Cardiol Young 2001; 11: 289–294
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ISSN 1047-9511

Original Article

Incidence and predictors for the development of significant supradiaphragmatic decompressing venous collateral channels following creation of Fontan physiology

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Abstract The occurrence of supradiaphragmatic decompressing venous collateral channels following construction of a bidirectional cavopulmonary connection or completion of the Fontan operation resulting in abnormal systemic hypoxemia has been infrequently described. In addition, the incidence and predictors of these channels have not been well delineated, especially in those patients without formation of such structures preoperatively. I evaluated, retrospectively, 40 patients who had undergone either construction of a bidirectional cavopulmonary shunt or completion of the Fontan operation, and who had complete pre and postoperative hemodynamic and angiographic data. Of the patients, 17 (43%) had developed a total of 21 decompressing venous collateral channels, of which 7 (18%) were considered to be hemodynamically significant requiring transcatheter coil occlusion. Of all variables examined, seven patients with significant decompressing collaterals had a greater transpulmonary gradient at follow-up catheterization (8 + /- 2 vs 5 + /- 2 mmHg, p= .01) and lower systemic saturations at routine clinical follow-up visits (82 + /- 5 vs 89 + /- 5 mmHg, p= .007) in comparison to the 33 others. When not evident preoperatively, decompressing venous collateral channels develop in a significant number of patients following conversion to Fontan physiology. If sufficiently large, they may produce lower than expected systemic saturations for the observed cardiac physiology. The larger decompressing channels are more likely to occur when a greater transpulmonary gradient exists postoperatively, which may require cardiac catheterization and transcatheter coil occlusion.

Keywords: Decompressing venous collaterals, coil occlusion

The Fontan operation has been performed for more than 20 years as definitive palliation for patients with functional univentricular physiology. The bidirectional cavopulmonary shunt, or the Hemi-Fontan operation, are now advocated as either interim palliation prior to completing the Fontan circulation or, less commonly, as a final procedure. Subsequent to such procedures, some patients, either early or late, will develop abnormally low systemic oxygen saturations related to the development of decompressing venous collateral channels. These vessels create a communication between the higher pressured superior caval venous system and the lower pressured inferior caval vein or the pulmonary venous atrium in the case of Hemi-Fontan physiology, and exclusively the pulmonary venous atrium in the case of completed Fontan physiology. A few previous studies have examined the etiologies of small and large decompressing venous collateral channels,[1–3] some of which, anatomically, are very unusual.[4] These studies, however, have also included patients with either preexisting venous collaterals, or insufficienct angiography to exclude the presence of such vessels prior to establishment of Fontan physiology. In this study, I have examined both the incidence and predictors for the postoperative development of significant supradiaphragmatic decompressing venous collateral channels

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Accepted for publication 8 October 2000

https://doi.org/10.1017/S1047951101000312 Published online by Cambridge University Press

exclusively in patients known to lack such collateral pathways prior to the establishment of Fontan physiology.

Materials and methods

For inclusion, patients were required to have complete hemodynamic and angiographic evaluation both before and after establishment of Fontan physiology. This included power angiographic injections within the superior caval vein (left, right or both), and if present, the brachiocephalic (innominate) vein, in order to determine whether any supradiaphragmatic decompressing venous collateral channels were present prior to or following a Fontan-type procedure. Balloon occlusion angiography was not utilized in any patients and, therefore, the method of assessment was consistent throughout the period of study. Decompressing venous collateral channels originating below the diaphragm subsequent to completion of the Fontan procedure were excluded in all patients by performing a power angiographic injection at the level of the inferior caval vein below the Fontan baffle. In patients who had undergone construction of a bidirectional cavopulmonary shunt, the azygos vein in presence of a right-sided superior caval vein, or the hemiazygos vein with a left-sided superior caval vein, had intentionally been ligated at the time of surgery. In those patients proceeding to a full Fontan procedure, without an intervening bidirectional cavopulmonary shunt, the azygos or hemiazygos veins were intentionally left open. Only those patients without decompressing venous collateral channels prior to construction of Fontan physiology, excluding the presence of a normal azygos or hemiazygos veins, were included in the analysis.

