Archives of Cardiovascular Disease (2019) 112, 135—143

Available online at ScienceDirect www.sciencedirect.com

Elsevier Masson France EM|consulte www.em-consulte.com/en

CLINICAL RESEARCH

Left superior vena cava draining into the left atrium: Clinical entities, diagnosis and surgical treatment

Veine cave supérieure gauche s'abouchant à l'oreillette gauche : entités cliniques, diagnostic et traitement chirurgical

Juan M. Aguilar[a], Fernando Rodríguez-Serrano[b,c], Andrea Ferreiro-Marzal[d], María Esteban-Molina[d], Antonio Gabucio[b], Enrique García[a], Lorenzo Boni[a], José M. Garrido[c,d,e,*]

[a] *Paediatric Heart Institute, Doce de Octubre University Hospital, 28041 Madrid, Spain*
[b] *Institute of Biopathology and Regenerative Medicine (IBIMER), University of Granada, 18100 Granada, Spain*
[c] *Biosanitary Research Institute of Granada (ibs.GRANADA), 18012 Granada, Spain*
[d] *Department of Cardiovascular Surgery, Virgen de las Nieves University Hospital, 18014 Granada, Spain*
[e] *Department of Surgery and its Specialities, University of Granada, 18016 Granada, Spain*

Received 24 January 2018; accepted 14 May 2018
Available online 1 September 2018

KEYWORDS
Congenital abnormality;

Summary Left superior vena cava draining into the left atrium in the absence of coronary sinus is an anomaly that can appear in heterotaxy syndrome and unroofed coronary sinus syndrome. Regardless of the origin of these syndromes, biventricular repair can be done through rerouting by intracardiac procedures or through disconnection-reconnection of the left superior vena

Abbreviations: LA:, left atrium; LSVC:, left superior vena cava; RA:, right atrium; RSVC:, right superior vena cava; TAPVC:, total anomalous pulmonary venous connection; UCSS:, unroofed coronary sinus syndrome.

* Corresponding author. Department of Surgery and its Specialities, University of Granada, Avd. de la Investigación 11, 18016 Granada, Spain.
E-mail address: josemgarrido@ugr.es (J.M. Garrido).

https://doi.org/10.1016/j.acvd.2018.05.007

Persistent left superior vena cava;
Heterotaxy syndrome;
Unroofed coronary sinus syndrome;
Surgical techniques

cava to the right atrium or right superior vena cava by extracardiac procedures. Different techniques can be used for this purpose, each of which has its own advantages and limitations. Therefore, appropriate selection is necessary to obtain the best results for each patient, and many factors, such as patient anatomy, age, associated cardiomyopathies, etc., have to be considered. In this review, we focus on heterotaxy and unroofed coronary sinus syndromes, associated cardiomyopathies, the state-of-the-art in their surgical treatment and our results in a sample of 10 patients. Our experience highlights the importance of accurate diagnosis and specific selection of surgical technique for the management of biventricular repair in patients with left superior vena cava draining into the left atrium in the absence of coronary sinus.

MOTS CLÉS
Anomalie congénitale ;
Veine cave supérieure gauche persistante ;
Syndrome d'hétérotaxie ;
Syndrome coronaire non affleuré ;
Techniques chirurgicales

Résumé La veine cave supérieure gauche en drainant à l'oreillette gauche en l'absence de sinus coronaire est une anomalie qui peut apparaître dans le syndrome d'hétérotaxie et le syndrome de l'agénésie du toit du sinus coronaire. Indépendamment de l'origine de ces syndromes, la réparation biventriculaire peut être effectuée par réacheminement par des procédures intracardiaques ou par déconnexion-reconnexion de la veine cave supérieure gauche à l'oreillette droite ou à la veine cave supérieure droite par des procédés extracardiaques. Il y a différentes techniques qui pourraient être utilisées à cette fin et toutes ont leurs avantages et leurs limites. Par conséquent, une sélection appropriée est déterminante afin d'obtenir les meilleurs résultats pour chaque patient, sans oublier de prendre en compte de nombreux facteurs tels que l'anatomie du patient, son âge, les cardiomyopathies associées, etc. Dans cette révision, nous nous intéressons aux cardiomyopathies associées aux syndromes coronariens hétérotaxiques et non-affleurés, aux traitements chirurgicaux d'avant garde les plus récents sans oublier les résultats que nous avons obtenus dans un échantillon de 10 patients, l'état de l'art dans leur traitement chirurgical et notre expérience souligne l'importance d'un diagnostic précis et d'une sélection spécifique de la technique chirurgicale adéquate pour la prise en charge de la réparation biventriculaire chez les patients ayant une veine cave supérieure gauche s'abouchant à l'oreillette gauche en l'absence de sinus coronaire.

