CASE STUDY

Reopening of persistent left superior caval vein after bidirectional cavopulmonary connections

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Abstract

Persistent left superior vena cava (SVC) is a not uncommon finding in patients with congenital heart disease. This anatomical variant must be recognised before doing a Glenn anastomosis, bidirectional cavopulmonary connection or a Fontan-type procedure. Following these procedures, reopening of a left SVC leading to clinical cyanosis can occur. Five cases are described in whom persisting left SVCs were excluded before performing a bidirectional cavopulmonary connection or Fontan procedure but (re-)opened after surgery, leading to cyanosis either by reducing effective pulmonary blood flow (bidirectional cavopulmonary connection) or by an obligatory right to left shunt (Fontan). These observations suggest that, embryologically, the lumen of the left SVC obliterates rather than disappears. Balloon occlusion angiography of the innominate vein before cavopulmonary connections or Fontan procedures might improve detection of these collateral vessels.

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Keywords: persistent left superior vena cava; cavopulmonary connection; Fontan procedure; congenital heart disease

Clinical deterioration following the establishment of cavopulmonary connections can result from the appearance of venous collaterals bypassing the lung and thus compromising the effective pulmonary blood flow.¹⁻³ Various types of systemic venous collaterals thought to result from altered venous haemodynamics have been described.⁴⁻⁵ However, reopening of a persistent left superior vena cava (SVC) as a cause for such clinical deterioration is uncommon. We describe five patients (table 1) in whom a persistent left SVC was excluded at initial assessment before cavopulmonary surgery, but angiography performed after establishment of the cavopulmonary shunt or Fontan-type procedure showed the presence and thus reopening of a left SVC draining to the coronary sinus. Embryological development of the anterior cardinal venous system and correlation with current findings are discussed.

Case reports

PATIENT 1

Patient 1 was diagnosed with situs solitus, dominant left form of atrioventricular septal defect, hypoplastic right ventricle, ventriculo-arterial (VA) concordance, and persistent arterial duct. A left SVC was excluded at initial assessment by a hand injection in the innominate vein. Pulmonary artery banding with ligation of the arterial duct was performed at the age of 4 months, followed at 4 years by a right sided classic Glenn anastomosis, and at 8 years by a left modified Blalock-Taussig shunt. At age 16, repeat cardiac catheterisation was performed in view of increasing cyanosis. Injection in the innominate vein demonstrated a small left SVC, draining to the coronary sinus, as well as other venovenous collaterals. The ostium of the coronary sinus was felt to be slightly stenotic. No transcatheter intervention was performed.

PATIENT 2

Patient 2 was diagnosed with situs solitus, double inlet left ventricle, hypoplastic right ventricle, restrictive ventricular septal defect, VA discordance, subvalvar and valvar pulmonary stenosis. Echocardiography excluded a persistent left SVC.⁶ A bidirectional cavopulmonary connection (BDCPC) with main pulmonary artery ligation, atrial septectomy, ventricular septal defect enlargement, and patch enlargement of the subaortic region was performed at the age of 6 months. One year later, a lateral tunnel Fontan procedure with fenestration was performed. Over the next months, there was clinical evidence of raised pressures in the venous circuit. Cardiac catheterisation one year after the Fontan procedure showed high venous pressures caused by diastolic dysfunction. Despite alleviation of symptoms with afterload reduction, the child remained cyanotic. Repeat cardiac catheterisation showed low venous pressures, lateral tunnel patch detachment, and a left SVC draining to the coronary sinus. The ostium of the coronary sinus was mildly stenotic. Transoesophageal echocardiography showed ascending flow in the left SVC, suggestive of obstruction at the coronary sinus ostium. No transcatheter intervention was performed.

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Table 1 Patient characteristics

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Diagnosis	Unbalanced AVSD	Double inlet LV	Tricuspid atresia	Double inlet LV	Ventriculoarterial discordance with hypoplastic LV
Exclusion of left SVC before BDCPC	Hand injection in innominate	Echocardiography	Hand injection in innominate	Hand injection in innominate	Power injection in innominate
Time between surgery and post-op catheterisation	8 years	1 year	10 months	14 months	8 days
Clinical symptoms	Increasing cyanosis	Increasing cyanosis	None	None	Increasing cyanosis
Intervention	None	None	Coil occlusion	Surgical ligation	Surgical ligation

AVSD, atrioventricular septal defect; BDCPC, bidirectional cavopulmonary connection.

