



Identifying Posterior Venous Collaterals Associated with Inordinate Hypoxemia After the Superior Cavopulmonary Anastomosis

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Abstract: Systemic venous collaterals may cause an inordinate amount of hypoxemia or an unsustainable oxygen requirement after single ventricle palliation with a superior cavopulmonary anastomosis. Anterior venous collaterals are easily identified by injections of contrast in the superior vena cava or innominate vein during heart catheterization (venography), while posterior venous collaterals may be overlooked. This report describes how well posterior systemic venous collaterals were identified by venography and CT angiography in five affected patients after a superior cavopulmonary anastomosis. The medical records and images of patients with evidence of prominent posterior systemic venous collaterals after a superior cavopulmonary anastomosis were retrospectively reviewed. Five patients with prominent posterior systemic venous collaterals were identified between 2008 and 2019. Posterior venous collaterals were identified by venography after initial identification by CT angiography in one patient; by CT angiography when overlooked by venography in three patients; and by repeat venography when overlooked by venography during an initial heart catheterization. Three patients survived without inordinate hypoxemia following closure of posterior venous collaterals. The collateral veins were occluded with access through vessels entering the inferior vena cava in two patients and vessels originating from the innominate vein in one patient. Two patients died without closure of posterior venous collaterals. All patients were treated with pulmonary vasodilators without sufficient improvement to prevent a need for intervention or death. In conclusion, posterior venous collaterals were seen well by CT angiography even when overlooked by venography in patients with inordinate hypoxemia following a superior cavopulmonary anastomosis. CT angiograms may also define the course of collaterals and provide guidance for interventions during heart catheterization.

Keywords: CT Angiography, Heart Catheterization, Hypoxemia, Superior Cavopulmonary Anastomosis, Systemic Venous Collaterals

1. Introduction

Single ventricle palliation with a bidirectional superior cavopulmonary anastomosis (SCPA) is usually performed during infancy to provide a stable and reasonably durable source of pulmonary blood flow. However, some patients develop an excessive amount of hypoxemia for this stage of palliation to succeed. Inordinate hypoxemia may result from several factors that limit the amount of pulmonary blood flow or lower the oxygen saturation of blood returning to the heart from the lung or the inferior vena cava. Systemic venous

collaterals may be present if an inordinate amount of hypoxemia occurs in the absence of pulmonary factors, poor systemic blood flow or relative anemia. Systemic venous collaterals allow blood in upper body veins to circumvent the pulmonary circulation and return to the heart through venous connections with the inferior vena cava or pulmonary veins [1-5]. Large collaterals result in unacceptably low oxygen saturation measurements early after palliation. Collaterals may also enlarge over time, limit the durability of palliation and force early treatment with a Fontan procedure or heart transplantation.

Anterior venous collaterals are easily identified when angiograms are performed in the superior vena cava or innominate vein (venography). However, posterior systemic venous collaterals may be overlooked and may be more difficult to close. Zampeli and associates reported their experience identifying collaterals by venography during heart catheterization [5]. Closure of the imaged collaterals failed to alleviate prohibitive hypoxemia or improve the outcome of palliation with a SCPA [5]. Perhaps, some major posterior venous collaterals were overlooked and not successfully closed. In contrast, McElhinney and associates observed a significant increase in oxygenation when collaterals were occluded in their series [2]. They describe imaging and closing some posterior venous channels in some patients [2]. Previous reports have not considered a role for CT angiography in defining the size and course of systemic venous collaterals in this setting. This report describes how well posterior systemic venous collaterals were identified by venography during heart catheterization and CT angiography in five patients after a SCPA.

2. Methods

The Institutional Review Board of the University of Utah approved this retrospective study. Individual consent for the study was waived. The medical records and images of

patients with evidence of prominent posterior venous collaterals were reviewed. The clinical course of each patient is reported descriptively without a statistical analysis.

