biventricular repair (the so-called “one and a half ventricle repair”). In a growing number of centers, many Fontan candidates are now undergoing staged palliation in the form of a BCPA, regardless of the presence or absence of risk factors (3).

After a “classic” Glenn anastomosis (end-to-end anastomosis of the superior vena cava [SVC] to the pulmonary artery [PA]), the development of venous connections between the superior and the inferior caval veins or right atrium can lead to progressive cyanosis (4,5) due to reduction in effective pulmonary blood flow, coupled with the increasing oxygen demands with growth. Systemic venous to hepatic collaterals have also been reported after the BCPA (6), particularly in the setting of left atrial isomerism (7), and may also result in profound and progressive cyanosis. Such collateral vessels may develop in the absence of mechanical obstruction, and represent “run-off” from the higher pressure caval venous system to the lower pressure atrium or hepatic vein. Recent reports have also highlighted the development of systemic venous collaterals after the Fontan procedure (8).

The aims of this study were to review those patients who had been palliated with a BCPA to determine the prevalence and morphology of such systemic venous channels, whether their presence was disadvantageous, and if anatomical and hemodynamic factors were associated with their presence.

Methods

Patient population. Between June 1982 and September 1995, 192 patients underwent a BCPA. Patients were excluded if pre- and/or postoperative cardiac catheterization data were unavailable. The remaining study population consisted of 103
Abbreviations and Acronyms

BCPA = bidirectional cavopulmonary anastomosis
IVC = inferior vena cava (caval)
PA = pulmonary artery
SVC = superior vena cava (caval)

In cases where doubt existed regarding morphology, the designation “indeterminate” was assigned. The right and left PA diameters were measured immediately proximal to their first branches using catheter diameter to correct for magnification and used to calculate cross-sectional areas. The combined PA areas were expressed as the Nakata index (9).

Because some PAs were distorted due to previous arterial shunts or constriction of ductal tissue, an attempt was made to categorize the degree of distortion: 1) no distortion, 2) mild distortion of one or both PAs not requiring intervention, 3) severe distortion of one PA requiring intervention either before or at the time of the next surgery, and 4) severe distortion of both PAs.

While most postoperative catheterizations were performed as scheduled assessments, a number were performed early after surgery in patients with low saturations and/or high SVC pressures. At the time of the postoperative assessment, the Nakata index was remeasured, the presence or absence of pulsatile flow in the pulmonary circuit was noted, and PA distortion was characterized as above.

Systemic venous collaterals were defined as venous channels allowing flow from the BCPA circuit to the inferior vena cava (IVC), atrium, or pulmonary veins, and could represent reversal of flow within an existing vessel or opening of a new channel. The location, origin, and course of such channels were recorded and subdivided into those running in the anterior, mid, or posterior mediastinum from lateral angiographic views.

Analysis of hemodynamics. Oxygen saturations were obtained in the aorta and the SVC, and the inspired oxygen concentration was noted. The majority of patients were breathing room air, however, several of those studied in the immediate postoperative setting were ventilated in an enriched oxygen mixture. Mean pressures were recorded in the SVC, the right atrium, and the main or branch PAs, together with the end-diastolic pressure of the dominant ventricle. Stenosis of the anastomosis was detected by both mean pullback pressures between the PA and SVC and by the angiographic appearance. Cardiac index was calculated using measured oxygen consumption in patients breathing room air. In those situations where it was possible to measure pulmonary blood flow (single source flow), the pulmonary arterial resistance index was calculated.

Surgical data. The cavopulmonary anastomosis was performed on cardiopulmonary bypass following a midline sternotomy. An end-to-side anastomosis of the SVC to the PA was constructed using interrupted sutures with 6-0 Prolene (Ethicon, Johnson and Johnson). The atrium was closed in two layers also using 6-0 Prolene. The type of cavopulmonary anastomosis, whether unilateral or bilateral, was noted. Excluding cases of left atrial isomerism with azygous continuation of the IVC, the azygous vein was ligated in all but seven patients by the surgeon’s preference. In one patient the azygous vein could not be identified. An atrial septectomy was required in 11 patients, the remaining having unrestricted atrial septal defects.

Summary of risk factors. Possible preanastomosis risk factors for the development of systemic venous collaterals and those associated with the presence of collaterals postanastomosis are summarized in Table 2. Ventricular function and degree of atrioventricular valve regurgitation were not included in the analysis, as this information was qualitative and thought to be of limited validity.