Background

There are two different clinical entities in which a persistent left superior vena cava (LSVC) can drain into the left atrium (LA) in the absence of coronary sinus: heterotaxy syndrome and unroofed coronary sinus syndrome (UCSS). In these cases, biventricular repair can be done by intracardiac rerouting or by disconnection-reconnection to the right atrium (RA) or to the right superior vena cava (RSVC) using extracardiac procedures. In the period from 1998 to 2016, 10 patients with LSVC draining into the LA underwent 11 surgeries for biventricular repair. The present study focuses on heterotaxy syndrome and unroofed coronary sinus syndrome (UCSS), associated cardiomyopathies, the state of the art in their surgical treatment and our results in our sample. Our experience highlights that accurate diagnosis and appropriate selection of surgical technique, considering different characteristics, such as patient anatomy, age, associated cardiomyopathies, etc., are important for patient outcome.

UCSS

UCSS consists of partial or total absence of the coronary sinus. Xie et al. classified UCSS into three categories: type I (total absence of coronary sinus); type II (partial forms with one or more defects in the midportion); and type III (partial forms in the coronary sinus outlet). Each of these categories is subdivided into subgroups A and B, which differ according to the persistence or not of LSVC, respectively. However, there are also mixed forms of types II and III [1].

Subtypes IB, IIB and IIIB correspond to coronary sinus-type atrial septal defects, which are easier to manage surgically. The repair can be done by closing the coronary sinus roof defect (coronary sinus drained to the RA) or by closing the atrial septal defect, leaving the coronary sinus draining into the LA. Throughout this paper we will discuss the surgical management of subtypes IA, IIA and IIIA.

The UCSS in its complete form is also known as Raghib's syndrome [2], and belongs to subtype IA (as classified by

Xie et al.); it is characterized by the LSVC draining into the upper left part of the LA, complete absence of the roof of the coronary sinus and coronary sinus-type atrial septal defect, which means that it is located at the posteroinferior edge of the interatrial septum. UCSS is a rare anomaly, which is accompanied by the absence of the innominate vein in 80—90% of cases [3]. Clinically, desaturation and left-right shunt are produced, and it is usually associated with other cardiac abnormalities, such as partial or complete atrioventricular channel, tetralogy of Fallot, interventricular communication, heterotaxia syndrome and double outlet right ventricles, among others [3,4]. Some complications, such as embolism or brain abscess, have been described. Therefore, surgical repair is required after a diagnosis of Raghib's syndrome [1].

Heterotaxy syndrome

Heterotaxy, visceral heterotaxy or heterotaxy syndrome can be defined as an abnormality in the distribution of the internal thoracoabdominal organs along the left-right body axis. By convention, heterotaxy does not include patients with normal distribution along the left-right axis (situs solitus) or those with a reversed mirror image of the normal position of the thoracoabdominal organs (situs inversus totalis) [5].

Left or right atrial appendage isomerisms are subtypes of heterotaxy, where appendages of both atria have the same morphology as the LA or RA. Table 1 shows the most frequent findings in both heterotaxy subtypes [6,7]. These abnormalities are of great interest during the surgical repair of heterotaxy-associated cardiopathies. Particularly noteworthy is the large number of anomalies that can appear in the pulmonary and systemic venous return. In both isomerisms, there is the frequent existence of the LSVC and RSVC, or a persistent LSVC in the absence of the RSVC, draining into the roof of the LA.

Right atrial appendage isomerism is associated with total anomalous pulmonary venous connection (TAPVC). TAPVC is usually extracardiac, although the pulmonary veins can be connected to one atrium or both atria. In other cases, the pulmonary veins can drain into a single atrium through a fibrous non-muscular collector, which can be obstructive (cor triatriatum). In the normal cardiac venous system, it is possible to distinguish a circumflex component within the atrioventricular groove and a longitudinal component coursing on the epicardial surface of the ventricular mass. The coronary sinus, which belongs to the circumflex component, is formed by the union of the greater cardiac vein and the oblique vein of Marshall. In a sample of 99 specimens with right atrial appendage isomerism, Uemura et al. did not find elements of the circumflex component, including the

Table 1 Most frequent findings in left and right atrial appendage isomerism [6,7].