3. Results

Five patients were identified with large posterior venous collaterals by venography or CT angiography following a SCPA between 2008 and 2019. All patients experienced periods of inordinately low oxygen saturation measurements or a prolonged need for 50% to 100% oxygen after palliation. Posterior venous collaterals were identified by venography after initial identification by CT angiography in one patient; by CT angiography when overlooked by venography in three patients; and by repeat venography when overlooked by venography during an initial heart catheterization. Three patients survived without inordinate hypoxemia following closure of known posterior venous collaterals. Collateral veins were accessed through venous connections with the innominate vein in 1 patient and venous connections with the inferior vena cava in 2 patients. Two patients died without closure of known posterior venous collaterals. Table 1 lists the diagnoses, previous interventions, ages when the SCPA was performed, postoperative complications, use of pulmonary vasodilators and outcomes for each patient.

Table 1. Demographic information and outcome.

Survival with postoperative occlusion of posterior venous collaterals	
Patient 1	
Year of birth	2008
Diagnoses	Hypoplastic left heart syndrome
Prior interventions	Sano modification of stage I palliation
Age of SCPA	6 months, bilateral SCPA
Postoperative complications	Chylothorax (bilateral), sternal wound infection
Pulmonary vasodilators	Inhaled nitric oxide, amlodipine
Outcome	Hospital discharge on supplemental oxygen 60 days after the SCPA and 8 days after occlusion of posterior venous collaterals Successful palliation with a Fontan procedure
Patient 2	
Year of birth	2015
Diagnoses	Dextrocardia, unbalanced atrioventricular septal defect, double-outlet right ventricle, subvalvar and valvar pulmonary stenosis and I-malposed great arteries
Prior interventions	Bilateral SCPA, Fontan procedure with thrombosis of the Fontan conduit (outside institution)
Age of SCPA	43 months, takedown of the Fontan conduit leaving bilateral SCPA
Postoperative complications	Chylothorax (left), paresis of the right side of the diaphragm, right hemothorax
Pulmonary vasodilators	Inhaled nitric oxide, sildenafil
Outcome	Hospital discharge on supplemental oxygen 102 days after takedown of the Fontan conduit and 88 days after occlusion of posterior venous collaterals.
Patient 3	
Year of birth	2017
Diagnoses	Double-outlet right ventricle, aortic valve atresia and hypoplasia of the aortic arch
Prior interventions	Banding of the right and left branch pulmonary arteries
Age of SCPA	3 months
Postoperative complications	Chylothorax (bilateral), sinus node dysfunction requiring placement of an atrial and ventricular epicardial pacing system
Pulmonary vasodilators	Inhaled nitric oxide, inhaled iloprost, sildenafil, bosentan
Outcome	Hospital discharge on supplemental oxygen 45 days after the SCPA and 41 days after creation of connection between the right carotid artery and the right innominate vein Closure of posterior venous collaterals 95 days after the SCPA

Death without postoperative occlusion of posterior venous collaterals

Patient 4

Year of birth	2017
Diagnoses	Hypoplastic left heart syndrome
Prior interventions	Sano modification of stage I palliation
Age of SCPA	3 months
Postoperative complications	Chylothorax (bilateral)
Pulmonary vasodilators	Inhaled nitric oxide, sildenafil
Outcome	Death 49 days after the SCPA

Patient 5

Year of birth	2018
Diagnoses	Pulmonary atresia with intact ventricular septum
Prior interventions	Pulmonary valve perforation and placement of a modified Blalock-Taussig shunt
Age of SCPA	6 months
Postoperative complications	Chylothorax (right), sternal wound infection
Pulmonary vasodilators	Inhaled nitric oxide, sildenafil
Outcome	Hospital discharge on supplemental oxygen 61 days after the SCPA and death related to severe hypoxemia 237 days following the SCPA

SCPA: superior cavopulmonary anastomosis (anastomoses)

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3.1. Patient 1



Figure 1. Access to posterior venous collaterals in Patient 1.

Biplane images of the tortuous course of the catheter (white arrows) passing through the inferior vena cava, right-sided paravertebral veins near the level of the renal veins, and paravertebral veins below and above the diaphragm. A. (LAO 90°, Cranial 1°) and B. (RAO 0°, Cranial 1°).

