ESC European Society of Cardiology

European Heart Journal - Case Reports (2022) **6**, 1–5
https://doi.org/10.1093/ehjcr/ytac382

CASE REPORT
Congenital heart disease

Case report of a left superior vena cava to left atrial connection treated with percutaneous covered stent placement

Phuoc Duong[1], Vasileios Papaioannou[2], Sarah Moharam-Elgamal[2], and Sok-Leng Kang [1,2]*

[1]Department of Pediatric Cardiology, Alder Hey Children's Hospital, Liverpool, UK; and [2]Department of Adult Congenital Heart Disease, Liverpool Heart and Chest Hospital, Liverpool, UK

Received 23 March 2022; first decision 20 April 2022; accepted 13 September 2022; online publish-ahead-of-print 3 October 2022

Background Persistent left superior vena cava (LSVC) with connection to the left atrium (LA) is a rare anomaly with serious clinical implications. Depending on the direction of flow through the intracardiac shunt, clinical presentation varies from cyanosis or paradoxical embolism to overt right heart failure.

Case summary A 26-year-old man with repaired ventricular septal defect (VSD) during infancy presented with symptoms of progressive exercise intolerance. Cardiac imaging revealed a large defect at the posterior wall of the LSVC above its entry to the coronary sinus (CS), connecting the LSVC with the LA and resulting in diversion of pulmonary venous return to the CS. All pulmonary veins connect normally to the LA. The large left-to-right intracardiac shunt led to significant right ventricular dilation and tricuspid regurgitation. He underwent successful anatomical repair with transcatheter implantation of covered stent from LSVC to the CS, redirecting pulmonary venous return to the LA. At 1 year follow up, his exercise capacity had improved, and cardiac imaging showed complete seal of the LSVC defect without obstruction to pulmonary venous return.

Discussion Our case is the first to our knowledge to report this unusual anatomic variant of LSVC to LA connection, and complete repair by transcatheter treatment. Previous case reports of other forms of LSVC to LA connection were treated with surgery or device occlusion without reconnection of LSVC. This case highlights the efficacy and safety of innovative percutaneous techniques in the management of complex congenital heart lesions. Meticulous procedural planning through 3D modelling and simulation is vital to mitigate the risks of these innovative procedures.

Keywords Percutaneous vascular intervention • Left superior vena cava • Anomalous venous return • Case report

ESC Curriculum 9.7 Adult congenital heart disease • 2.4 Cardiac computed tomography • 2.1 Imaging modalities

Learning points

- Despite an extremely rare anomaly, LSVC to LA connection should be suspected in the presence of cyanosis, paradoxical embolism, or right heart volume overload without other intracardiac shunt.
- In the anatomical variant of LSVC to LA connection in our case, percutaneous treatment by covered stent is a feasible option.
- Pre-procedure planning with 3D modelling and simulation facilitates the application of novel interventional techniques in the treatment of complex congenital heart lesions.

* Corresponding author. Tel: +44 416 813 6141, Fax: 0044 (0) 151 252 5643, Email: sokleng.kang@alderhey.nhs.uk
Handling Editor: Filippo Puricelli
Peer-reviewers: Jan Henzel; Parag Bawaskar
Compliance Editor: Gal Tsaban
Supplementary Material Editor: Katharine Kott

Introduction

Persistence of left superior vena cava (LSVC) is the most common variant of systemic venous drainage with an incidence of ~0.5% in the general population and up to 10% of patients with congenital heart disease.[1,2] It results from a failure of obliteration of the left common cardinal vein in foetal life and typically drains into the right atrium (RA) via the coronary sinus (CS) with no adverse haemodynamic effect. Atypical LSVC connection with the left atrium (LA), however, carries important clinical consequences. Recognized forms of this anomaly include indirect drainage through an unroofed CS or direct termination of the LSVC in the LA.[2] Depending on the direction of flow and shunt volume, clinical presentation varies from cyanosis or paradoxical embolism (right to left shunt) to overt right heart failure (left-to-right shunt), necessitating complex surgical repair.[2–4] Here, we report an unusual case of LSVC to LA connection through a large posterior wall defect of the LSVC as it enters the CS. Diversion of pulmonary venous flow through the LSVC defect into the RA resulted in symptomatic right ventricular dilation and significant tricuspid regurgitation. To our knowledge, this is the first report describing complete anatomical correction of this unique variant with percutaneous covered stent implantation.