Data analysis. Population characteristics are reported as frequencies, medians with ranges, and means with 1 standard deviation. Risk factors for the presence or absence of venous collaterals in a patient were sought using Fisher’s exact tests, chi-square tests, and Kruskal-Wallis ANOVA. Significant predictors associated with collateral formation were further explored in multiple logistic regression analysis with a forward and backward stepwise variable selection with entry criteria set at p < 0.05. All analyses were performed using SAS statistical software (SAS Institute Inc.).
Table 2. Risk Factors Assessed for the Development of Systemic Venous Collaterals

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Site</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td>Age at surgery</td>
<td>504</td>
</tr>
<tr>
<td>Time between surgery and postoperative catheterization</td>
<td>13.8 years</td>
<td>32</td>
</tr>
<tr>
<td>Morphology of dominant ventricle (right, left or indeterminate)</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Type of caval connection</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Ligation of a left superior vena cava</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Presence of pulsatile flow in the pulmonary circuit</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Degree of pulmonary artery distortion</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Presence of previous ipsilateral Blalock-Taussig shunt</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Mean pressure in the superior vena cava</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Mean pressure in the pulmonary artery*</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Mean pressure in the right atrium*</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>End-diastolic pressure in the dominant ventricle*</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Mean pressure gradient between superior vena cava and right atrium</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Mixed venous oxygen saturation*</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Systemic arterial oxygen saturation*</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Cardiac index*</td>
<td>1.3 years</td>
<td>32</td>
</tr>
<tr>
<td>Pulmonary arterial resistance index (after surgery only)</td>
<td>1.3 years</td>
<td>32</td>
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</tbody>
</table>

*The difference between pre- and postoperative values for these risk factors was also assessed as a predictor.

Results

Demographic and anatomical data. The median age of catheterization prior to the BCPA was 1 year (range 1 day to 13.8 years), the median age at surgery was 1.3 years (range 0.3 to 14.3 years), and the median time from surgery to postoperative catheterization was 1.3 years (range 1 day to 8.4 years), with eight patients requiring catheterization within 2 weeks of surgery. There were 14 deaths (14% mortality): four occurring <2 weeks after surgery, one patient with widespread thrombus within the pulmonary circulation died 3 months after surgery, and the remaining nine patients died after further surgical procedures.

The morphology of the dominant ventricle was left in 64 patients (62%), right in 38 patients (37%) and in one patient both ventricles were of equal size (1%). The SVC connection was single in 85 patients (82%) and bilateral in 18 patients (18%) (the left SVC draining into the coronary sinus in nine patients and to the roof of a common atrium in nine patients). Four of these patients underwent ligation of the left SVC, 13 underwent bilateral BCPA, and in one patient with left atrial isomerism, the left SVC was left in continuity with the heart.

Postoperative PA flow appeared pulsatile during angiography in 49 patients (48%), either having the main PA in continuity with the heart (n = 36) or having a systemic arterial shunt (n = 13). Postoperative angiography showed that 73 patients had no PA distortion (71%), 15 patients had mild distortion (14%), 11 patients had severe distortion affecting one PA (11%) and four patients had severe distortion affecting both PAs (4%).

Prevalence and types of collaterals. No abnormal systemic venous channels were identified at the time of preoperative catheterization. At the time of postoperative catheterization, a total of 51 sites of collateral venous drainage were identified in 32 patients (31%). Types of channels are summarized in Table 3.

The eventual destination of collateral flow was to the right upper pulmonary vein, to a common atrium (left isomerism, see below) and to the coronary sinus each in one patient. In this last patient (Fig. 1), a tiny superior intercostal vein became a large left SVC draining to the coronary sinus following the BCPA. In the remainder, blood eventually drained to the IVC. Two examples are shown (Figs. 2 and 3). The patient in Figure 2 had a dilated pericardial vein that drained to the IVC, and the patient in Figure 3 had decompression via a right internal mammary vein. Of the seven patients who did not have azygous vein ligation, decompression into the azygous vein developed in five, and in two of these patients, this was the only source of venous decompression. Only one patient with left atrial isomerism and azygous vein continuation of the IVC developed collateral run-off, which drained via the azygous vein (Fig. 4A) to the left renal vein, and then from there through a new channel to the hepatic portal system (Fig. 4B). Blood then drained through the liver parenchyma to the hepatic vein, and thence to the right atrium.

Forty-nine channels coursed through the mediastinum (anterior mediastinum in 4, mid-mediastinum in 16, and posterior mediastinum in 29). Two channels were outside the mediastinum: the left renal to hepatic portal channel mentioned above and cervical veins draining from the left to the right SVC in one additional patient.