Hemodynamic variables examined before and after establishment of Fontan physiology are listed in Table 1. Pulmonary vascular resistance in each patient was calculated by obtaining the mean transpulmonary gradient divided by the index for pulmonary flow via a measured consumption of oxygen indexed to body surface area. In those few patients with an additional source of pulmonary blood flow subsequent to the Hemi-Fontan procedure, the pulmonary vascular resistance was

Table 1. Predictors for the development of significant decompressing venous collaterals

	Significant Collaterals (N= 7)	Insignificant Collaterals (N= 33)	p value
% Male vs Female	57 vs 43%	36 vs 64%	NS
LV hypoplasia (%)	43%	45%	NS
Unilateral SCV (%)	100%	85%	NS
Heterotaxia syndrome	29%	24%	NS
Anatomic diagnosis	Unbalanced AVSD (3) Functionally single ventricle/pulmonary atresia(3) Hypoplastic left heart (1)	Hypoplastic left heart variants (10) Tricuspid atresia/small RV (7) Functionally single ventricular physiology (5) Unbalanced AVSD (4) Pulmonary atresia/intact septum(3) Double outlet RV/mitral atresia (4)	NS
RAm pre(mmHg)	6 + /- 2	6 + /- 3	NS
Additional PBF	2 (29%)	2 (6%)	NS
Hemi-fontan physiology	71%	27%	0.07
Age @ surgery (mths)	14 + /- 11	20 + /- 20	NS
Interval since surgery (mths)	24 + /- 15	26 + /- 18	NS
SCVm (mmHg)	14 + /- 4	11 + /- 4	NS
Change in SCVm (mmHg)	6 + /- 5	5 + /- 5	NS
TPG (mmHg)	8 + /- 2	5 + /- 2	0.001
PVR post (units/M2)	2.5 + /- 1.1	2.0 + /- 1.0	NS
Systemic saturation (%)	82 + /- 5	89 + /- 5	0.005
Qs (L/min/M2)	3.2 + /- 0.6	2.8 + /- 0.9	NS

Additional PBF: number of patients with an additional source of pulmonary blood flow (pulmonary artery banding or a systemic to pulmonary artery shunt); Age @ surgery: age in months at the time of hemi or complete Fontan surgery; Anatomic diagnosis: cardiac diagnosis preoperatively. Change in SCVm: change in superior caval venous pressure pre to post hemi or complete Fontan; Hemi-fontan physiology: percentage of patients with a hemi-fontan procedure Heterotaxia syndrome: number of patients with isomeric atrial appendages; Interval since surgery: time since hemi or complete Fontan to catheterization procedure; LV hypoplasia: percentage of patients with hypoplastic left ventricle vs right ventricle;PVR post: pulmonary vascular resistence post hemi or Fontan physiology; Qs: systemic flow index; RAm pre: mean right atrial pressure pre hemi or Fontan physiology; Systemic saturation: systemic oxygen saturation measured at clinical followup pre catheterization;TPG: transpulmonary gradient between superior caval vein and pulmonary venous atrium measured at catheterization; Unilateral SCV: percentage of patients with unilateral vs bilateral caval veins

AVSD - atrioventricular septal defect

https://doi.org/10.1017/S1047951101000312 Published online by Cambridge University Press

estimated by averaging saturations of oxygen and the mean transpulmonary gradients in both the left and right pulmonary arteries.

From 1983–1998, 118 patients have undergone either a bidirectional cavopulmonary connection or completion of the Fontan procedure at our institution. Of these, 40 had complete hemodynamic and angiographic data making them suitable for inclusion in the study. Of the patients, 36 underwent cardiac catheterization electively either prior to completion of the Fontan circulation, or at approximately 1 year after its completion, as part of their routine clinical assessment. The remaining 4 patients underwent cardiac catheterization non-electively due to a significantly lower than expected systemic saturation for their cardiac physiology during routine clinical follow-up. These patients had undergone 3 bidirectional cavopulmonary shunts and 1 fenestrated Fontan procedure, respectively. At cardiac catheterization, a venous collateral channel was considered significant if, via non-selective power angiography, there was significant opacification of the pulmonary venous atrium or inferior caval vein, thus representing a reduction in effective pulmonary blood flow with or without abnormal systemic hypoxemia. The maximal diameter of the decompressing channel was not utilized to differentiate between significant and non-significant vessels, since most were tortuous, with areas of both dilation and constriction. I also did not exclusively use the systemic saturation at cardiac catheterization in patients with the Hemi-Fontan procedure in this differentiation, since with conscious sedation and the patient lying supine, flow may have been altered through the abnormal venous channel. This is supported by my observation that most patients at catheterization with functionally univentricular physiology before and after the Hemi-Fontan procedure in the absence of decompressing venous collateral channels have higher systemic saturations than what is observed during routine clinical follow-up. This is likely related to alterations in extraction and consumption of oxygen.