Timeline

Age	Event
Infant	Surgical repair of ventricular septal defect (VSD)
18 years	Lost to follow up following discharge from paediatric cardiology services
25 years	Progressive exercise intolerance, fatigability
25 years	Echocardiogram showed severe tricuspid regurgitation and significantly dilated right heart, and LSVC draining into CS. Bubble study through left arm showed bubbles appearing in left heart.
25 years	Cardiac catheterization confirmed atrial level left-to-right shunt, Qp:Qs 3:1
25 years	Cardiac magnetic resonance imaging (MRI) showed large defect of posterior wall of LSVC.
25 years	3D model printed to assess feasibility of transcatheter covered stent correction.
25 years	Discussed in cardiothoracic meeting with consensus for covered stent to seal the LSVC defect
25 years	Customised 80 mm covered CP stent ordered
26 years	Interventional cardiac catheterization with placement of uncovered and covered stents in the LSVC extending to CS.
27 years	No complications at 1-year follow-up post procedure. Symptomatic improvement. Cardiac imaging confirmed good result with unobstructed LSVC flow to CS, unobstructed pulmonary venous return, reduced RV volume and reduced severity of tricuspid regurgitation.

Case summary

A 26-year-old Caucasian man with repaired VSD during infancy who had been lost to follow up, presented with symptoms of progressive exercise intolerance and breathlessness during sporting activities over a 1 year period. He was otherwise generally well with no limitations with daily activities. He has mild scoliosis and no other significant medical history. A thorough physical examination did not yield any abnormal findings. His saturation was 94%. Echocardiography showed good biventricular function a volume overloaded right ventricle (RV) with moderate tricuspid regurgitation. The LSVC drains into a severely dilated CS and right SVC drains into RA. There was no bridging innominate vein. The roof of the CS and atrial septum was intact. Contrast study via the left arm showed microbubbles appearing in the left heart, prompting an MRI which showed a large defect at the posterior wall of the LSVC above its entry to the CS. Pulmonary venous drainage to LA was normal. The LSVC defect connects with the LA, resulting in diversion of pulmonary venous return to the CS. Cardiac catheterization confirmed atrial level shunting with Qp:Qs of 3.2 to 1 and normal pulmonary pressures. After extensive discussions, our group decided that transcatheter implantation of a covered stent from the LSVC to the CS was a viable treatment strategy. A 3D printed model from a prior computed tomography (CT; *Figure 1*) was used to simulate the procedure with a balloon catheter, confirming good seal of the LSVC defect without compromising pulmonary venous return.

The procedure was performed under general anaesthesia. On initial angiography, the LSVC measured 12 × 13 mm and CS measured 32 × 30 mm in orthogonal planes (*Figure 2*). A 24F Gore Dry Seal sheath (W.L. Gore & Associates, Inc., Arizona, USA) was introduced over a veno-venous rail from right femoral vein to the left internal jugular vein to facilitate balloon testing and subsequent stent deployment. A 4F Pigtail catheter was advanced through the CS to the left upper pulmonary vein (LUPV) for continuous pressure monitoring. A 34 mm Amplatzer sizing balloon (Abbott Medical, Minnesota, USA) was inflated in the LSVC until transoesophageal echocardiogram (TOE) confirmed elimination of the shunt. To guide stent placement, an 18 × 45 mm Cristal balloon (Bard, Düsseldorf, Germany) was inflated at the high LSVC, and the balloon waist was used as a landmark (*Figure 2*). A 45 and 60 mm uncovered CP stent mounted on an 18 mm balloon-in-balloon (BIB) catheter was deployed sequentially at the LSVC, serving as 'anchor' stents. Finally, a customized 80 mm covered CP stent mounted on a 40 × 60 mm BIB was deployed to overlap the existing stent assembly with the caudal end of this stent ending within the CS. The caudal end of this 80 mm stent was dilated with a 30 × 60 mm Cristal balloon (Bard, Düsseldorf, Germany) until satisfactory coverage of the shunt was confirmed by TOE and angiography. The LUPV pressure remained constant throughout the procedure.

