REVIEW

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Unroofed coronary sinus: update on diagnosis and treatment

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Abstract

Unroofed coronary sinus (UCS) is among the rarest congenital heart malformations in pediatric practice. Often, they are associated with persistent left superior vena cava (PLSVC) draining into the left atrium, and in this situation, there is a double intracardiac shunt, right-to-left and left-to-right. They are asymptomatic in the majority of the cases. Each ordinary imagistic method (transthoracic echocardiography, contrast transesophageal echocardiography, angiography) can be helpful in finding the diagnosis, but enhanced computed tomography scan can give an irrefutable diagnosis. The surgical intervention may consist in the recreation of a roof for the coronary sinus, and anastomosis of the PLSVC to the right atrial appendage.

Keywords: unroofed coronary sinus, agenesia of the coronary sinus, congenital heart disease, dyspnea, persistent left superior vena cava.

Introduction

Unroofed coronary sinus (UCS) is a congenital heart disease (CHD) characterized by defects in the formation of the coronary sinus (CS). CS is an anatomical structure of the heart connecting the coronary venous collecting system to the right atrium (RA). It opens into the RA superior to the tricuspid valve, being guarded by a crescent-shaped valve, the Thebesian valve. Between the coronary ostium and the inferior vena cava, there is a commissure which is connected to the membranous septum by the tendon of Todaro. The position of the CS is in the left posterior atrioventricular groove along with the circumflex coronary artery, at the inferior part of the left atrium (LA). Although it is closely attached to the LA, both CS and LA have their own walls. CS defects are considered "occult" type of atrial septal defects (ASDs) and they may have different aspects [1, 2].

CS anomalies may appear as rupture of the wall, as complete absence of the wall of the CS, as CS agenesis or as stenosis of the orifice. They are very rare representing less than 1% of all ASDs. Seventy-five percent of them are associated with persistent left superior vena cava (PLSVC) [3, 4].

History and classification

Raghib *et al.* were the first to describe the UCS for the first time in 1965 [5]. There are two forms of UCS – partial and complete. In the partial form, there is a mild/moderate opening into the roof of the CS, and practically, CS communicates with both LA and RA. The complete form may be simple or associated with persistence of the left superior vena cava (LSVC) to the LA. In fact, the last one was the form described by Raghib *et al.* as an association of PLSVC into the LA with the absence of the CS and a defect in the posteroinferior angle of the atrial septum. In many cases of UCS, the associated PLSVC masks the UCS. Congenital malformations of the CS were classified differently according to the anatomical and pathological presentation [6].

Kirklin & Barrat-Boyes [7] described four types, dividing the partial form into two: partially unroofed mid portion and partially unroofed terminal portion. They are described as type I, completely unroofed with LSVC; type II, completely unroofed without LSVC; type III, partially unroofed midportion; type IV, partially unroofed terminal portion [7–10].

In the complete form of this defect, there is total agenesia of the roof of the coronary sinus (UCS) with a complete, large communication between the LA and the CS, without any wall between them. The UCS associated with the PLSVC into the LA is called Raghib defect [11]. Raghib *et al.* described eight cases with this association: absence of CS, a persistence of PLSVC to the LA, and a posteroinferior ASD [5]. The most part of all these was associated with the heterotaxic syndrome and asplenia [10].

In the partial form, there is a mild/moderate opening into the roof of the CS, and practically, CS communicates with both LA and RA.

PLSVC was described in 1787. The most part of them (92%) are draining into the RA through the CS. The rest of them are draining into the LA being associated with partially or completely UCS [2].

Mantini *et al.*, taken into consideration all aspects of the CS anomalies, classified them as follows [9]:

(I) Enlargement of the CS:
  (A) Without left-to-right shunt into the CS:
    (1) PLSVC;
    (2) Partial anomalous hepatic venous connection to the CS;
    (3) Continuity of inferior vena cava with left superior vena cava (SVC) through hemiazygos vein.
  (B) With left-to-right shunt into the CS:
    (1) Low pressure shunts;
      (a) Communication of CS with LA;
      (b) Pulmonary venous connection to the CS.
    (2) High pressure shunts:
      (a) Coronary artery–CS fistula.
(II) The absence of CS:
  (A) ASD localized to the position normally occupied by the CS;
  (B) An ASD involving the entire lowermost portion of the septum:
    (1) Persistent common atrioventricular canal;
    (2) Asplenia with congenital cardiac disease.
(III) Atresia of the right atrial CS ostium:
  (A) PLSVC;
  (B) Gross communication of CS with the LA;
  (C) Multiple communications between the CS and related atria.
(IV) Hypoplasia of the CS.

