Cardiol Young 2002; 12: 218–223

ISSN 1047-9511

Review

Physiological impact and transcatheter treatment of the persisting left superior caval vein

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Abstract A left superior caval vein frequently occurs in the malformed, as well as in the structurally normal, heart. Its physiological impact varies, and is determined by its connections and whether there are associated cardiac lesions. In this review, we describe 3 patients with such a vein, 1 without other lesions and 2 with the anomalous venous channel as a component of a complex cardiac malformation. In all cases, transcatheter techniques were used to treat the physiological dysfunction caused by the presence of the vein. The connections of the vein, and the complexity of the associated cardiac lesions, determine the options for treatment. We define the role of surgical as opposed to transcatheter intervention, and discuss strategies for transcatheter treatment.

Keywords: Systemic veins; superior vena cava; coronary sinus; interventional cardiology

A LEFT SUPERIOR CAVAL VEIN FREQUENTLY occurs in association with complex cardiac lesions, as well as in the structurally normal heart.[1] As the connection of the vein is variable with the other systemic venous tributaries, its physiological effects can vary. It may cause positional or persistent systemic desaturation, or paradoxical embolism due to an obligatory right-to-left shunt.[2] Alternatively when seen in association with obstructive lesions of the left heart, the shunting may be left-to-right.[3,4] Such shunts can be masked by associated cardiac lesions, and may manifest only after surgical repair. Presentation can be with inappropriate systemic desaturation during or after staged functionally univentricular palliation. A left superior caval vein has also been implicated in the causation of obstruction to left ventricular inflow. When connected appropriately to the right heart, its presence may be inconsequential.[2–15]

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Accepted for publication 11 January 2002

Transcatheter[9,16–18] and surgical intervention are well established for treatment of the vein[6,19–25]. In this review, we illustrate the options in the setting of 3 cases, 1 in the absence of, and 2 in association with, complex cardiac lesions. In all these cases, transcatheter intervention served to correct the physiological dysfunction. We reviewed the physiologic impact of the persisting left superior caval vein, and discuss the options for transcatheter treatment.

Illustrative cases

Our first patient, a 6-year-old boy came to clinical attention due to mild systemic desaturation with oxygen saturation of 89% in room air upon presentation to the emergency room for suspected aspiration of a foreign body. Clinical examination, chest radiography and electrocardiography were all normal. Analysis of arterial blood gases revealed a saturation of oxygen 60 mmHg when breathing 100% oxygen through a facemask. Echocardiography suggested a persisting left superior caval vein connecting to the left atrium in the setting of usual atrial arrangements and concordant segmental connections. Both atrial and ventricular septums were intact. At cardiac catheterization, angiography confirmed bilateral superior caval veins,

https://doi.org/10.1017/S1047951102000495 Published online by Cambridge University Press

with a bridging brachiocephalic vein and azygous and hemiazygous veins connecting to the right and left superior caval veins, respectively. There was mild stenosis of the insertion of the brachiocephalic vein to the right superior caval vein. The left superior caval vein was capacious, measuring 17.5 mm in diameter, and connected distally to the coronary sinus. There was partial unroofing of the coronary sinus, and stenosis at its mouth. When a balloon was used to occlude the left superior caval vein, there was an increase in left internal jugular venous pressure from 12 to 20 mmHg, and an increase in systemic arterial saturation to 100%, with a partial pressure of oxygen of 413 mmHg when breathing 100% oxygen. The left superior caval vein was occluded by transcatheter device such that the coronary sinus remained draining in unobstructed fashion to the left atrium. The strategy consisted of transeptal puncture from the right femoral vein, a balloon catheter (Tyshak II; 18 mm diameter, 3 cm balloon length, Numed Inc, Nicolville, NY) being advanced from the left atrium into the distal end of the left superior caval vein. The inflated balloon was placed to prevent embolization of the subsequently positioned occlusion device (Fig. 1). The device chosen was an Amplatzer Spider (Cook, Bloomington IN), deployed from the left internal jugular vein in the midsegment of the left superior caval vein. The device was compacted with detachable controlled release and Gianturco coils. Angiography confirmed total occlusion of the left superior caval vein. The hemiazygous connection to the left superior caval vein was inconsequentially sacrificed. The unroofed segment of the coronary sinus allowed unobstructed decompression of the coronary venous return into the left atrium.