PATIENT 3

Patient 3 was diagnosed with situs solitus, tricuspid atresia, hypoplastic right ventricle, ventricular septal defect, and normally related great arteries. Initial angiograms excluded a persistent left SVC. A modified right Blalock-Taussig shunt was performed at the age of 8 months, followed by a BDCPC at 18 months. At the age of 4 years a modified fenestrated extracardiac Fontan procedure was performed. The main pulmonary artery was ligated but not divided. Cardiac catheterisation 10 months later showed a low pressure unobstructed venous circuit, a patent fenestration, small epicardial veins connecting the hepatic veins with the left atrium, and a reopened left SVC to coronary sinus. There was also persistent flow across the ligated main pulmonary artery. The left SVC and collateral veins were successfully occluded in the catheterisation laboratory. The persistent main pulmonary artery shunt will be addressed in the near future.

PATIENT 4

Patient 4 was diagnosed with situs solitus, double inlet left ventricle, hypoplastic right ventricle, VA discordance, pulmonary atresia, and right aortic arch. A modified left Blalock-Taussig shunt was done in the neonatal period for initial palliation. At the age of 10 months the Blalock-Taussig shunt was taken down and a right sided BDCPC with right pulmonary artery plasty was performed. Cardiac catheterisation before the BDCPC failed to show a persistent left SVC (fig 1). At 2 years of age repeat

cardiac catheterisation as a pre-Fontan assessment showed unequivocally a left SVC draining to the coronary sinus with moderate stenosis of the left SVC to coronary sinus junction, as well as small venous collaterals from the left SVC to the coronary veins (fig 2). The child was scheduled for Fontan procedure with left SVC ligation.

PATIENT 5

Patient 5 was diagnosed with situs solitus, atrioventricular (AV) concordance, hypoplastic left atrioventricular valve, straddling right atrioventricular valve, large inlet muscular ventricular septal defect, hypoplastic left ventricle, VA discordance, valvar and sub-valvar pulmonary stenosis. At 3 days old he underwent a balloon atrial septostomy followed 4 months later by a right sided BDCPC with atrial septectomy and azygos vein ligation. The pulmonary artery was not ligated at that stage. Selective angiography in the innominate vein before the BDCPC had shown a minute vein not clearly proven to be a left SVC, but possibly a left superior intercostal vein (fig 3). Echocardiography before Fontan procedure excluded a left SVC. Eight days after undergoing a fenestrated Fontan operation with additional MPA division the patient was progressively cyanotic. Angiography showed a widely patent left SVC draining to the coronary sinus with areas of stenosis throughout its course (fig 4). The child subsequently underwent division and ligation of the reopened left SVC.

Figure 1 Patient 4 before bidirectional cavopulmonary connection.

Figure 2 Patient 4 after bidirectional cavopulmonary connection. Reopened left SVC is arrowed. RPA, right pulmonary artery; LPA, left pulmonary artery.

Figure 3 Patient 5 before bidirectional cavopulmonary connection. Arrow points to left superior intercostal vein.

Figure 4 Patient 5 after bidirectional cavopulmonary connection. Star is at coronary sinus; left SVC (LSVC) is widely patent with multiple stenosis throughout its course.