In the presence of atresia of the CS ostium associated with PLSVC, the blood that flows from the coronary veins will drain either through the PLSVC or through the innominate vein into the right SVC. However, if a PLSVC is missing, then the coronary venous blood will arrive into the related atria by large or multiple small communications, intracardiac Thebesian veins, generating a right-to-left shunt, insignificant from hemodynamical and clinical point of view [12–14].

Embryology

Embryologically, both UCS and persistence of LSVC are related with the venous system connected to the *sinus venosus*, which is a part of the primitive cardiac tube. *Sinus venosus* has two symmetrical horns, which are connected to the entire venous system of the embryo – the cardinal, umbilical, and vitelline veins. Both left and right SVC origin from the ipsilateral common and anterior cardinal veins [12]. Later, during the development of the left innominate vein, the LSVC regress and becomes the ligament of Marshall. The CS has its origin into the left horn of the *sinus venosus* and the left common cardinal vein. The transverse part of the *sinus venosus* suffers a rightward shifting and pulls the left horn into the posterior atrioventricular groove. A defect into the wall of the CS can be explained by resorption of the wall while persistence of the LSVC, as a failure in the involution of the left anterior and left common cardinal veins. Therefore, it is normal for a PLSVC to drain into the CS, because they both have the same embryological origin and to be connected to the LA only in the presence of an UCS. These anomalies are reported also in association with the isolated right isomerism [12, 15, 16].

Clinical presentation

Clinical manifestations are variable. They may be asymptomatic for a long time and many cases have been described in the literature in which the diagnosis of UCS has been established in adulthood.

Clinical manifestations may embrace the classic clinical appearance of left–right shunt malformations, which can be manifested by dyspnea of effort or rest, fatigue, tachypnea, hepatomegaly and other signs of heart failure, increasing delay, malnutrition [17]. Palpitations may be a reason for presentation, especially in adults, and are associated with right cavity dilatation [18]. Also, syncope, peripheral edema, and recurrent lung infections have been described as modalities of onset [19].

Also, when the PLSVC is associated with the UCS, the patient may experience central cyanosis or other severe complications such as cerebral abscess, paradoxical embolism. Cardiac abnormalities of the CS are less noisy and difficult to diagnose and may associate systemic desaturation, which can be diagnosed with the occasion of various other presentations [6, 20–24]. In case of association of more complex anomalies, clinical manifestations may be different, depending on associations. Also, if there is stenosis or atrophy of the coronary artery, the clinical manifestations will be characteristic of right–left shunt [2].

At the clinical examination, a hyperkinetic cord, the Harzer sign, systolic murmur especially in newborns and infants (corresponding to functional pulmonary stenosis) may be perceived. Cardiac rhythm may be rapid, showing tachyarrhythmias. Arrhythmias occur especially in patients diagnosed later, and predominant supraventricular arrhythmias (supraventricular tachycardia, atrial fibrillation) [19, 25, 26].

Investigations

Investigations follow the typical diagnostic evaluation.

Chest radiogram, which is done with a different purpose, may show a shadow along the left upper border of the mediastinum, corresponding to the LSVC. Characteristic signs of the left-to-right shunt are cardiomegaly, dilated pulmonary artery shadow and increased pulmonary vascularity [2, 19].

The electrocardiogram (ECG) may show typical signs of ASD, enlargement of the right atria, right ventricle hypertrophy, and incomplete right bundle branch block. The axis may be to the right, superior for associated *ostium primum* defect, inferior for *sinus venosus* ASD, and abnormal in cardiac isomerism with a left sinoatrial node or ectopic cardiac rhythm [2, 19].

As imagistic tools, transthoracic echocardiography is the first to evaluate each child and adult with a suspicion of a CHD [27, 28]. The transthoracic echocardiographic examination may reveal right-side heart enlargement, an enlarged opening of the CS into the RA or absence of the CS and PLSVC (Figure 1). Sometimes, an UCS in the presence of an *ostium secundum* ASD may be missed and in such a case, the patient needs a second intervention [21, 29].