Oxygen saturation measurements were marginal for a prolonged period despite treatment with supplemental oxygen. A large azygous vein and paravertebral vessels were identified when a CT angiogram was performed to evaluate for a cause of prolonged fever 46 days after the SCPA. Heart catheterization and venography were performed 52 days after the SCPA. An injection of contrast was performed with transient balloon occlusion of the superior vena cava. Contrast filled a plexus of multiple small veins flowing into a large azygous vein. The collaterals could not be accessed through these small anterior veins. The inferior vena cava and renal veins were probed with a catheter and floppy guide wire. As shown in Figure 1, a tortuous venous pathway was entered allowing a guide wire and catheter to be advanced to the azygous vein and the hemiazygous vein or a left-sided paravertebral vein. Multiple coils were deployed into these right- and left-sided venous collaterals. She was also noted to

have angiographic evidence of prominent systemic to pulmonary arterial collaterals. The right internal mammary artery was occluded with multiple coils. The patient was sent home on 0.75 liters per minute nasal cannula oxygen 60 days after the SCPA and 8 days after occlusion of systemic venous collaterals.

3.2. Patient 2

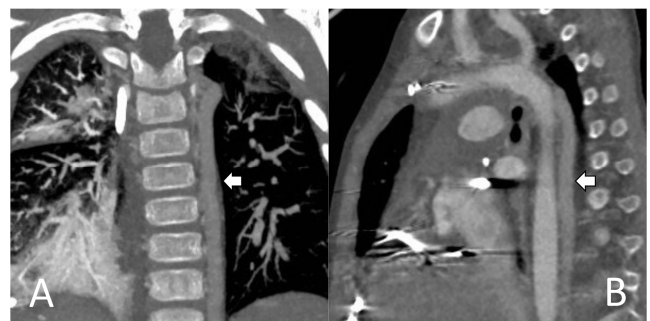


Figure 2. Left posterior venous collateral in Patient 2.

CT angiography of a prominent hemiazygous or left paravertebral vein (white arrows) posterior to the thoracic descending aorta. A. Coronal image. B. Sagittal image.

The patient developed thrombosis of a complex conduit from the inferior vena cava and hepatic veins to the pulmonary arteries after a Fontan procedure at an outside hospital. The patient was transported to our hospital and underwent emergent takedown of the conduit, leaving bilateral SCPA. Oxygen saturation measurements were often less than 75% despite treatment with 75% to 100% oxygen. Heart catheterization was performed 8 days after surgical conversion to bilateral SCPA. Injections of contrast in the right superior vena cava and left superior vena cava failed to identify any prominent systemic venous collaterals. Though small anterior venous

collaterals originating from the left superior vena cava were occluded with coils. As shown in Figure 2, a large hemiazygous or left-sided paravertebral vein was identified when a CT angiogram was performed 14 days after surgical conversion. The inferior vena cava and renal veins were probed with a catheter and floppy guide wire during repeat heart catheterization 16 days after surgical conversion. A tortuous venous pathway was entered allowing a guide wire and catheter to be advanced to the left-sided paravertebral vein. This vessel and additional contiguous right-sided paravertebral veins were occluded with vascular plugs. His oxygen requirement readily improved. However, he required a long period to recover from postoperative complications. He was sent home on 1 liter per minute nasal cannula oxygen during the day and 3.5 liters per minute oxygen with bilevel positive airway pressure during the night 102 days after surgical conversion to bilateral SCPA and 88 days after occlusion of the systemic venous collaterals.

3.3. Patient 3

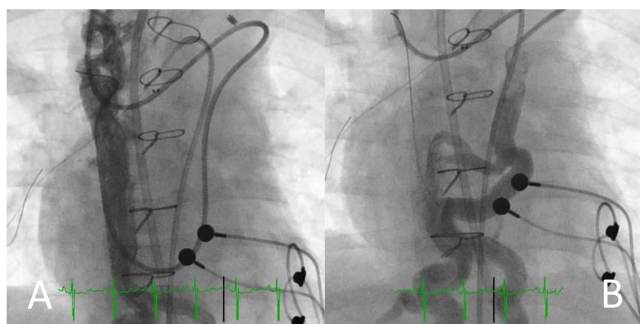


Figure 3. Posterior venous collaterals in Patient 3.