Discussion

During embryological life, the bilateral anterior cardinal venous system, consisting of the left and right common cardinal veins, develops into a unilateral right sided system in cases with solitus arrangement. This is believed to result from obliteration of the left cardinal vein, leaving the coronary sinus as a remnant. This process is not well understood but is speculated by some to result from intrauterine compression of the left sided system between the left atrium and the hilum of the left lung, with subsequent luminal obliteration.⁷⁻⁸ In 0.1–0.4% of the general population a left SVC persists.⁷⁻⁹ This could be secondary to reduced compression by the left atrium and flow redistribution at an early stage in embryonic development.⁷ This correlates well with the fact that persistent left SVC is more frequent in patients with congenital cardiac abnormalities, where it is as high as 10%.⁷⁻¹⁰⁻¹¹ In particular, congenital heart defects with undetermined atrial situs are highly correlated with left SVC with an incidence up to 72%.¹²

In most cases the left SVC connects to the right atrium via the coronary sinus,³⁻¹³⁻¹⁴

however, direct drainage into the left atrium with absent or unroofed coronary sinus or normal coronary sinus has been described.¹³⁻¹⁶ Persistent left SVC with stenosis or atresia of the coronary sinus ostium are other variants.¹⁶⁻¹⁹ Buirska *et al* suggested an angiographic classification of persistent left SVC in five groups ranging from bilateral superior vena cava without interconnecting veins to rudimentary persistent left SVC.¹²

Detailed study of the systemic venous anatomy is mandatory before performing cavopulmonary connections, to exclude pre-existing anatomical variants of the venous system such as a persistent left SVC, as they may require different surgical approaches.¹⁴⁻¹⁵ Increasing cyanosis after classic Glenn anastomosis and BDCPC has been well described, as a result of the development of abnormal venous channels, which reduce effective pulmonary blood flow. These abnormal vessels, connecting the superior with the inferior vena cava, atrium or pulmonary veins, could be angiogenesis *de novo* or result from (re-)opening of pre-existing channels.¹⁻³⁻⁵⁻¹⁴⁻²⁰⁻²¹ Altered haemodynamics following the establishment of cavopulmonary connections with increased pressure gradient between the pulmonary and systemic venous circuit may promote their development.²⁻³⁻²² Magee *et al* diagnosed a persistent left SVC at preoperative catheterisation in 18 of 103 patients who underwent a BDCPC and for whom preoperative and postoperative angiograms were available over a 13 year period (1983–95).³ At surgery, the left SVC was either ligated or connected to the pulmonary artery (bilateral BDCPC). Venous collaterals were present in 32 of the 103 patients on postoperative catheterisation (median time of postsurgical catheterisation 1.3 years, range 0.3–14.3 years), one of them being a (re-)opened left SVC.³ Surgical or more recently transcatheter closure of these collaterals can improve the patient's condition.⁵⁻²⁰

Our study reports five patients in whom the absence of a patent left SVC was demonstrated before construction of a cavopulmonary connection. Following classic Glenn anastomosis, bidirectional cavopulmonary shunt or Fontan-type procedure, all reopened an left SVC, resulting in venovenous right to left shunt. Similarly, Gatzoulis *et al* described a patient with left atrial isomerism who reopened a left sided inferior vena cava not detectable on preoperative screening.⁴ These findings support the concept that primitive venous structures obliterate but do not disappear during embryological life. Angiography performed with balloon occlusion or high pressure might improve detection of pre-existing vessels susceptible to reopening. After surgery, serial echocardiographic controls specifically aimed to detect reopened left SVC can be performed, especially in patients with clinically progressive cyanosis, as this modality has been shown to detect this anatomical variant reliably.⁶⁻²³ In two of our patients the left SVC was not occluded during the cardiac catheterisation or by surgery as the coronary sinus was thought to be stenotic. Although it remains difficult to

prove this in our cases, this emphasises the need to visualise the coronary ostium in an adequate way before occluding the left SVC.