Findings	Atrial appendage isomerism	
	Right	Left
Conduction system	Usually two sinusal nodes; two AV nodes can be present	In most cases, the sinusal node is absent; when it is present, it usually has an unusual position and is hypoplastic
Interatrial septal defects	Usually a single atrium	A single atrium, ASD ostium primum or ASD ostium secundum may appear
AV connection	Complete AV channel with a common AV valve; a univentricular connection can appear in 50—75% of cases	A normal AV valve in 50% of cases; rarely, a complete AV channel with a common AV valve; a biventricular connection is usually present
Interventricular septal defects	Interventricular communication (100% of cases), commonly related to complete AV channel	Very frequent but not constant; when present, the localization is variable
Arterial-ventricular coupling	Discordant or double outlet in the RV with pulmonary stenosis or atresia	Usually concordant and associated with subaortic stenosis and aortic coarctation; double outlet in the RV and pulmonary stenosis or atresia can be present
Systemic venous connection anomalies	Frequently, both the LSVC and RSVC, or a persistent LSVC in the absence of the RSVC, draining into the roof of the LA; inferior vena cava usually normal	Frequently, both the LSVC and RSVC, or a persistent LSVC in the absence of the RSVC, draining into the roof of the LA or coronary sinus; the most frequent venous drainage anomaly is interruption of the intrahepatic segment of the inferior cava vein with azygos-hemiazygos continuation
Pulmonary venous connection anomalies	Extracardiac TAPVC is frequent; pulmonary veins can drain into a single atrium through a fibrous non-muscular collector that can be obstructive (cor triatriatum)	The drainage of pulmonary veins by a collector that can be obstructive (cor triatriatum) or into the ipsilateral atrium

ASD: atrial septal defect; AV: atrioventricular; LA: left atrium; LSVC: left superior vena cava; RSVC: right superior vena cava; RV: right ventricle; TAPVC: total anomalous pulmonary vein connection.

coronary sinus [8]. In fact, the coronary sinus is universally absent in the setting of right atrial appendage isomerism [5,9], so LSVC draining into the LA is not generally considered as UCSS.

In left atrial appendage isomerism, extracardiac TAPVC is infrequent, but not unusual. The most frequent venous drainage anomaly is interruption of the intrahepatic segment of the inferior cava vein with azygos or hemiazygos continuation. Drainage of the LSVC through the coronary sinus can only be produced in cases of left atrial appendage isomerism. Because of this, if the LSVC drains into the LA roof it could be interpreted as an unroofed coronary sinus [5,6]. However, these conceptual differences are irrelevant during the biventricular repair of cardiopathies associated with heterotaxy syndromes, because if the LSVC is draining into the LA, the repair process implies a rerouting and/or disconnection-reconnection to the RA or RSVC. However, if there is communication between the RSVC and the LSVC, such as through the innominate vein, ligation of the section and suture of the distal segment of the LSVC would be enough to re-establish a normal systemic venous return.

Diagnosis

The diagnosis is usually made using echocardiographic techniques (Fig. 1). The coronary sinus is generally identified in the parasternal long-axis and apical four-chamber view, with a slightly backward deviation. The LSVC is usually identified along the suprasternal axis. The diagnosis is confirmed by identification of the LSVC without dilated coronary sinus. Occasionally, a channel-like drainage structure between the left atrial appendage basement and the left superior pulmonary vein can be found in the suprasternal short axis [1]. When some diagnostic doubts persist, right heart contrast echocardiography or echocardiography with an air-bubble study must be used. In this technique, agitated saline serum is injected into a peripheral vein from the left superior arm. Thus, if the LSVC is connected to the LA, the bubbles are found in the LA before the RA. This technique is especially useful when there is partial absence of coronary sinus roof. In subtype III, an atrial septal defect can be found in the usual location of the coronary sinus ostium [1]. Contrast computed tomography scans and magnetic resonance imaging can help during exploration of the intracardiac anatomy, in identification of dysfunction in the systemic or pulmonary venous return and to complete the diagnosis of thoracoabdominal anomalies in heterotaxy syndromes [10].

Surgical techniques

Numerous intracardiac and extracardiac surgical techniques have been described (Fig. 2 and Fig. 3). A thorough knowledge of all of these techniques is required for the appropriate selection to be made, because it depends on