The dilated CS ostium, which appears as an inferior interatrial communication, may be confused with an *ostium primum* defect. The enlarged CS has to be visualized from different views. One of these is the parasternal long-axis

view in which a gap between the CS and the LA is seen and it is confirmed by color Doppler evaluation. In the case of absence of a clear CS, a complete UCS can be suspected. A partial UCS can be suspected if the CS can be identified, but a part of it cannot be seen. Using the subcostal view, the chances for a correct diagnosis are reported at 100% of *primum* ASDs, 89% of *secundum* ASDs, and 44% of *sinus venosus* ASDs [19, 29–31].

Special attention has to be paid for the presence of the PLSVC, a component of the Raghib syndrome. The termination of the PLSVC is very difficult to be visualized even using color Doppler. The position is anterior to the left upper pulmonary vein and posterior to the left atrial appendage [2, 29]. LSVC can be best visualized from subcostal, precordial or suprasternal view.

However, in some cases, the transthoracic examination may not be as helpful as it is needed due to obesity, scarce transthoracic window. Therefore, the next evaluation for all patients consists of contrast transthoracic. This is inappropriate for a scarce transthoracic window (Figure 2) for which transesophageal echocardiography (TEE) is the next choice. For contrast transthoracic echocardiography, an injection of bubble contrast into the left arm is needed. In the presence of a PLSVC draining into the LA, the contrast will appear initially into the LA and subsequent into the RA [2, 29, 32].

Establishing the diagnosis of the UCS by using contrast echocardiography can be misleading in the case of the coexistence of an innominate vein between the left and right SVC with the PLSVC (Figures 3 and 4). In this situation, injection of contrast media/bubbles into the left arm will be associated with opacifying of both atria at the same time, with or without UCS. Sometimes, a washout effect can be seen at the level of the CS, due to the left-to-right shunt, especially if there is only a partial agenesia of the CS.

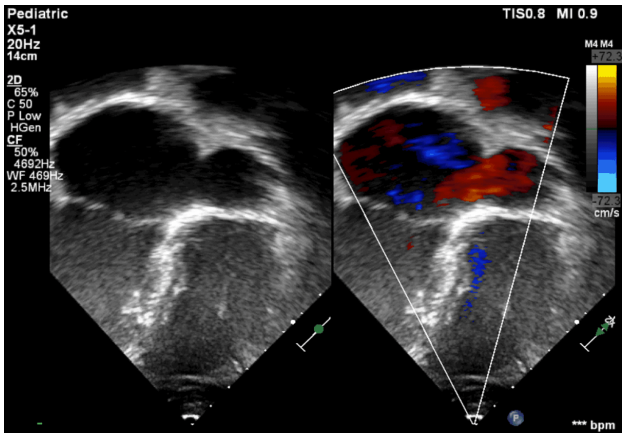

Figure 1 – ***12-year-old male patient with Raghib syndrome admitted in "Maria Skłodowska Curie" Emergency Children's Hospital (Bucharest, Romania) with unroofed coronary sinus associated with persistent left superior – transthoracic echocardiography in apical four chambers showing dilated right chambers and dilated coronary sinus.***

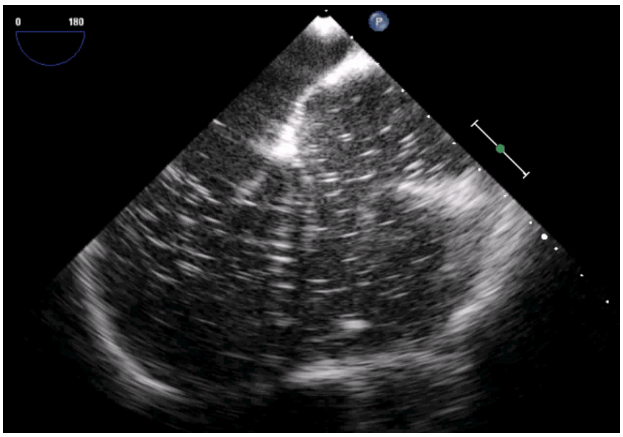

Figure 2 – ***The same patient. Contrast transesophageal echocardiography showing opacification of both atria in the same time due to the presence of an innominate vein connecting the left superior vena cava to the right superior vena cava.***