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- 1 Barger LM, Karp TB, Barcia A, et al. Late deterioration of patients after superior vena cava to right pulmonary anastomosis. *Am J Cardiol* 1972;30:211–16.
- 2 Laks H, Mudd GJ, Standeven JW, et al. Longterm effect of the superior vena cava-pulmonary artery anastomosis on pulmonary blood flow. *J Thorac Cardiovasc Surg* 1980;74:253–60.
- 3 Magee AG, McCrindle BW, Mawson J, et al. Systemic venous collaterals after the bidirectional cavopulmonary shunt: prevalence and risk factors [abstract]. XXXI annual meeting of the Association of European Pediatric Cardiologists, Göteborg, Sweden, 1996.
- 4 Gatzoulis MA, Shinebourne EA, Redington AN, et al. Increasing cyanosis early after cavopulmonary connection caused by abnormal systemic venous channels. *Br Heart J* 1995;73:182–6.
- 5 Stumper O, Wright JGC, Sadiq M, et al. Late systemic desaturation after total cavopulmonary shunt operations. *Br Heart J* 1995;74:282–6.
- 6 Hutha JC, Smallhorn JF, Macartney FJ, et al. Cross-sectional echocardiographic diagnosis of systemic venous return. *Br Heart J* 1982;48:388–403.
- 7 Nsah EN, Moore GW, Hutchins GM. Pathogenesis of persistent left superior vena cava with a coronary sinus connection. *Pediatric Pathology* 1991;11:261–9.
- 8 Raghib G, Ruttenberg HD, Anderson RC, et al. Termination of left superior vena cava in left atrium, atrial septal defect, and absence of coronary sinus. *Circulation* 1965;xxxi:906–18.
- 9 Page Y, Tardy B, Comtet C, et al. Catheterisme veineux et anomalies congenitales de la veine cave superieure. *Annales Francaises d'Anesthesie et de Reanimation* 1990;9:450–5.
- 10 Campbell M, Deuchar DC. The left-sided superior vena cava. *Br Heart J* 1954;16:423–39.
- 11 Parikh SR, Prasad K, Iyer RN, et al. Prospective angiographic study of the abnormalities of systemic venous connections in congenital and acquired heart disease. *Cathet Cardiovasc Diagn* 1996;38:379–86.
- 12 Buirski G, Jordan SC, Joffe HS, et al. Superior vena caval abnormalities: their occurrence rate, associated cardiac abnormalities and angiographic classification in a paediatric population with congenital heart disease. *Clin Radiol* 1986;37:131–8.
- 13 Wiles HB. Two cases of left superior vena cava draining directly to a left atrium with a normal coronary sinus. *Br Heart J* 1991;65:158–60.
- 14 de Leval M, Ritter DG, McGoon DC, et al. Anomalous systemic venous connection. Surgical considerations. *Mayo Clin Proc* 1975;50:599–610.
- 15 Mazzucco A, Bortolotti U, Stellin G, et al. Anomalies of the systemic venous return: a review. *J Cardiovasc Surg* 1990;5:122–33.
- 16 Adatia I, Gittenberger-de Groot AC. Unroofed coronary sinus and coronary sinus orifice atresia implications for management of complex congenital heart disease. *J Am Coll Cardiol* 1995;25:948–53.
- 17 Santoscoy R, Walters GK, Ross RD, et al. Coronary sinus ostial atresia with persistent left superior vena cava. *Ann Thorac Surg* 1996;61:879–82.
- 18 Chiang CE, Chen SA, Yang CR, et al. Major coronary sinus abnormalities: identification of occurrence and significance in radiofrequency ablation of supraventricular tachycardia. *Am Heart J* 1994;127:1279–89.
- 19 Gerlis LM, Gibbs JL, Williams GJ, et al. Coronary sinus orifice atresia and persistent left superior vena cava. *Br Heart J* 1984;52:648–53.
- 20 Bridges ND, Jonas RA, Mayer JE, et al. Bidirectional cavo-pulmonary anastomosis as interim palliation for high-risk Fontan candidates: early results. *Circulation* 1990;82:170–6.
- 21 McElhinney DB, Reddy VM, Hanley FL, et al. Systemic venous collateral channels causing desaturation after bidirectional cavopulmonary anastomosis. *J Am Coll Cardiol* 1997;30:817–24.
- 22 Pennington DG, Nouri S, Ho J, et al. Glenn shunt: long-term results and current role in congenital heart operations. *Ann Thorac Surg* 1980;31:532–9.
- 23 Sunaga Y, Okubo N, Hayashi K, et al. Transesophageal echocardiographic diagnosis of coronary sinus orifice atresia. *Am Heart J* 1992;124:794–6.