Management of collaterals. Twelve patients with collaterals required treatment (38%). Coil occlusion during cardiac catheterization was performed in four patients, one of whom also underwent dilatation of a previously implanted endovascular stent at the anastomotic site and another who underwent placement of an endovascular stent within a narrowed left PA. One patient underwent successful balloon angioplasty of the anastomotic site and of a narrowed distal left PA with disappearance of collateral flow, and one patient underwent endovascular stent placement within the left PA with similar reduction in collateral flow. Collaterals in two patients were ligated at the time of Fontan completion, and one patient...
underwent successful surgical revision for BCPA stenosis with subsequent reduction in collateral flow.

There were three deaths: one patient after attempted thrombolysis for widespread thrombosis following homograft reconstruction of disconnected PAs and bilateral BCPA; one patient after attempted balloon occlusion of a large hemi-azygous vein, and one patient during attempted surgical decompression for SVC syndrome.

**Hemodynamics.** Hemodynamic measurements before and after BCPA are given in Table 4. Only the SVC and aortic saturations, the PA pressures and the Nakata index changed significantly. Saturation data collected while patients were receiving enriched oxygen (FiO$_2$ = 0.25) were not included for analysis. The difference in pulmonary arteriolar resistance was not assessed as pulmonary blood flow could not be calculated in the majority of preoperative studies.

**Univariate analysis of risk factors.** Comparing patients with and without collaterals, the following features were associated with their appearance: preexisting bilateral SVC connection with an incidence of 56% compared with 26% for single right SVC (p = 0.01), and postoperative factors including distortion of the PAs with an incidence of 53% versus 22% for no distortion (p = 0.002), increased SVC mean pressures of 14 ± 5 mm Hg versus 11 ± 4 mm Hg for no collaterals (p = 0.0002), higher PA mean pressures of 13 ± 4 mm Hg versus 11 ± 4 mm Hg for no collaterals (p = 0.02), and lower right atrial mean pressures of 5 ± 2 mm Hg versus 6 ± 3 mm Hg for no collaterals (p = 0.04). The mean postoperative gradient

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**Figure 1.** Patient with double outlet right ventricle, multiple ventricular septal defects, and a straddling right atrioventricular valve who underwent a BCPA at 1 year of age. A) A preoperative left brachiocephalic vein injection is shown in the frontal projection. There is a good-sized right superior vena cava (SVC) and filling of a tiny superior intercostal vein (arrow). Postoperative left SVC injection (LSVC) can be seen in the frontal (B) and lateral (C) projections. An enlarged LSVC is now seen connecting to coronary sinus (CoS) with areas of dilatation and narrowing. The SVC was ligated at the time of Fontan completion.
between SVC and right atrium was 8 ± 3 mm Hg for those with, versus 5 ± 4 mm Hg for those without collaterals (p = 0.0002). Eight patients had evidence of obstruction at the anastomotic site, with a mean pullback gradient of ≥3 mm Hg (range 3–9 mm Hg), however, these gradients were not associated with collateral development (p = 0.1). No preoperative hemodynamic factors predicted the development of collaterals, although mean PA pressure tended to be higher at 14 ± 4 mm Hg versus 12 ± 4 mm Hg for no collaterals (p = 0.1).

Regression analysis. When these predictors were tested using multiple logistic regression, only the postoperative gradient between the SVC and right atrium emerged as independent for collateral development. The odds ratio per 1 mm Hg was 1.33 (95% CI 1.12 to 1.58, p = 0.001).

Discussion

Prevalence and predictors. With careful angiography, systemic venous anomalies can be found frequently after the BCPA. The incidence in this series of 31% is greater than the 17% found by Gross et al. (10), probably because the presence of collaterals was actively sought. Of the variables examined, only the postoperative gradient between the SVC and the right atrium was independently associated with the presence of collaterals. This gradient may be increased by the relative reduction in caval or PA cross-sectional area after ligation of one SVC (in the setting of bilateral cavae), the presence of PA hypoplasia or distortion, or obstruction of the anastomosis. An inadequate PA distribution, increased pulmonary resistance, and pulmonary venous obstruction could also contribute to an elevated caval vein pressure. It might also be possible for marked cyanosis to lead to an increase in transpulmonary gradient by increasing blood viscosity (11).

All patients in this series had acceptable preoperative PA pressures, none had pulmonary venous obstruction, and eight demonstrated obstruction at the anastomotic site, but this factor alone did not predict the appearance of venous anom-
An elevated ventricular end-diastolic pressure, which can reflect ventricular diastolic dysfunction, was also not independently associated with their development. In any case, such an elevation in pressure sufficient to result in collateral development would not be compatible with single ventricle physiology.