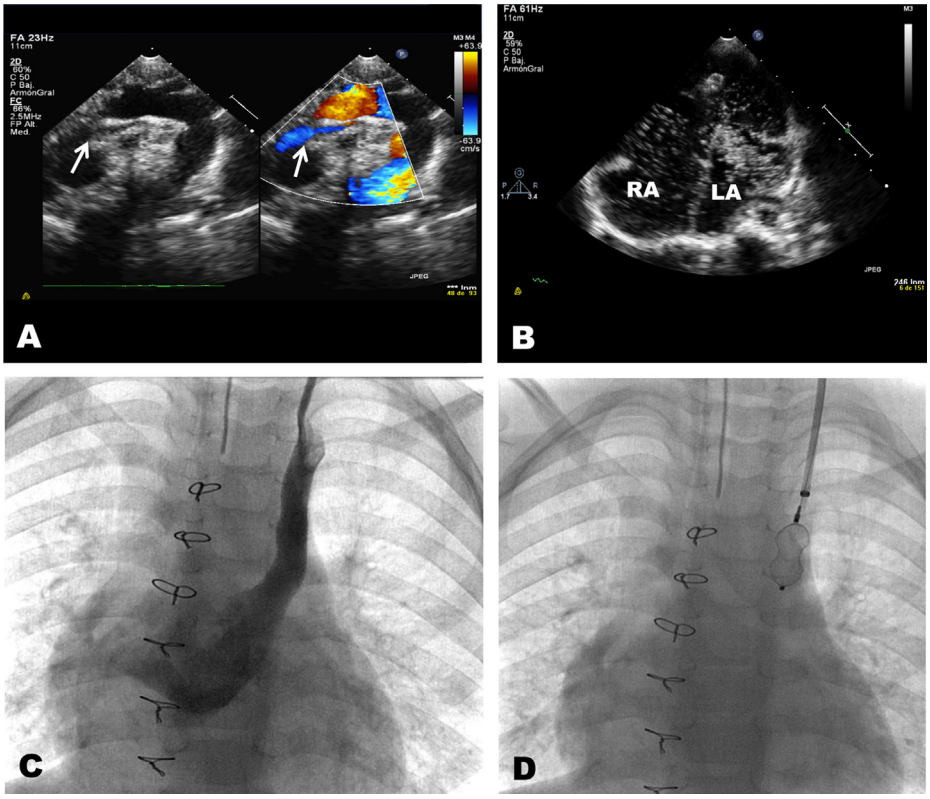

Figure 1. Diagnostic images. A. Transthoracic suprasternal long-axis echocardiography, showing the left superior vena cava (LSVC) draining into the right superior vena cava behind the aorta (white arrow). B. Transthoracic echocardiography apical four-chamber view during an air-bubble study, showing that after injection of agitated serum through a vein in the left arm, the bubbles first enter the left atrium (LA). C and D. Angiographic images showing the LSVC draining into the roof of the LA (C), and the releasing of the percutaneous closure device (D). RA: right atrium.

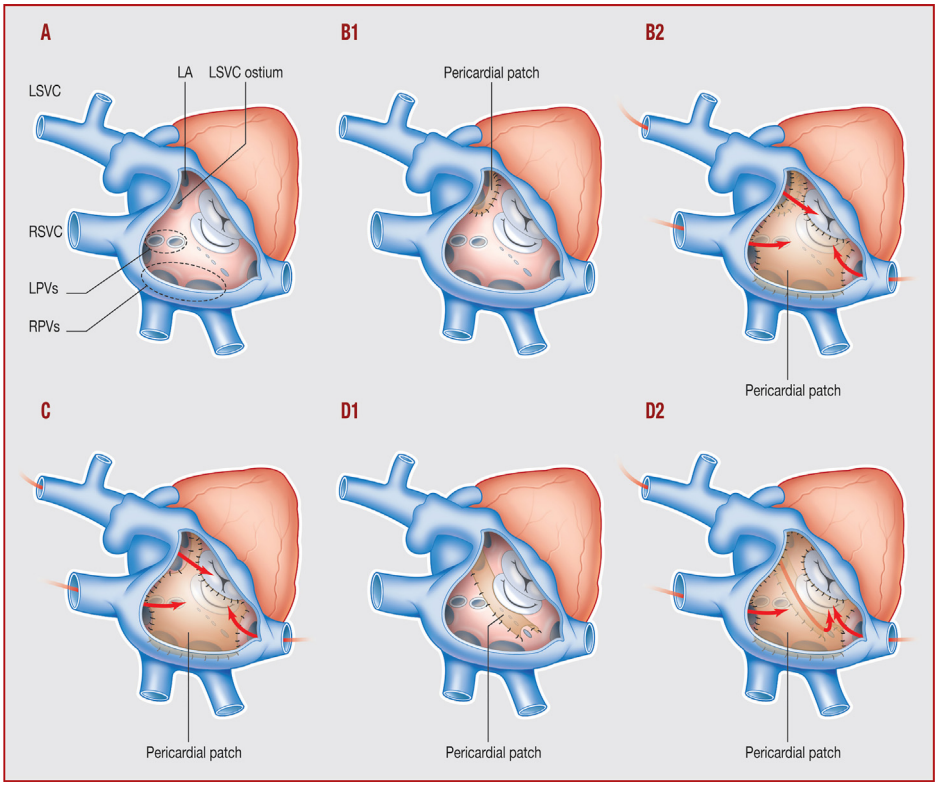

Figure 2. Schematic representations of intracardiac repair techniques. A. Anatomy of unroofed coronary sinus syndrome. B. Two-patch repairing technique: the first patch reroutes the flux from the left superior vena cava (LSVC) to the interatrial septum plane (B1); the second patch closes the atrial septal defect (B2). C. Single-patch repairing technique: the patch is used to redirect the LSVC flux to the tricuspid valve, and to close the atrial septal defect. D. Two-patch repairing technique for reconstruction of the coronary sinus: the first patch creates a new coronary sinus that redirects the flux from the LSVC to the tricuspid valve (D1); the second patch closes the atrial septal defect (D2). LA: left appendage; LPVs: left pulmonary veins; RPVs: right pulmonary veins; RSVC: right superior vena cava.

important characteristics, such as the patient anatomy, age and concomitant cardiopathies and abnormalities.