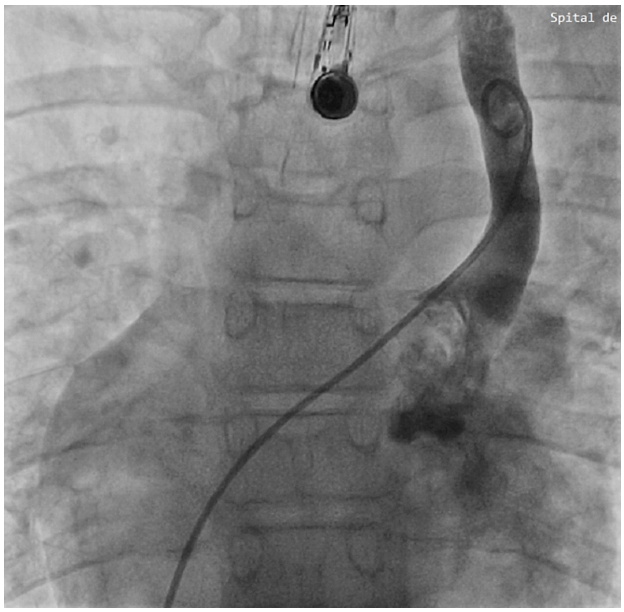

Figure 3 – ***Angiographic evaluation of the persistent left superior vena cava connected to the left atrium and the absence of the roof of the coronary sinus.***

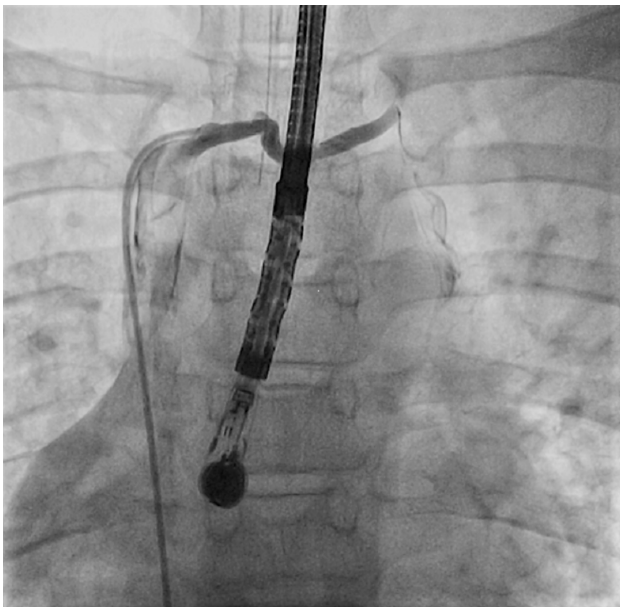

Figure 4 – ***Angiographic evaluation of the hypoplastic innominate vein in the same patient.***

TEE and three-dimensional (3D) echocardiography may play an important place in UCS evaluations. There are cases in literature in which the contribution of these techniques was essential for a positive diagnosis, sometimes in obese patients or after cardiac surgery. Best images for CS evaluation using TEE are the high four-chamber view and the midesophageal long axis view [33–39].

Computed tomography (CT) scan may also not visualize exactly the dimensions of the opening of the UCS and also may be imprecisely related to the opening of the PLSVC but using the cardiac ECG-gated 64-slice multi-detector-row computed tomography (MDCT) all this detailed information is obtained (Figure 5) [40, 41]. Sometimes, CT scan may reveal asymptomatic types of UCS with CS stenosis. For these patients, there is no need for surgery, only follow-up [42]. Besides the severe stenosis in the orifice, a dilated CS may be present also in CS atresia. Not always, the CS atresia presents with enlarged CS. The most important views for CS evaluation are the cardiac short axis and long axis views in the plane of the atrio-ventricular groove [4, 43–48].