Our second patient was a 7-month-old infant with hypoplastic left heart syndrome and bilateral superior caval veins in absence of a bridging brachiocephalic vein who had been palliated with the first stage of the Norwood operation. Cardiac catheterization was performed at 4 months of age, and showed a large atrial septal defect and an unobstructed aortic arch. Venography revealed a left superior caval vein draining to a dilated coronary sinus, with stenosis of midsegment of the left superior caval vein. Resonance imaging was undertaken to delineate further the venous anatomy. The right internal jugular vein, right subclavian vein, and the right superior caval vein were patent, as was the left subclavian vein. The left internal jugular vein was not visualized. The coronary sinus connected to the contiguous segment of the left superior caval vein, which was a string like channel coursing in its expected location. The infant underwent a right-sided bidirectional cavopulmonary connection and the left superior caval vein was ligated. The postoperative course was punctuated by respiratory distress. Despite optimized respiratory support, the infant had persistently low systemic oxygen saturation of oxygen of 55% in room air. Cardiac catheterization was performed at 7 months of age to identify the cause of the desaturation, and to assess the suitability for possible cardiac transplantation.

Figure 1.
An anterioposterior angiogram in our first patient showing an inflated balloon catheter in the distal end of the left superior caval vein, placed to prevent embolization of the device (left panel). An angiogram (right panel, same projection) shows complete occlusion of the left superior caval vein using an Amplatzer Spider compacted with coils.

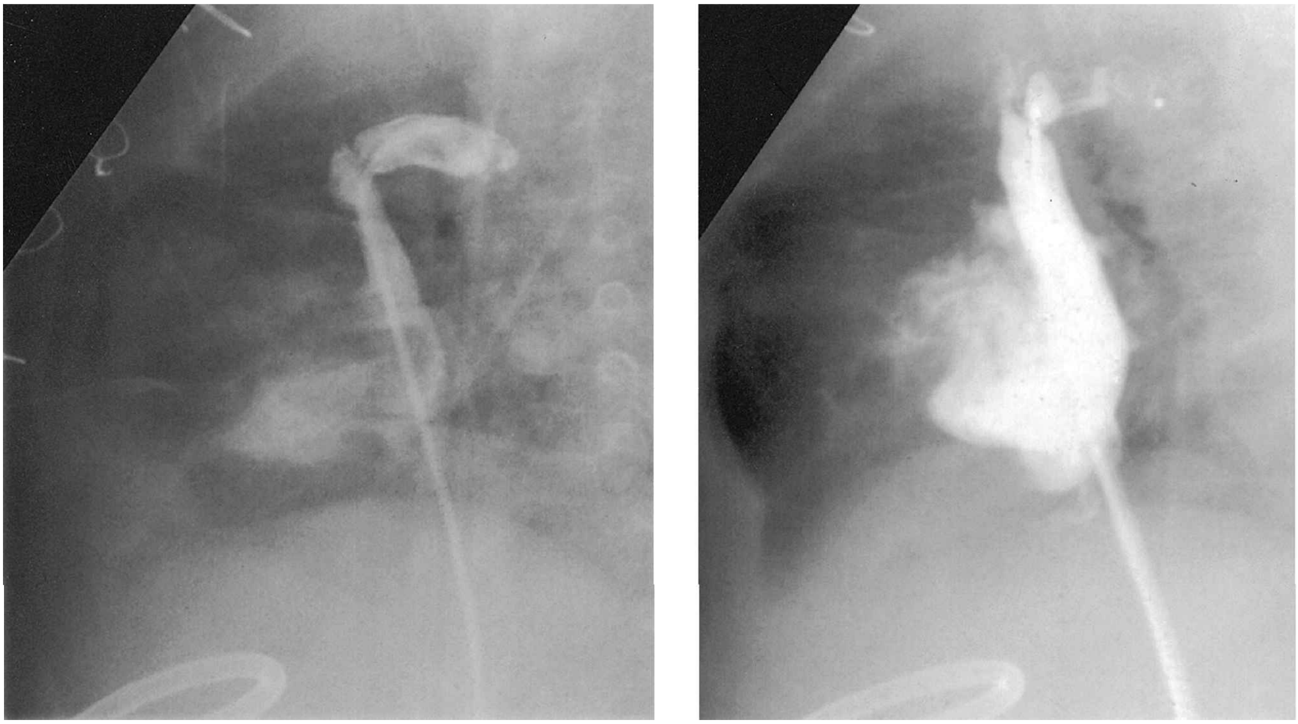

Figure 2.
A lateral angiogram in our second patient, following injection in the mouth of the hemiazygous vein, which drains into a remnant of the left superior caval vein (left panel). This was successfully occluded with an Amplatzer Duct Occlude implant delivered into the left superior caval vein through the unroofed segment of the coronary sinus (right panel; lateral view).