A. Image of a large azygous or right paravertebral vein (RAO 0°, Caudal 1°). B. Image of a large hemiazygous or left paravertebral vein (RAO 0°, Caudal 1°). The right and left collateral veins connect through enlarged vessels of the anterior external vertebral venous plexus.

Oxygen saturation measurements were often less than 75% despite treatment with 75% to 100% oxygen. Heart catheterization was performed 3 days after the SCPA. Injections of contrast in the superior vena cava, without and with balloon occlusion of the lower superior vena cava, failed to identify any prominent systemic venous collaterals. Four days after the SCPA, the patient was transiently placed on support with extracorporeal membrane oxygenation and an arteriovenous connection was made between the right carotid artery and right innominate vein. Over time, the patient developed adequate oxygen saturation measurements and was sent home on 0.25 liters per minute nasal cannula oxygen 45 days after the SCPA. Due to concerns of a large volume load associated with the arteriovenous connection, heart catheterization was repeated 95 days after the SCPA to re-evaluate for potential posterior venous collaterals. An injection of contrast in the superior vena cava failed to identify systemic venous collaterals. Only after probing the innominate vein and performing selective manual injections

of contrast in small contiguous veins were massive left and right posterior venous collaterals identified, as shown in Figure 3. The collateral veins were occluded with vascular plugs. The arteriovenous connection was also occluded with a vascular plug with no need for additional supplemental oxygen. The size of the posterior venous collaterals early after the SCPA is unknown because CT angiography was not performed.

3.4. Patient 4

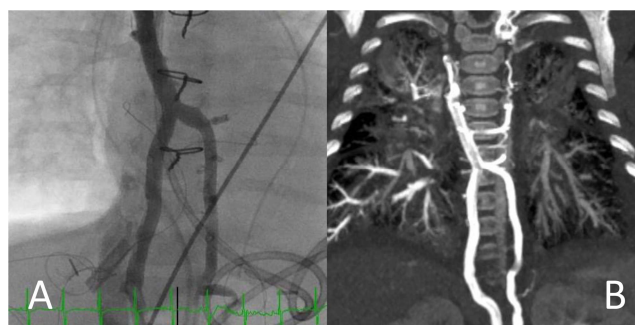


Figure 4. Posterior venous collaterals in Patient 4.

A. The azygous vein and inferior paravertebral veins were imaged by angiography before the azygous vein was occluded with a vascular plug near its connection with the superior vena cava. B. Coronal image of persistent prominent azygous and inferior right and left paravertebral veins that were identified by CT angiography after the azygous vein was occluded near its connection with the superior vena cava.

The patient developed thrombosis of the iliac veins and inferior portion of the inferior vena cava after stage I palliation. The azygous vein was not ligated at the time of the SCPA. Oxygen saturation measurements were often less than 75% despite treatment with 100% oxygen and frequent hemoglobin values near 15 g/dl. Heart catheterization and angiography were performed 3 days after the SCPA. Injections of contrast were performed in the superior vena cava and azygous vein. Large azygous and right paravertebral veins were allowing blood to flow inferiorly to the left renal vein. A single vascular plug was placed in the azygous vein near its connection with the superior vena cava. The severity of hypoxemia and the oxygen requirement only improved a small amount over time. Heart catheterization was repeated 36 days after the SCPA. No systemic venous collaterals were identified with an injection of contrast in the superior vena cava; however, he now had evidence of prominent systemic to pulmonary arterial collaterals. As shown in Figure 4, persistently large azygous and paravertebral venous collaterals were identified when a CT angiogram was performed 39 days after the SCPA. The collateral veins could no longer be entered through the occluded upper azygous vein, or through an inferior left renal venous approach due to prior thrombosis of the iliac veins and lower portion of the inferior vena cava. He was ultimately withdrawn from assisted ventilation and died 49 days after the SCPA.