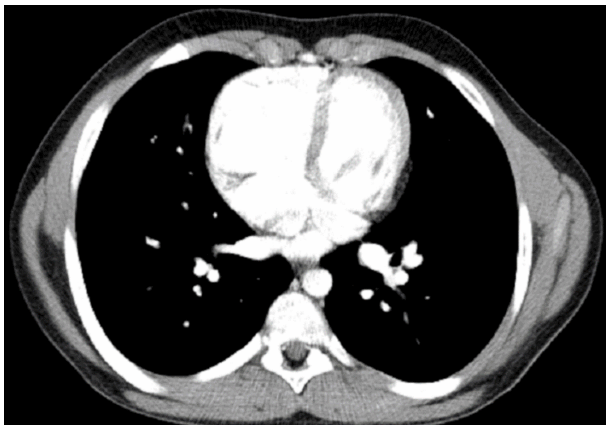

Figure 5 – ***CT scan image showing dilated right heart with dilated coronary sinus in the same patient. CT: Computed tomography.***

Cardiovascular magnetic resonance (CMR) is an important tool in the diagnosis of the UCS, but frequently it is an incidental finding. CMR may be extremely important for an anatomical confirmation, seeing the defect in the axial, oblique, sagittal planes. By CMR, Qp/Qs may be calculated using phase contrast CMR [49].

Cardiac catheterization and angiography had to be performed in order to clarify the anatomy. However, angiography also has its limitations because there is a limitation to a volume of contrast agent especially in a monoplane angiograph and in the case of dual shunting, enlarged heart chambers, sometimes this can offer an imprecise diagnosis [50].

At cardiac catheterization, sometimes by sampling at different levels, a significant decrease of oxygen saturation samples between the pulmonary veins and the LA may be evident [2, 10]. Also, measuring the oxygen concentration by fiber-optic catheter pull-back, during cardiac surgery, one may have a confirmation of the diagnosis [51]. Not always, the initial imagistic is able to diagnose the UCS [11] or if the contrast agent is injected through the right arm even with enhanced CT scan [28]. The best technique for such cases is the left arm contrast injection during the enhanced CT scan [28].

Associated lesions

Other CHD, besides PLSVC, may be associated with this defect such as atrial isomerism, ASD, atrioventricular septal defect, mitral stenosis, mitral atresia, tricuspid atresia, tetralogy of Fallot, all being described for the first time by Raghib *et al.* [5, 11, 20, 52]. They may function predominantly as right-to-left heart disease, like in cases of abnormal systemic venous return [53] increasing the risk for cerebral embolism and abscess or predominantly like an "occult" left-to-right shunt with dyspnea and enlarged ride-sided heart. Sometimes, the diagnosis is missed due to dropping attention of other CHD, such as the single ventricle, associated atrial and ventricular septal defect, atrioventricular septal defect, etc. [8, 54], which may explain the desaturation or the dyspnea and heart enlargement [55–58].

There were also other associated lesions described like absent right SVC (in 1% of all cases with PLSVC) [59], *ostium secundum* ASD [21], partial anomalous pulmonary venous return [60], interrupted inferior vena cava with hemi-azygos continuation [3], double outlet right ventricle, ventricular septal defect, aortic coarctation [61, 62], and anomalous hepatic vein drainage [30], complete atrio-ventricular canal (CAVC) associated with total anomalous pulmonary venous return and UCS [63].

Interventions

Indications for correction of the UCS are related to the amplitude of the ASD. For large defects, significant UCS correction is needed, but for small defects, the indication is only for regular follow-up [64]. Also, if a complication related to the right-to-left shunt occurs, the indication is clear [24].

Classically, these defects were surgical ones. Recently, there were attempts for interventional correction.

The anatomy and the associated CHD are very important for decision making regarding the indication and the type of procedure.

There are many operatory techniques for this CHD. The best technique is chosen accordingly with the dimensions of the PLSVC and innominate vein or of the UCS. If there is not an innominate vein or if the dimensions of the PLSVC are important, the UCS cardiac surgery consists of patching the defect, by reroofing the CS on the posterior wall of the LA (Figure 6). In UCS associated with PLSVC into the LA, surgical techniques aimed at redirecting the blood from the CS or from PLSVC to RA by resecting the existing interatrial septum and reconstructing it from bovine pericardium or synthetic material so as to keep the non-mixing of the two systemic and pulmonary circulations [11].

For the situation where the PLSVC is of significant flow and the defect in the CS is small, it is possible to opt for interventional PLSVC closure using a vascular plug, after having previously undergone an occlusion test of the PLSVC with observation of the increase of the overlying pressures and the presence of the collateral circulation to the right SVC [65].