Angiography showed a large hemiazygous vein draining to the coronary sinus through the remnant of the left superior caval vein. An Amplatzer Duct Occlude implant (AGA Medical, Golden Valley, MN) was placed at the junction of the hemiazygous vein with the left superior caval vein, the device being positioned through the unroofed distal end of the coronary sinus from a right femoral venous approach (Fig. 2). Angiography confirmed total occlusion of the left superior caval vein, with systemic saturation improving to 70%. Cardiac transplantation was deferred.

Our third patient, a 4-year-old girl, had undergone an extracardiac total cavopulmonary connection with a 4 mm fenestration in the setting of double outlet right ventricle, pulmonary stenosis, and a non-committed ventricular septal defect. The initial surgical palliation included banding of the pulmonary trunk as a newborn, followed by Damus-Kaye-Stanzel anastomosis with an atrial septectomy, a bidirectional cavopulmonary connection and a patch pulmonary arterioplasty all completed at 5 months of age. Cardiac catheterization before the total cavopulmonary connection revealed a large left superior caval vein draining into the coronary sinus, which was ligated surgically during the total cavopulmonary connection. Multiple interventions were required following the surgery for persistent pleural effusions including placement of a stent in the left pulmonary artery, coil embolization of aortopulmonary collateral arteries, surgical ligation of the thoracic duct, decortication of the right lung and pleurocentesis. Only after these procedures could pulmonary rehabilitation be achieved. Ten months after the total cavopulmonary connection, cardiac catheterization was performed for evaluation of inappropriate systemic desaturation. The mean pressure within the total cavopulmonary connection was 18 mmHg, and angiography demonstrated a tiny fenestration unlikely to cause a systemic oxygen saturation of 70%. Angiography in the inferior caval vein showed a prominent hemiazygous vein draining to the pulmonary venous atrium though a remnant of the left superior caval vein itself draining to the coronary sinus. Test occlusion of the left superior caval vein, resulted in 100% systemic oxygen saturation. The left superior caval vein was occluded with an Amplatzer Duct Occlude implant (AGA Medical) from a right femoral venous approach (Fig. 3). A small hepatic vein forming a left atrial collateral channel was also occluded with a single Gianturco coil.

Discussion

The embryological precursors of the right and the left superior caval veins are the right and the left anterior cardinal veins. The unilateral right-sided system with usual arrangement is speculated to