Within the group of intracardiac techniques, rerouting of the LSVC and coronary sinus must be highlighted. Rerouting of the LSVC implies atrial septation with a single patch [11—14] or two patches [4,15] of the pericardium or synthetic materials. Reroofing of the coronary sinus is focused on the creation of a new coronary sinus using the pericardium, synthetic materials or even autologous left atrial tissue [3,11,16]. Then, a new atrial septation must be created with a new patch.

Regarding extracardiac techniques, first, the atrial septal defect has to be closed or the single atrium has to be septated, according to the specific case. The different extracardiac techniques that can be selected are: simple ligation of the LSVC or LSVC occlusion in the catheterization laboratory, independent of the presence or not of the innominate vein, measuring and controlling the blood pressure into the vein after clamping and taking care not to exceed 15 mmHg [4] or 30 mmHg [17]; terminolateral anastomosis between the LSVC and RSVC in front of [18] or behind [19] the aortic arch, or otherwise using a left atrial appendage flap [20]; anastomosis of the LSVC to the right atrial appendage, directly [19] or interposing left atrial tissue [21,22], a PTFE graft [23] or a venous homograft [24]; terminolateral anastomosis between the LSVC and left pulmonary artery [19]; and when a vestige of the innominate vein is present, it can be extended in size using an autologous pericardial patch or other material [25].

Surgical series

In the period from 1998 to 2016, 11 surgical procedures in 10 patients were performed by our surgical team (Table 2). Six patients had been diagnosed with heterotaxy syndrome and four with UCSS. Some intraoperative images are shown in Fig. 4. In the patients with heterotaxy syndrome, an intracardiac repair procedure (single-patch or two-patch rerouting) was used, with the exception of patient 6, in whom ligation of the LSVC was done because of the presence of innominate vein and RSVC. Among the patients with UCSS, a surgical procedure was selected, considering patient characteristics. Extracardiac repairing was done in two patients: disconnection-reconnection to the RA (patient 9) or to the RSVC via a retroaortic approach (patient 8). Patient 7 was treated by two-patch intracardiac rerouting, and patient 10 was diagnosed after surgical repair of tetralogy of Fallot. In this case, because patient 10 presented innominate vein and RSVC, we performed percutaneous embolization of the LSVC using a 16 mm Amplatzer® vascular plug (St. Jude Medical, St. Paul, MN, USA).