3.5. Patient 5

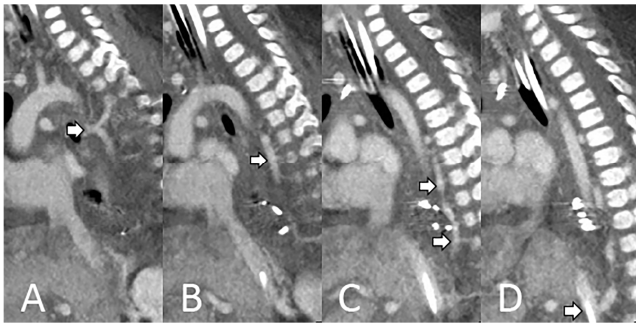


Figure 5. Posterior venous collaterals in Patient 5.

A series of rightward to leftward sagittal images are shown with arrows delineating a prominent collateral vein. A. A superior posterior paravertebral vein joins with the ligated azygous vein. B. and C. The azygous vein courses inferiorly and leftward. D. The azygous vein joins directly with the inferior vena cava below the level of the diaphragm.

Oxygen saturation measurements were initially acceptable with supplemental oxygen. Heart catheterization and angiography were performed 13 days after the SCPA due to a persistent chylothorax. No systemic venous collaterals were identified with an injection of contrast in the superior vena cava. His oxygen requirement increased when thoracic duct ligation was performed through a right thoracotomy 18 days after the SCPA. He subsequently required 50% to 100% oxygen to maintain oxygen saturation measurements near 80% for several weeks. CT angiography was performed 32 days after the SCPA and identified prominent right and left paravertebral veins with an inferior course and a direct connection with the inferior vena cava below the level of the diaphragm, as shown in Figure 5. Heart catheterization was repeated 56 days after the SCPA. Again, no prominent systemic venous collaterals were identified with injections of contrast in the superior vena cava and the innominate vein. There was angiographic evidence of thrombosis of the right iliac vein and a catheter was present in the left femoral vein. Due to the presence of prominent systemic to pulmonary arterial collaterals, providers hoped the severity of hypoxemia and the oxygen requirement would gradually improve over time. The left femoral venous catheter was not exchanged to establish vascular access and his collateral veins were not closed through an inferior vena cava approach. He was sent home 61 days after the SCPA. He was readmitted to the hospital for management of a small right pleural effusion, dehydration and an increased oxygen requirement approximately 6 months later. He developed severe hypoxemia and terminal bradycardia when nasal secretions were being suctioned 237 days after the SCPA.

Care providers at Primary Children's Hospital frequently use inhaled nitric oxide and sildenafil empirically in patients with low oxygen saturation measurements following the SCPA. There was no evidence that pulmonary vasodilators had a compelling impact on the postoperative course of patients in this report. Supplemental oxygen is also

frequently used for two reasons. First, supplemental oxygen sometimes helps to maintain oxygen saturation measurements near 80%. Second, an increase in alveolar oxygen often helps to decrease pulmonary vascular resistance. Care providers typically administered transfusions of red blood cells to avoid anemia; however, they did not uniformly perform transfusions to achieve a minimum hemoglobin or hematocrit in these patients. Though systemic oxygenation could have potentially been improved in patients by increasing hemoglobin values, and in turn increasing the oxygen saturation of blood returning to the heart from the inferior vena cava.

4. Discussion

This report illustrates that posterior systemic venous collaterals can be identified well by CT angiography. The patients in this report who underwent closure of posterior collateral veins following the SCPA survived, while the patients who did not undergo closure of similar collaterals ultimately died.

The focus of this report is using CT angiography to accurately image posterior venous collaterals. One cannot infer from a small number of patients that closure of venous collaterals will guarantee early or long-term survival. However, some patients with inordinate hypoxemia may ultimately be treated with extracorporeal membrane oxygenation when collaterals are not successfully identified and closed. Greenberg and associates reported an increased risk for poor outcome when extracorporeal support is needed [6]. Further, Kendall and associates have reported that length of hospitalization after the SCPA is a risk factor for attrition before the total cavopulmonary anastomosis [7]. Early identification and successful closure of collateral veins might decrease the length of hospitalization, and thereby have an impact on long-term outcome for some patients.