Non-categoric variables are expressed as the mean + /- 1 standard deviation. Comparisons between groups were performed by utilizing either Student's t-test for unpaired data, or chi squared analysis with continuity correction. A p value of less than 0.05 was considered to be significant.

Results

Of the 40 patients with data suitable for inclusion in the study, 18 had undergone a complete Fontan procedure without an intermediate bidirectional Glenn, whereas the remaining 22 patients have undergone a staged approach. Of these, 10 are either currently waiting, while 12 have completed the Fontan. Amongst the group, 17 patients (43%) have developed 21 decompressing venous collateral channels subsequent to establishing Fontan physiology. In 13 patients, there was development of 1 decompressing channel, whereas 4 patients had 2 vessels, all of which originated from the superior caval venous system (Table 2). In 7 patients (18%), the collateral channels were considered to be hemodynamically significant and were therefore coil embolized at catheterization, resulting in an increase in systemic saturation from 82 + /- 5% to 86 + /- 5% (p= 0.9). The significant channels included a vein running from the brachiocephalic to the inferior caval vein just below the diaphragm in 4 patients following construction of a bidirectional cavopulmonary shunt; a vein running from the brachiocephalic vein and draining above and below the diaphragm to the venous atria in another patient with a bidirectional cavopulmonary shunt; a vein extending from the brachiocephalic vein to the coronary sinus above the diaphragm in one patient following completion of the Fontan circulation, and a channel running from the azygos vein, communicating to the hemiazygos vein and thence to a left superior caval vein and eventually to the coronary sinus above the diaphragm in a patient undergoing completion of the Fontan circulation without an intervening bidirectional cavopulmonary shunt (Figs. 1–3). When the seven patients with haemodynamically significant venous collateral channels were compared to the remaining 33 patients, the only differentiating variables were the presence of a greater transpulmonary gradient at catheterization and a lower systemic saturation by pulse oximetry during routine clinical follow-up immediately prior to their catheterization (Table 1). Although not statistically significant, significant decompressing venous collateral channels were more common in patients with a bidirectional cavopulmonary connection. There was no relationship, however, with the presence of an additional source of pulmonary blood flow or the time interval since performing the bidirectional cavopulmonary shunt or completion of the Fontan procedure.

Discussion

Since the introduction of the Fontan operation more than 20 years ago, the procedure has undergone several modifications, including the application of the bidirectional cavopulmonary shunt or Hemi-Fontan procedure as interim palliation prior to completing the Fontan operation. As

https://doi.org/10.1017/S1047951101000312 Published online by Cambridge University Press

Table 2. Types of decompressing venous collaterals (N= 21)

Cardinal vein from brachiocephalic vein posteriorly to pulmonary venous atriums or ICV (N= 8)
Cardinal vein from brachiocephalic vein posteriorly to CS (N= 6)
Right SCV to ICV or pulmonary venous atriums (N= 4)
Brachiocephalic vein anteriorly to ICV (N= 1)
Azygous vein to hemiazygous vein to CS to left SCV to pulmonary venous atriums (N= 1)
Azygous vein to hemiazygous vein to left SCV to CS (N= 1)

CS: coronary sinus; ICV: inferior caval vein; SCV: superior caval vein

a result of these changes, and better selection of patients, the long term hemodynamic results and survival have improved dramatically for the majority of patients with functionally univentricular physiology. An unusual, but serious, postoperative complication in this group of patients is the development of significant cyanosis out of proportion to either Hemi-Fontan physiology, or the presence of an intentional fenestration in those patients undergoing a completed Fontan procedure. Etiologies include baffle leaks in those with a completed Fontan procedure, development of pulmonary arterio-venous fistulas primarily in older patients with a classic Glenn or completed Fontan which excludes hepatic venous return from the pulmonary circulation, or significant ventilation/perfusion mismatch secondary to lung disease. An additional cause, described by Gross et al,[5] is found when the bidirectional cavopulmonary connection is constructed in patients older than 4 years of age, or with an elevated indexed pulmonary vascular resistance of greater than 5.0. This apparently results in a redistribution of systemic blood flow away from the upper body,