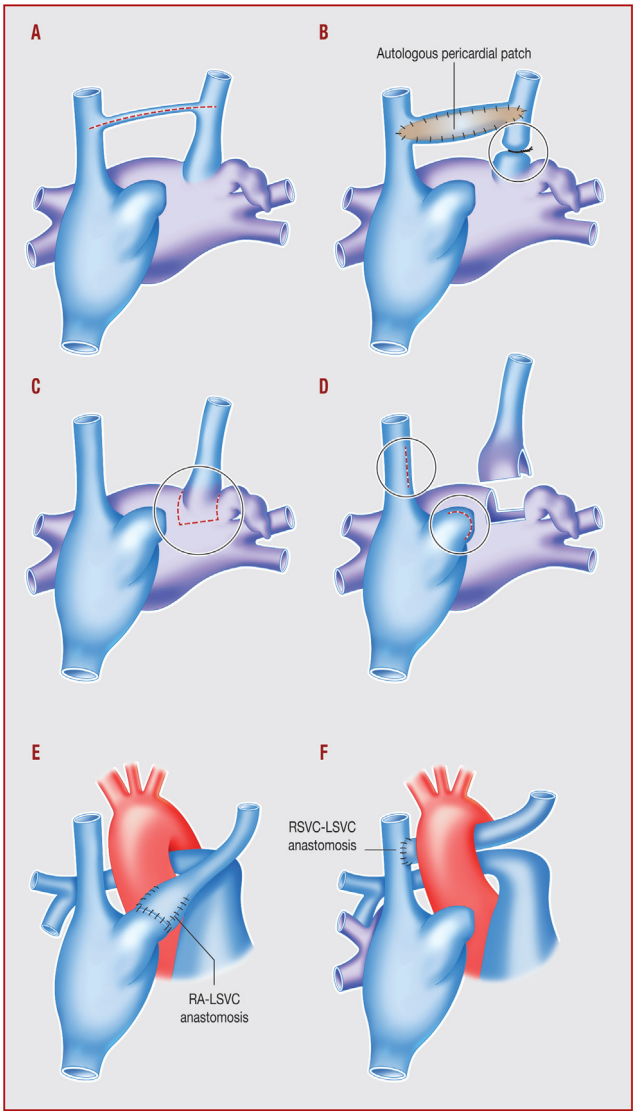

Figure 3. Schematic representations of extracardiac repair techniques. A. Persistent left superior vena cava (LSVC) draining into the left atrium roof, with a hypoplastic innominate vein. B. The extracardiac repair can be performed by expansion of the innominate vein using an autologous pericardial patch in addition to ligation of LSVC drainage into the left atrium roof. C. Persistent LSVC draining into the left atrium roof without hypoplastic innominate vein; the circle shows the surgical incision lines for disconnection of the LSVC from the left atrium roof. D and E. Then, the disconnected LSVC (D) can be reconnected (circles) to the right atrial (RA) appendage (E) or to the right superior vena cava (RSVC) behind the aorta (F).

Patient 3 required a reintervention 36 months after surgery because of a partial dehiscence of the LSVC rerouting patch. For that, we performed reconstruction of the coronary sinus and, later, closure of the atrial septal defect, using a heterologous pericardial patch in both interventions. The mean follow-up for the overall cohort was 65.0 months (range 4.0—212.0 months). Two patients experienced complications, although no reintervention was needed during the conduction of this study: patient 1 with moderate left superior pulmonary vein stenosis; and patient 4 with light supramitral stenosis.

Overall assessment of the surgical procedures

Embryologically, the persistence of the LSVC is caused by the absence of atrophy of the left common cardinal vein, independent of the presence of heterotaxy syndrome or UCSS [26]. As stated before, because the coronary sinus is absent in the setting of right atrial appendage isomerism, LSVC draining into the LA is not generally considered as UCSS [5,8,9]. In left atrial appendage isomerism, if the LSVC exists, it would drain into the coronary sinus. Therefore, if the LSVC drains into the roof of LA, the diagnosis would be UCSS.

Independent of the clinical entity, biventricular repair is directed to the rerouting or disconnection-reconnection of the LSVC to the RA or RSVC. As we have stated before, there are numerous surgical techniques for this purpose, which can be divided into two main groups: extracardiac and intracardiac. Because of potential complications in intracardiac techniques, such as stenosis in the pulmonary veins, stenosis in the LSVC tunnel or supramitral stenosis [19,25,27], most authors prefer extracardiac techniques. These techniques are easier to apply, and reconnection of the LSVC to the RSVC or right atrial appendage can be realized after intra-atrial communication closure or after single atrium septation, according to each individual case, thereby diminishing aortic clamping time [19,22]. Reddy et al. highlighted the terminolateral anastomosis between the LSVC and retroaortic RSVC, arguing that this represents a more natural pathway, closer to that seen when a retroaortic innominate vein exists. This technique was employed for patient 8, in whom other extracardiac techniques (such as left atrial roll) could not be used because of possible compression of the LSVC by the surrounding great vessels. Consequently, we performed a retroaortic anastomosis to avoid compression [19].

There are patients in whom extracardiac techniques are especially difficult, as a result of the distance between the LSVC, once sectioned, and the RSVC or right atrial appendage. In these cases, a useful option is the interposing of a polytetrafluoroethylene (PTFE) conduit for the connection. In infants and young children, the duct has to be of sufficient size (at least 8 mm in diameter) to avoid, as far as possible, its future replacement [23]. Patient 7 was a 4-month-old baby, who was treated surgically for the closure of an interventricular communication. Intrasurgical examination revealed a persistent LSVC totally unroofed (Raghib's syndrome). The presence of pulmonary trunk dilatation along with absence of dilatation of the right atrial appendage led us to discard an extracardiac repair, because of its complexity, and perform an intracardiac repair with two patches.

In heterotaxy syndrome, single atrium association is rather usual. In these cases, septation of the single atrium for biventricular repairing is necessary, and it can be done using single-patch or two-patch procedures. The inclusion of the LSVC hole in the septation patch does not usually increase the difficulty. For all these issues, intracardiac repair is the usual treatment for single atrium [14].

Table 2 Characteristics of surgical series.