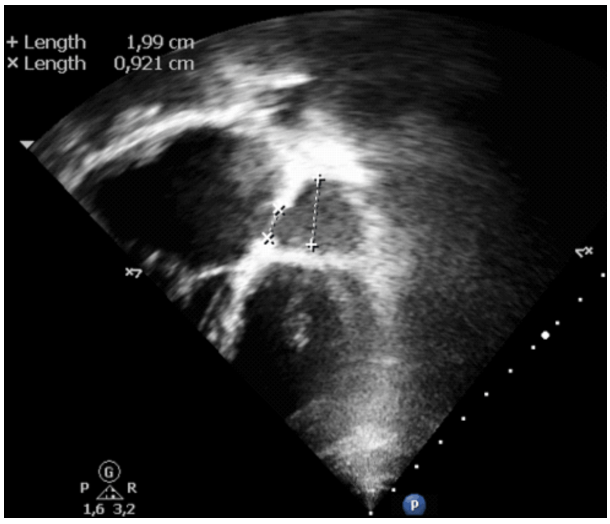

Figure 6 – ***Postoperative transthoracic echocardiography of the reconstructed coronary sinus with patching of the roof of the coronary sinus in the patient with Raghib syndrome. The persistent left superior vena cava was extracardiac connected to the right atrial appendage.***

Many other techniques were described for PLSVC connected to the LA. The surgery must be done for ligation of the PLSVC when there is a good connection to the right SVC. For absent bridging vein (innominate vein), it is mandatory to redirect the flow from the left part of the neck and head, and the left arm towards the right. Other techniques may be applied by using an intra-atrial baffle [66], transection and reconnection of the PLSVC either to the right SVC, to the right auricular appendage (RAA) or to the RAA through the left auricular appendage together with the reconstruction of the CS roof [67, 68]. Also, redirection of the PLSVC to the left pulmonary artery was also, described, the cavopulmonary anastomosis [11]. This is feasible if the left pulmonary artery has a good caliber and the pulmonary pressures are low. Some authors decide the type of intervention after intraoperative measurement of the pressure into the LSVC. If the pressure is less than 15 mmHg (or 16 mmHg according to other authors) and there is a good caliber innominate vein, the PLSVC will be ligated.

Transcatheter closure of the PLSVC even in the presence of a UCS was described, using a vascular plug or other devices [69, 70].

The PLSVC has to be taken into account regarding the cardiopulmonary bypass circulation [71]. Extracardiac repair also has a reduced bypass time comparing with the intracardiac technique.

Operatory techniques are synthetized in Table 1 [8, 9, 12, 20, 24, 30, 34, 63, 67, 72–77].

Table 1 – ***Surgical interventions feasible for UCS***

(I) For UCS without LSVC
(a) Correction of the defect by reroofing the CS – especially in midportion defects.
(b) Closure of the CS ostium with a patch. It is indicated more in cases in which the defect is nearby the ostium of the CS. Disadvantage – the venous coronary blood will drain into the LA generating a desaturation (insignificant) because the flow from the CS is responsible for 4–5% of the cardiac output. Contraindication – if there is a LSVC present. Some authors chose this method for type II of UCS (completely unroofed without LSVC).
(II) For UCS associated with PLSVC. The PLSVC has to be ligated or redirected, depending on the caliber of the innominate vein.
(a) If there is a good calibre-connecting vein, the LSVC can be ligated below the bifurcation and the UCS reroofed.
(b) If the innominate vein is hypoplastic:
(*i*) Its augmentation using a pericardial patch and ligation of the distal LSVC.
(c) If the innominate vein is hypoplastic or absent:
(*i*) Intracardiac redirection of the LSVC and ostium closure. Redirection of the PLSVC towards the RA is done by resecting the existing interatrial septum and reconstructing it from bovine pericardium or synthetic material, as an intracardiac baffle. For intracardiac repair, the left atrial appendage may be used by plicating the posterior wall of the LA, by using a patch, or by using the inverted left atrial appendage as a baffle.
(*ii*) Anastomosis of the LSVC using extracardiac techniques – reimplantation of the LSVC into the pulmonary artery (bidirectional left cavopulmonary anastomosis), into the right atrial appendage or into the right SVC with anterior or retro-aortic course.

UCS: Unroofed coronary sinus; LSVC: Left superior vena cava; CS: Coronary sinus; LA: Left atrium; PLSVC: Persistent left superior vena cava; RA: Right atrium; SVC: Superior vena cava.