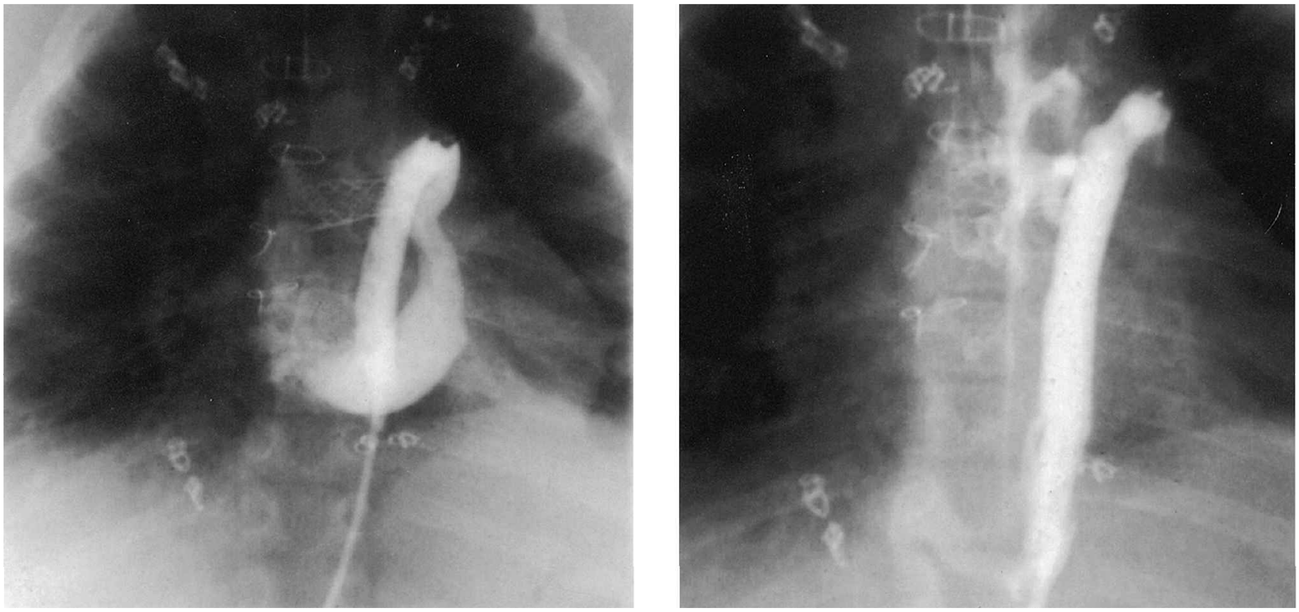

Figure 3.
An anterioposterior angiogram in our third patient shows the hemiazygous vein draining to the coronary sinus via the distal left superior caval vein following ligation of the proximal left superior caval vein (left panel). An angiogram (right panel) shows complete occlusion of the hemiazygous vein at the site of connection to the distal left superior caval vein following placement of an Amplatzer Duct Occlude implant.

develop due to obliteration of the left anterior cardinal vein between the left atrium and the hilum of the left lung.[8,26] Whilst up to one person in each 200 of the general population are said to have persistence of the left superior caval vein, the incidence ranges from one-tenth of those with congenital cardiac lesions in the setting of the usual arrangement, to three-quarters of those with cardiac lesions in the setting of isomerism of the atrial appendages.[1,8,13,26]

The physiological impact of the persisting vein is influenced by its connections and the associated cardiac lesions.[27] Most commonly, the left superior caval vein drains into the right atrium through the coronary sinus, generally causing no physiological embarrassment. Less commonly, it drains to the left atrium directly, or indirectly due to partial or complete unroofing of the coronary sinus.

When the drainage is to the right atrium via an otherwise intact coronary sinus, the presence of the left superior caval vein is inconsequential. Indeed, it can serve as a conduit for cerebral protection during surgical procedures on the aorta.[28,29] Its presence modifies the approach used during insertion of transvenous pacemakers or extracorporeal membrane oxygenation in the absence of a bridging brachiocephalic vein.[30,31] It may, however, cause or aggravate obstruction to left ventricular inflow by creating a subdivided left atrium with pulmonary venous obstruction,[5,6,10] particularly when there is no bridging brachiocephalic vein. In this case, the coronary sinus dilates and results in obstruction to the inflow of the left ventricle and the potential for a left-to-right shunt through the vein.[12]

An obligatory right-to-left shunt occurs when the persisting left superior caval vein connects to an unroofed coronary sinus, or when it is connected directly to the left atrium. Such connections cause positional or persistent cyanosis, clubbing of the nail beds and hypoxemia with the risk of paradoxical embolization.[7,11,27] When obstructive lesions of the left heart coexist, a left-to-right shunt can occur from the left atrium to the systemic venous circulation through the vein. As such, associated lesions that cause hypoxemia may mask the large left-to-right shunts.

In complex lesions requiring functionally univentricular palliation, the importance of recognizing the left superior caval vein if present is critical.[8,14,15] Its presence in the absence of a bridging brachiocephalic vein reduces the efficiency of a bidirectional cavopulmonary connection by reducing effective pulmonary blood flow, by channelling venous blood away from the pulmonary circulation. To avoid this, bilateral bidirectional cavopulmonary connections are required.[32] In the total cavopulmonary connection, a persisting left superior caval vein results in an obligatory right-to-left shunt through the coronary sinus which is generally incorporated into the pulmonary venous atrium by a wide atrial septectomy. If the coronary sinus drains into the systemic venous chamber, the left superior caval vein may undergo aneurysmal dilation.[33] It is, therefore, common practice to ligate a left superior caval vein if present, or perform bilateral bidirectional cavopulmonary connections if a bridging brachiocephalic vein is not present. A left superior caval vein may open as a new event, or its remnant may enlarge following the bidirectional cavopulmonary connection or a total cavopulmonary connection if surgical ligation has not been performed.[8]