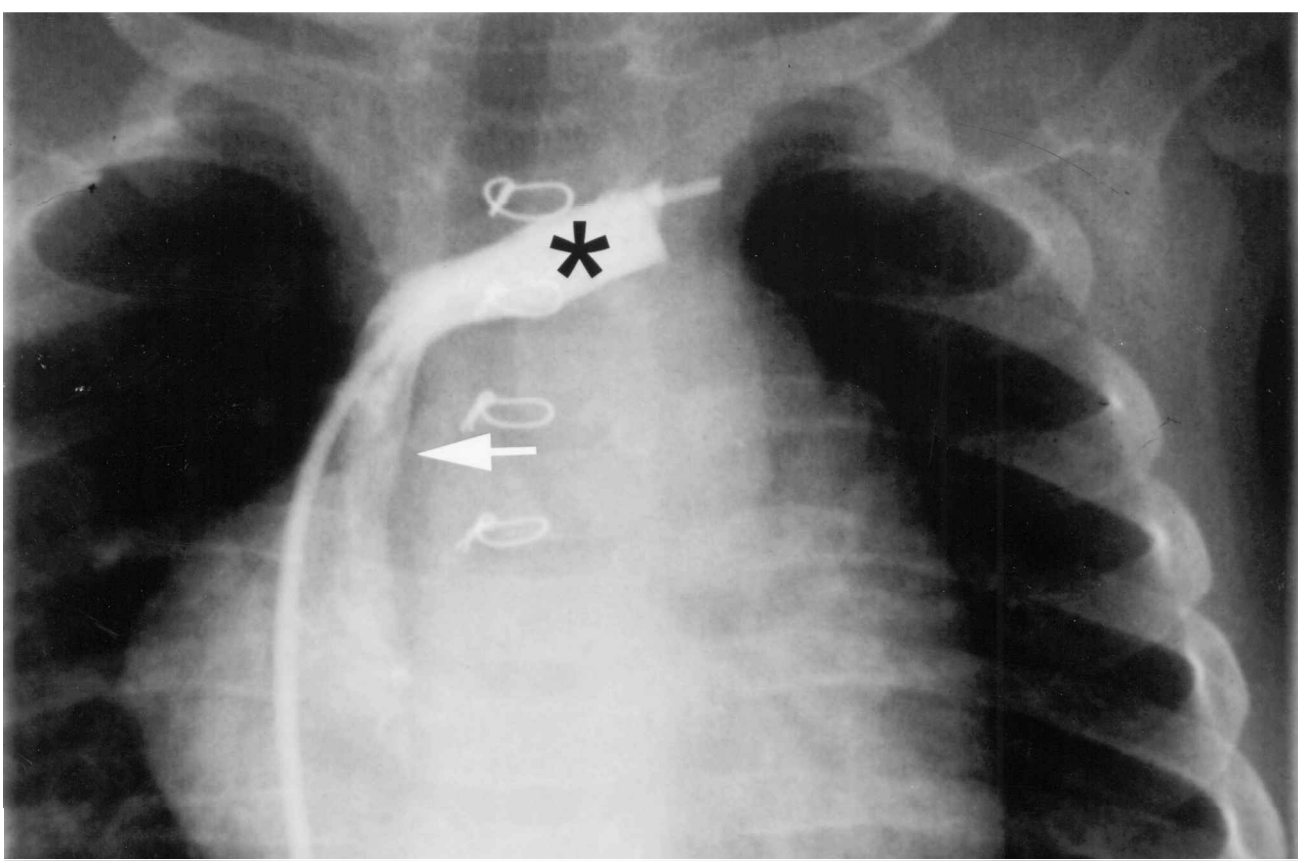

Figure 1.
Power injection within the brachiocephalic vein in a child with hypoplastic left heart syndrome subsequent to the first stage of a Norwood reconstructive procedure, and taken 2 months prior to performing a complete fenestrated Fontan operation without an intermediate hemi-Fontan procedure. There is no evidence of an anomalous venous collateral channel arising from the brachiocephalic vein (*) or the right superior caval vein (white arrow.

https://doi.org/10.1017/S1047951101000312 Published online by Cambridge University Press