Posterior venous collaterals typically include the azygous vein, hemiazygous vein and other paravertebral veins. Right and left collaterals may communicate at several levels through the anterior external vertebral venous plexus. Posterior venous collaterals may be overlooked when injections of contrast are performed in the superior vena cava or innominate vein. CT angiography may be a superior method for identifying posterior collateral veins. Imaging should be performed several seconds following the injection of contrast in order to allow contrast to enter and image the posterior vessels. CT angiography can also be used to plan interventions to occlude collaterals. Imaging should be extended from the neck to the level of the renal veins in order to identify how posterior collateral channels ultimately join with the inferior vena cava or pulmonary veins. The collaterals often join with the renal veins or nearby vessels; however, direct connections with the inferior vena cava may occur.

Posterior venous collaterals can be entered and occluded with coils or vascular plugs. However, entering and advancing guide wires and catheters through the tortuous

venous connections to the culpable paravertebral veins can be challenging. Vessels near the vertebral foramina and spinal nerves should not be occluded in order to avoid potential nerve injury. Most of the paravertebral collateral veins that are large enough to cause inordinately low oxygen saturation measurements after the SCPA are more anterior, and often include branches of the azygous and hemizygous veins. Day and associates have previously accessed and occluded posterior venous collaterals in this manner to increase the duration of palliation with a SCPA [8]. However, this report is the first to present images describing successful closure of collaterals with access through the inferior vena cava.

The azygous vein is surgically ligated when the SCPA is performed. Flow through the pulmonary arteries is usually sufficient for most patients to have oxygen saturation measurements near 80% with little or no supplemental oxygen. The anatomic and hemodynamic reasons why posterior venous collaterals develop, and are sufficiently large to impair systemic oxygenation in a subset of patients early after surgery, are unknown. However, the amount of the collective shunt might depend upon the size of collateral connections with the inferior vena cava or pulmonary veins. Patient 4 had persistent enlargement of posterior venous collaterals when only the upper azygous vein was occluded. Many upper body veins were allowed to flow into the azygous vein along the course of the thoracic and upper abdominal spine. In this patient, an opportunity to improve oxygenation was lost by not occluding the upper and lower segments of the major collateral channels. The azygous and hemiazygous systems could be routinely occluded with coils or vascular plugs prior to the SCPA. However, a preemptive intervention might only be necessary or worthwhile for a subset of patients.

All of the patients were treated with inhaled and enteral pulmonary vasodilators, even though Adatia and associates previously reported that inhaled nitric oxide does not improve oxygenation after the SCPA [9]. Frank and associates reported that circulating endothelin-1 levels are associated with inordinate hypoxemia following the SCPA [10]. However, Pearl and associates noted that endothelin-1 levels often increase when inhaled nitric oxide is used postoperatively [11]. Only Patient 3 was treated with an endothelin receptor antagonist in this series.

All of the patients in this report developed bilateral or unilateral chylous pleural effusions. Patients with a unilateral chylothorax had more prominent posterior venous collaterals on the ipsilateral side. More investigation is needed to determine whether the pressure or direction of flow in the collateral veins are factors that influence lymphatic function and contribute to the development of chylous effusions.

All of the patients with posterior venous collaterals and no source of pulmonary blood flow other than the SCPA developed prominent systemic to pulmonary arterial collaterals over several weeks. Arterial collaterals may improve oxygenation, but increase the volume load on the heart. More investigation is needed to determine whether posterior collateral veins provide a low resistance path that

fosters the progressive enlargement of systemic arterial to pulmonary arterial connections and the progressive enlargement of the posterior venous collaterals.

5. Conclusion

In conclusion, CT angiography may identify posterior venous collaterals and provide guidance for their access and closure. Systemic oxygen saturation and requirements for supplemental oxygen can be improved by identifying and closing posterior venous collaterals after the SCPA, particularly when other causes of hypoxemia have already been addressed.

Conflict of Interest Disclosure

The author declares that he has no competing interests.

Author Contributions

Ronald William Day is the sole author. He compiled the demographic information and prepared the images of all patients for publication.

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