Patient	Age (months)	Diagnosis	Type of surgery	Timing of diagnosis	Previous operation	Reoperation	Follow-up (months)	Complications
1	20	Heterotaxy; left isomerism; complete AVSD; PAPVC	Intracardiac: rerouting (one patch)	Presurgery	No	No	212	Moderate PV stenosis
2	39	Heterotaxy; left isomerism; complete AVSD; single atrium; no RSVC	Intracardiac: rerouting (one patch)	Presurgery	PAB	No	136	No
3	40	Heterotaxy; left isomerism; transitional AVSD; single atrium	Intracardiac: rerouting (two patches)	Intrasurgery	PAB	Yes: residual ASD; rerouting of CS	109	No
4	32	Heterotaxy; left isomerism; transitional AVSD; single atrium; Ellis-van Creveld syndrome	Intracardiac: rerouting (two patches)	Presurgery	No	No	77	Mild supramitral stenosis
5	153	Heterotaxy; left isomerism; transitional AVSD; single atrium; no RSVC	Intracardiac: rerouting (one patch)	Presurgery	No	No	33	No
6	32	Heterotaxy; right isomerism; complete AVSD; DORV; PA; TAPVC	Surgical closure of LSVC (innominate vein)	Presurgery	MBTS 3.5 mm; MBTS 5 mm	No	29	No
7	4	VSD; UCSS type IA	Intracardiac: rerouting (two patches)	Intrasurgery	No	No	22	No
8	88	VSDs; UCSS type IIIA	Extracardiac: retroaortic LSVC-RSVC anastomosis	Postsurgery	PAB; closure of VSDs (hybrid approach)	No	8	No
9	552	UCSS type IA	Extracardiac: LSVC-RAA anastomosis	Presurgery	No	No	20	No
10	53	TOF; UCSS type IIA	Percutaneous closure of LSVC (innominate vein)	Postsurgery	Complete repair; relief of residual RVOTO	No	4	No

ASD: atrial septal defect; AVSD: atrioventricular septal defect; CS: coronary sinus; DORV: double outlet right ventricle; LSVC: left superior vena cava; MBTS: modified Blalock-Taussig shunt; PA: pulmonary atresia; PAPVC: partial anomalous pulmonary vein connection; PV: pulmonary vein; PAB: pulmonary artery banding; RAA: right atrial appendage; RSVC: right superior vena cava; RVOTO: right ventricular outflow tract obstruction; TAPVC: total anomalous pulmonary vein connection; TOF: tetralogy of Fallot; UCSS: unroofed coronary sinus syndrome; VSD: ventricular septal defect; VSDs: multiple ventricular septal defects.

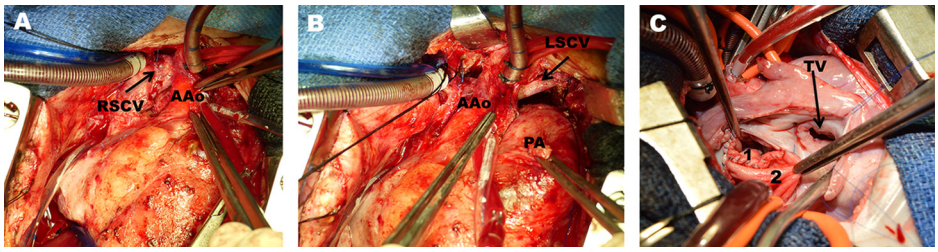

Figure 4. Intraoperative images. A and B. Extracardiac repair by disconnection-reconnection of the left superior vena cava (LSVC) to the right superior vena cava (RSVC) behind the aorta. A. Terminolateral anastomosis between both veins (black arrow). B. The LSVC course from its origin to the RSVC after surgery. C. Two-patch intracardiac repair in Raghib's syndrome: the image shows the opened right atrium (RA) and both heterologous pericardial patches; the first patch (1) reroutes the flux from the LSVC to the RA; the second patch (2) closes the atrial septal defect. Aao: ascending aorta; PA: pulmonary artery; TV: tricuspid valve.

Conclusions

Persistent LSVC draining into the LA is a systemic venous drain anomaly that includes different clinical entities; numerous techniques can be useful for their reparation. Depending on the clinical entity to be treated, the age of the patient, the anatomical constitution and the associated cardiopathies, a specific technique may be indicated.

Sources of funding

None.

Acknowledgments

This paper is related to the PhD thesis of J. M. Aguilar, who is enrolled in the PhD programme in Biomedicine, University of Granada, Spain.

Disclosure of interest

The authors declare that they have no competing interest.