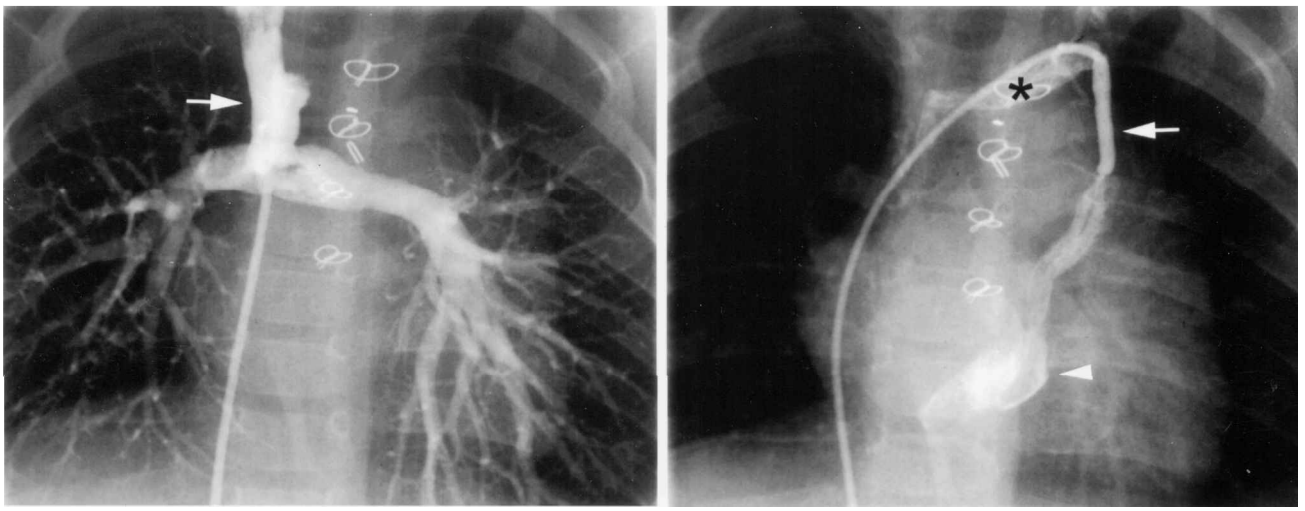

Figure 2.
Routine postoperative cardiac catheterization 13 months subsequent to construction of a fenestrated Fontan procedure in the patient shown in Fig. 1. Left panel: no evidence of a decompressing venous collateral or retrograde filling of the azygos vein, which was intentionally left open, from the right superior caval vein (solid white arrow). Right panel: selective hand injection of contrast in a small decompressing venous collateral (solid white arrow) which extends from the brachiocephalic vein (*) to the coronary sinus. This venous channel was initially identified via a power injection of contrast within the brachiocephalic vein similar to that shown in Fig. 1.