In our series, the only anatomical factors associated with the presence of collaterals were postoperative PA distortion and the presence of bilateral SVCs, although the mechanism for the latter would not appear to be due to ligation of one caval vein. As previously reported (12), the Nakata index fell significantly after the BCPA, however, a lower postoperative Nakata index was not associated with the presence of collaterals.

Obstruction to vena caval flow can also lead to systemic venous decompression after atrial repair of transposition of the great arteries (13,14), and pulmonary venous decompression has been described after repair of sinus venosus atrial septal defect (15). An anomalous systemic venous to left atrial channel has also been described between the SVC to left atrium in a case of tricuspid atresia and restrictive interatrial communication (16). Thus, a variety of obstructive lesions can form the substrate for the development of a host of possible decompressing channels.

**Mechanism of development.** The etiology of such anomalies is unsettled, representing angiogenesis de novo or the reopening of previously existing channels. To this end, the embryological development of the IVC (described in detail by McClure and Butler in 1925 [17]), has helped elucidate the considerable variations found in the adult. During the course of its development, many venous channels are known to “disappear.” We postulate that such channels reappear when the cavae are subjected to elevated pressures. This was underscored in our population with the reopening of the anterior and/or posterior cardinal systems from the innominate vein via the superior intercostal vein. The speed at which collaterals appear also suggests the reopening of preexisting channels.

**Physiological effects.** The physiologic effect of this collateralization takes two forms, either systemic venous to systemic venous connections, or systemic venous to pulmonary venous or left atrial connections. Increased cyanosis could therefore result from either a reduced effective pulmonary blood flow or increased admixture of pulmonary venous return. Clinically, the overall presence of systemic venous run-off was not associated with significant systemic desaturation, presumably as several were small and represented an insignificant right to left shunt or reduction in effective pulmonary blood flow at rest. Large collaterals did result in clinically unacceptable desaturation and required intervention. In many cases, it was safe to leave anomalous veins draining from superior to inferior vena cava.

### Table 4. Hemodynamic Data Before and After Bidirectional Cavopulmonary Anastomosis

<table>
<thead>
<tr>
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<th>Pre-Op</th>
<th>Post-op</th>
<th>p</th>
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<tbody>
<tr>
<td>SVC saturation</td>
<td>56 ± 9%</td>
<td>66 ± 13%</td>
<td>0.0001</td>
</tr>
<tr>
<td>Aortic saturation</td>
<td>78 ± 7%</td>
<td>85 ± 8%</td>
<td>0.0001</td>
</tr>
<tr>
<td>RA pressure</td>
<td>6 ± 3</td>
<td>6 ± 3</td>
<td>NS</td>
</tr>
<tr>
<td>PA pressure (mm Hg)</td>
<td>13 ± 4</td>
<td>11.5 ± 4</td>
<td>0.04</td>
</tr>
<tr>
<td>Ventricular EDP (mm Hg)</td>
<td>7 ± 2</td>
<td>7 ± 3</td>
<td>NS</td>
</tr>
<tr>
<td>Nakata index (mm²/m²)</td>
<td>341 ± 18</td>
<td>304 ± 145</td>
<td>0.002</td>
</tr>
<tr>
<td>Cardiac index (L/min/m²)</td>
<td>4.7 ± 2.0</td>
<td>4.5 ± 1.4</td>
<td>NS</td>
</tr>
</tbody>
</table>

EDP = end-diastolic pressure, PA = pulmonary artery, RA = right atrial.
cava, as this shunt would be negated after completion of the Fontan circuit, unless they were so large as to seriously reduce effective pulmonary blood flow in the short term. However, channels draining to the pulmonary veins or common atrium required occlusion.

No hemodynamic risk factors could be identified from the preoperative catheterization data, however, in many cases pulmonary vascular resistance could not be estimated as there were two sources of pulmonary blood flow. In addition, hemodynamic measurements during catheterization only represent a snapshot of prevailing conditions. It may be that collateral channels develop in hemodynamically borderline situations and are markers for a poor long-term outcomes. It is intriguing to speculate that these same patients may develop new collaterals after completion of the Fontan circulation and/or further collaterals after occlusion.

**Conclusions.** Systemic venous collaterals are frequently found after the bidirectional cavopulmonary anastomosis, the majority drain to the inferior vena cava, and they are related to a pressure gradient between the cava and right atrium. Collaterals probably represent reverse flow in the azygous or hemi-azygous systems or reopening of channels present during embryonic development.

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**References**