Transcatheter interventional treatment can be addressed to:

(1) Closure of the ostium of the CS, in the absence of the PLSVC to the LA, by using an Amplatzer™ Septal Occluder (ASO) [78].
(2) Closure of the communication by using an ASO device, in another case of an UCS associated with PLSVC. The solution was to close effectively the communication, and to let the LSVC to drain into the RA through the CS. In such a case, the right disc of the ASO should be less than one-third of the diameter of the CS and the flow should not be turbulent or accelerated [79, 80].
(3) Closure of the communication by using a covered stent [17].
(4) Closure of the communication between the LSVC and LA in the mid portion of the vessel, by using caval vein filter, coil [81], vascular occlusion device [69] or Amplatzer™ ASD Occluder. In this case, the simple approach is to access the left internal jugular vein. The device should be at least 2 mm larger than the vessel or the diameter of the vessel multiplicated by 1.3–1.5%.

Postprocedural course and complications

The evolution of UCS depends very much of the dimension of the defect and if there are other cardiac malformations. The defect of the UCS can close spontaneously. This was the case of complex anatomy (mitral atresia, double outlet right ventricle, persistent *ductus arteriosus*, patent *foramen ovale*) associated with a fenestration between the LA and the CS in which the closure followed the Blalock–Hanlon procedure and was documented at angiography [82].

In patients without cardiac intervention or late intervention, cardiac complications are more frequent: late cardiac failure, stroke, atrial fibrillation [83].

Postoperative, any obstruction on the route of the PLSVC or CS, any residual shunt at the CS level or other intracardiac shunts are to be ruled out by echocardiography or, in difficult situations, by CT or magnetic resonance imaging (MRI) [11].

Reported complications are presented in Table 2 [8, 24, 67, 72, 75, 76, 78, 80].

Table 2 – ***Complications pre- and postoperative of the UCS with or without persistence of the LSVC***

Preoperatory	Postsurgical	Postinterventional
▪ Cerebral abscess; ▪ Stroke; ▪ Heart failure; ▪ Pulmonary hypertension; ▪ Atrial fibrillation; ▪ Supraventricular tachycardia.	▪ Obstruction of the LSVC or of the CS; ▪ Residual shunt of the CS; ▪ Intracardiac shunt; ▪ Stenosis of the baffle; ▪ Early detachments and late deterioration of the baffle; ▪ Pulmonary venous obstruction and congestion due to pulmonary vein stenosis or mitral stenosis with abnormal hypertrophy of the vascular walls; ▪ Creation of a small left atrial compartment with repercusions on ventricular filling for intraatrial baffle repair; ▪ Homograft vein stenosis or occlusion [63]; ▪ Complete atrioventricular block [84]; ▪ Pulmonary venous obstruction; ▪ Cerebral and upper limb obstruction: venous engorgements, facial edema and chylothorax.	▪ Residual shunt of the CS; ▪ Obstructions of the CS; ▪ Obstruction with a device in ASO implantation to close UCS; ▪ Supraventricular tachycardia in UCS closure [80]; ▪ Clot formation at the proximal disc site of the vascular device (even under anticoagulant therapy); ▪ Covering the coronary vein ostium in covered stent implantation associated with complete atrioventricular block, ventricular dysfunction and hemodynamic decompensation [17]; ▪ Left disc herniation into the CS with moderate residual shunt in CS ostium device closure [72].

UCS: Unroofed coronary sinus; LSVC: Left superior vena cava; CS: Coronary sinus; ASO: Amplatzer™ Septal Occluder.

Conclusions

UCS associated with PLSVC is an extremely rare form of a CHD. They are functioning as an ASD with left-to-right shunt associated with right-to-left shunt by the abnormal systemic venous drainage of the PLSVC into the LA. Left-arm injection for enhanced CT is the method of choice and it should be used in any case of PLSVC or suspicion of UCS. The presence of the innominate vein may mislead the possible diagnosis provided by contrast echocardiography. PLSVC and UCS may be a cause of systemic hypoxemia, brain abscess, cerebral emboli but depending on the volume of the flow it can also lead towards dyspnea, right chambers enlargement, and pulmonary hypertension. Surgery is the method of choice with redirecting the flow from the PLSVC towards the RA and reroofing of the CS. Also, interventional methods may be used in special cases of small CS defects.

Conflict of interests
None declared.

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