References

[1] Xie MX, Yang YL, Cheng TO, et al. Coronary sinus septal defect (unroofed coronary sinus): echocardiographic diagnosis and surgical treatment. Int J Cardiol 2013;168:1258—63.
[2] Raghib G, Ruttenberg HD, Anderson RC, Amplatz K, Adams Jr P, Edwards JE. Termination of left superior vena cava in Left atrium atrial septal defect, and absence of coronary sinus; a developmental complex. Circulation 1965;31:906—18.
[3] Quaegebeur J, Kirklin JW, Pacifico AD, Bargeron Jr LM. Surgical experience with unroofed coronary sinus. Ann Thorac Surg 1979;27:418—25.
[4] Ootaki Y, Yamaguchi M, Yoshimura N, Oka S, Yoshida M, Hasegawa T. Unroofed coronary sinus syndrome: diagnosis, classification, and surgical treatment. J Thorac Cardiovasc Surg 2003;126:1655—6.
[5] Jacobs JP, Anderson RH, Weinberg PM, et al. The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy. Cardiol Young 2007;17(Suppl 2):1—28.
[6] Kirklin JW, Barratt-Boyes BG. Unroofed coronary sinus. In: Kirklin JW, Barratt-Boyes BG, editors. Cardiac Surgery. 2nd ed New York: Churchill Livingston; 1993. p. 1383—467.
[7] Park MK. Cardiopatías congénitas cianosantes. In: Park MK, editor. Cardiología Pediátrica. 5th ed Barcelona S.L: Elsevier España; 2008. p. 215—301.
[8] Uemura H, Ho SY, Anderson RH, et al. The surgical anatomy of coronary venous return in hearts with isomeric atrial appendages. J Thorac Cardiovasc Surg 1995;110:436—44.
[9] Anderson RH, Spicer DE, Loomba R. Is an appreciation of isomerism the key to unlocking the mysteries of the cardiac findings in heterotaxy? J Cardiovasc Dev Dis 2018:5.
[10] Cohen MS, Anderson RH, Cohen MI, et al. Controversies, genetics, diagnostic assessment, and outcomes relating to the heterotaxy syndrome. Cardiol Young 2007;17(Suppl 2):29—43.
[11] de Leval MR, Ritter DG, McGoon DC, Danielson GK. Anomalous systemic venous connection. Surgical considerations. Mayo Clin Proc 1975;50:599—610.
[12] Komai H, Naito Y, Fujiwara K. Operative technique for persistent left superior vena cava draining into the left atrium. Ann Thorac Surg 1996;62:1188—90.
[13] Miller GA, Ongley PA, Rastelli GC, Kirklin JW. Surgical correction of total anomalous systemic venous connection: report of case. Mayo Clin Proc 1965;40:532—8.
[14] Vodiskar J, Clur SA, Hruda J, Bokenkamp R, Hazekamp MG. Left atrial isomerism: biventricular repair. Eur J Cardiothorac Surg 2010;37:1259—63.
[15] Sand ME, McGrath LB, Pacifico AD, Mandke NV. Repair of left superior vena cava entering the left atrium. Ann Thorac Surg 1986;42:560—4.
[16] Rastelli GC, Ongley PA, Kirklin JW. Surgical correction of common atrium with anomalously connected persistent left superior vena cava: report of case. Mayo Clin Proc 1965;40:528—32.
[17] Zimand S, Benjamin P, Frand M, Mishaly D, Smolinsky AK, Hegesh J. Left superior vena cava to the left atrium: do we have to change the traditional approach? Ann Thorac Surg 1999;68:1869—71 [discussion 71-2].
[18] Ugaki S, Kasahara S, Fujii Y, Sano S. Anatomical repair of a persistent left superior vena cava into the left atrium. Interact Cardiovasc Thorac Surg 2010;11:199—201.
[19] Reddy VM, McElhinney DB, Hanley FL. Correction of left superior vena cava draining to the left atrium using extracardiac techniques. Ann Thorac Surg 1997;63:1800—2.
[20] Palacios-Macedo AX, Fraser Jr CD. Correction of anomalous systemic venous drainage in heterotaxy syndrome. Ann Thorac Surg 1997;64:235—7 [discussion 7-8].
[21] Shumacker Jr HB, King H, Waldhausen JA. The persistent left superior vena cava. Surgical implications, with special reference to caval drainage into the left atrium. Ann Surg 1967;165:797—805.

[22] Vargas FJ. Reconstructive methods for anomalous systemic venous return: surgical management of persistent left superior vena cava. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2008:31—8.
[23] Gontijo B, Fantini FA, de Paula e Silva JA, Barbosa JT, Vrandecic MO, Masci Mda G. The use of PTFE graft to correct anomalous drainage of persistent left superior vena cava. J Cardiovasc Surg (Torino) 1990;31:815—7.
[24] Alexi-Meskishvili V, Dahnert I, Beyer E, Hetzer R. Successful total correction of complete atrioventricular canal, total anomalous pulmonary venous drainage and unroofed coronary sinus in an infant. Eur J Cardiothorac Surg 1999;15:95—6.
[25] van Son JA, Hambsch J, Mohr FW. Repair of complex unroofed coronary sinus by anastomosis of left to right superior vena cava. Ann Thorac Surg 1998;65:280—1.
[26] Ramos N, Fernandez-Pineda L, Tamariz-Martel A, Villagra F, Egurbide N, Maitre MJ. Absent right superior vena cava with left superior vena cava draining to an unroofed coronary sinus. Rev Esp Cardiol 2005;58:984—7.
[27] Gandy K, Hanley F. Management of systemic venous anomalies in the pediatric cardiovascular surgical patient. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2006:63—74.