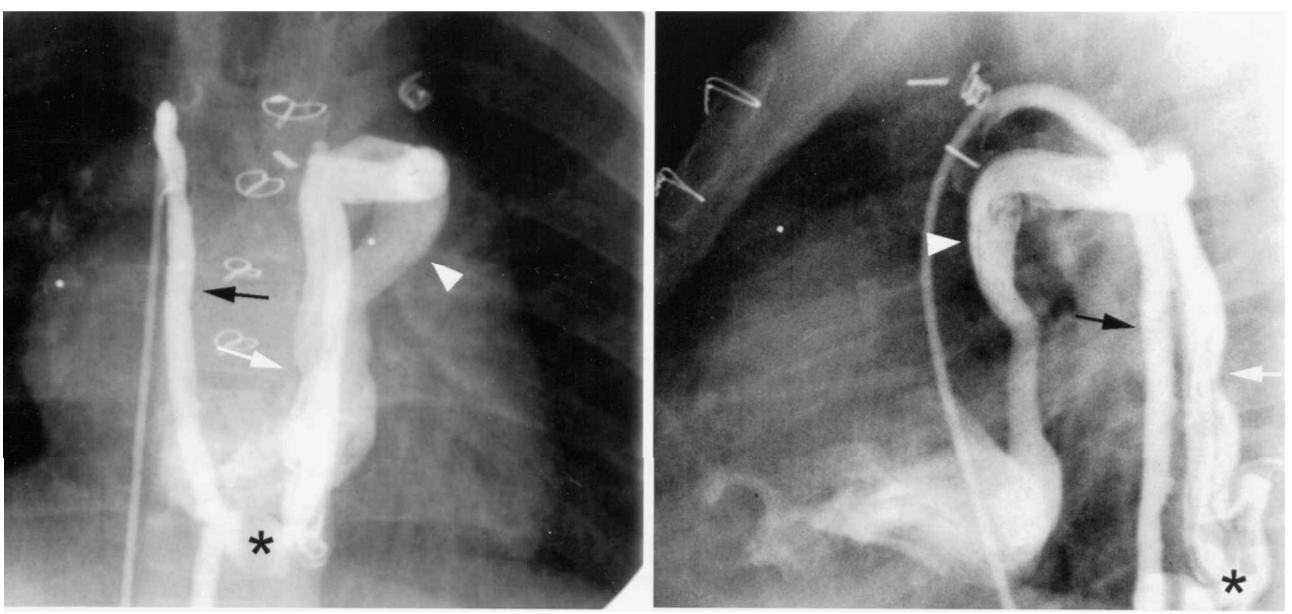

Figure 3.
Repeat cardiac catheterization performed in the same patient 46 months after construction of the fenestrated Fontan procedure because of systemic hypoxemia detected during routine follow-up with an oxygen saturation of 80% by pulse oximetry. A power angiogram within the right superior caval vein demonstrated retrograde filling of the azygos vein and pulmonary venous atrium, following which selective power angiography was performed. Left panel: the azygos vein (solid black arrow) is in communication with a hemiazygos vein (solid white arrow) via a bulbous-like structure (*) with opacification of an enlarged distal vein (solid white arrowhead) and the coronary sinus, which eventually fills the pulmonary venous atrium. This vessel was occluded with several coils (at the level of the asterisk) with an increase in systemic saturation to 90%. The previously placed coil in the proximal portion of the vein demonstrated in the earlier catheterization (Fig. 2) was confirmed to be closed. Right panel: Lateral image of the same angiogram.

https://doi.org/10.1017/S1047951101000312 Published online by Cambridge University Press

thereby reducing effective pulmonary blood flow and systemic oxygen saturation. If the etiology of the cyanosis is unclear, however, then investigation should be undertaken to search for the presence of decompressing venous collateral channels via cardiac catheterization and power angiography within the superior and inferior caval veins.

Although I have demonstrated an incidence of supradiaghragmatic decompressing venous collateral channels of variable size in almost half the patients who had no evidence of such channels prior to establishment of Fontan physiology, only about one-fifth had channels sufficiently large to warrant catheter intervention. For my analysis, I specifically differentiated those patients with significant decompressing venous channels delineated by angiography from all others, since this is the subset of patients at risk for developing lower than expected systemic saturations following construction of a hemi-Fontan circulation, or completion of the Fontan procedure.

A previous report by Magee et al[2] demonstrated an incidence of supradiaghragmatic decompressing venous collateral channels of variable size in approximately one third of patients exclusively following construction of the bidirectional cavopulmonary shunt. They also demonstrated that the presence of a greater transpulmonary gradient at follow-up catheterization was a risk factor for the development of the decompressing channels. Their analysis, however, did not differentiate between patients with small as opposed to those with hemodynamically significant channels, whilst they also included patients who developed reversal of flow within a pre-existing venous collateral that had been evident prior to performing the bidirectional cavopulmonary shunt. The significance of a greater transpulmonary gradient in patients with small decompressing venous collaterals remains unclear, specifically whether, with continued observation, these abnormal vessels will become larger. I did not discover any difference in the length of follow-up between those patients with and without significant decompressing venous channels.

McElhinney et al[3] also demonstrated that a greater transpulmonary gradient was predictive for the development of both small and large decompressing venous collateral channels in 18 of 54 patients following a bidirectional cavopulmonary shunt. This association was evident only in the immediate postoperative period, and not at follow-up catheterization. In addition, two thirds of their patients with collaterals either did not have angiograms performed in either the superior caval vein or brachiocephalic vein prior to construction of the bidirectional cavopulmonary connection, or else they were unavailable for review. Thus, the existence preoperatively of decompressing collateral vessels in some or all of their patients cannot be excluded.

Intuitively, a greater transpulmonary gradient should be a stimulus for the development of a decompressing venous collateral as a way of reducing the elevated pressure in the superior caval venous system subsequent to creation of the Fontan physiology. Although speculative, it is possible that, in my patients, the transpulmonary gradient was even higher immediately following the construction of the bidirectional cavopulmonary connection or completion of the Fontan procedure. As the venous collateral enlarged over time, the transpulmonary gradient could have become smaller. Confirming this hypothesis would require the presence of not only a monitoring catheter in the superior caval vein, but also placement of one within the pulmonary venous atrium. This is not our standard clinical practice.

In conclusion, there is an increased risk of developing significant supradiaphragmatic decompressing venous collateral channels in those patients with a greater transpulmonary gradient following a Fontan-type procedure. This should be suspected whenever abnormally low systemic oxygen saturations are detected in the early or late postoperative period. Transcatheter coil embolization is the treatment of choice for occluding these abnormal venous channels. If successful, the procedure should alleviate the cyanosis and avoid the need for immediate surgical intervention.

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