Transcatheter treatment of a persisting left superior caval vein requires angiographic definition of its connections, and the anatomy of the coronary sinus. Stenosis or atresia of the mouth of the coronary sinus precludes any effort to occlude the caval vein, as it is the only route for decompression of coronary venous return. The absence of a bridging vein also precludes occlusion if occlusion with a balloon produces a rise in the pressure in the left internal jugular vein to above 30 mmHg.[34] The location of a hemiazygous vein should be defined to direct the placement of devices either across its orifice, or between it and the left atrium. Failure to do so can result in right-to-left flow from the inferior caval vein through the hemiazygous venous system and the distal segment of the left superior caval vein. An occlusion device can be delivered into the left superior caval vein from several routes. The device can be placed through the mouth of the coronary sinus from the inferior caval vein. Alternatively, if there is an unroofed coronary sinus, the approach can be through the right atrium as in our second patient, or from the hemiazygous vein through the inferior caval vein as in our third. This is necessary when the left superior caval vein can be ligated proximally, precluding an approach from the left internal jugular vein. An approach from the left internal jugular vein is the most direct route if the anatomy is suitable. A combination of transeptal needle puncture, placing a balloon at the distal end of the left superior caval vein to prevent embolization, with delivery of the device through the left internal jugular vein as in our first patient, is also an effective strategy. This is helpful when the left superior caval vein is a large vessel with a wide connection to the left atrium. The technique is an extension of a strategy described for occlusion of systemic-to-pulmonary arterial shunts.[35]

Numerous devices have been used for occlusion of a persisting left superior caval vein, including the Gianturco Grifka Vascular Occlusion Device (Cook),[15] the Greenfield filter packed with Gianturco coils (Cook),[18] a Rashkind double umbrella device (CR Bard Inc, Billerica, MA),[17] a Palmaz stent (Johnson & Johnson Interventional, Warren NJ) packed with a Gianturco Grifka Vascular Occlusion Device and platinum detachable embolization coils,[36] and the Gunther Tulip Vena Cava Mreye Filter packed with Cook detachable coils.[16] Our experience records additional devices used for transcatheter approaches to a left superior caval vein.

If surgical intervention is required to address associated lesions, a left superior caval vein can be treated in a number of ways. The approach can be intracardiac in the form of an intraatrial baffle, channelling the left superior caval vein to the right atrium by plication of the left atrial posterior wall. Extracardiac approaches, in the absence of a bridging vein, include an end-to-side connection of the left superior caval vein to the right superior caval vein. The connection may be routed anterior to the aorta, or through the space between the aorta and the pulmonary trunk. An anastomosis of the distal end of a left superior caval vein to the tip of the right atrial appendage can be performed, and is facilitated if there is juxtaposition of the atrial appendages. A left-sided bidirectional cavopulmonary connection is another alternative.[6,19,25] Surgical ligation of the left superior caval vein can also be undertaken in the absence of a bridging vein taking care to measure pressures to avoid potential development of a caval syndrome.[34] When considering surgical ligation, the connections of the left superior caval vein should be determined. If the site of ligation is superior to the site of entry of the hemiazygous vein, the latter vein can be the source of an obligatory right-to-left shunt through the distal left superior caval vein, as occurred in our third patient. The distal left superior caval vein, inferior to the entry of the hemiazygous vein, is the preferred site for surgical ligation. Occlusion of a persisting left superior caval vein in the presence of orificial stenosis or atresia of the coronary sinus can result in high coronary venous pressures and impair myocardial function.[20,37,38] Stenosis or atresia, therefore should be excluded prior to intervention.

In summary, a left superior caval vein may be of no consequence. It can, however, cause physiological compromise due to its connections or associated cardiac lesions. It can be occluded by several different transcatheter approaches, and by using a variety of devices. We have reviewed its physiologic impact, highlighted the role of surgical intervention, and defined the strategies of transcatheter and surgical treatments.

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