The patient was discharged the following day on dual antiplatelet therapy. At 1 year follow up, the patient was well and reported symptomatic improvement. Cardiopulmonary exercise test at 3 months post procedure showed improved peak VO_2 of 32 mL/min/kg compared with pre-procedure measurement of 24.1 mL/min/kg. Comparison of pre- and post-procedure CT (performed after 3 months) showed a reduction in right ventricle to left ventricle (RV:LV) volume ratio from 2.1 to 1.3. The CT also confirmed complete coverage of the LSVC defect with no residual shunt and unobstructed

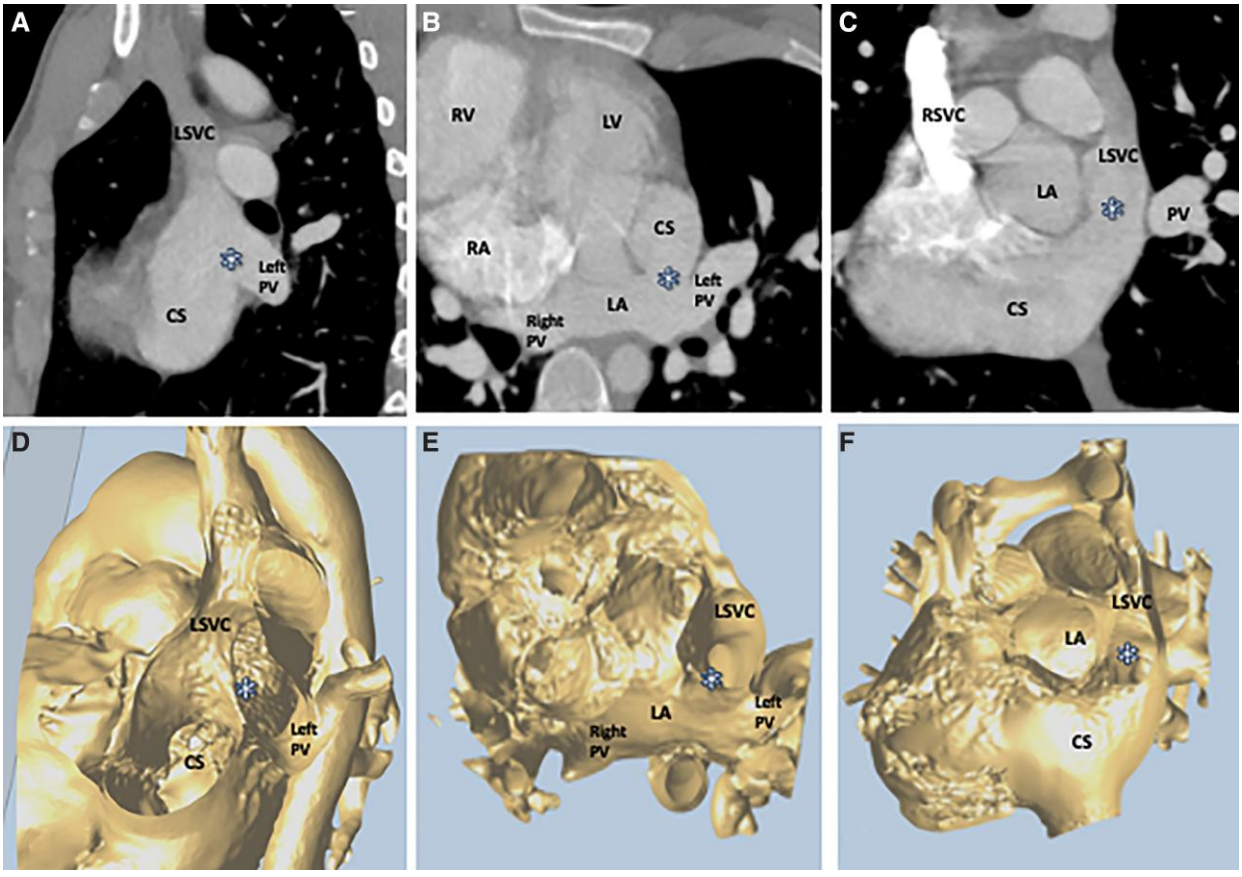

Figure 1 Top panel shows computed tomography orthogonal views of the left superior vena cava defect (*) in multiplanar reformat and bottom panel demonstrates the defect (*) in equivalent crop from the 3D model: (*A* and *D*) sagittal plane, (*B* and *E*) axial plane, and (*C* and *F*) coronal plane. The left superior vena cava drains to the severely dilated coronary sinus and pulmonary veins connect to normal sized left atrium left atrium. There is deficiency of the left superior vena cava posterior wall as it joins the coronary sinus .

pulmonary venous return (*Figure 3*). His most recent echocardiogram showed an improvement in the severity of tricuspid regurgitation.

Discussion

Management of LSVC to LA connection is guided by clinical symptoms and surgical repair remain the mainstay of treatment. When associated with an unroofed CS, various techniques with intracardiac baffling and/or extracardiac re-routing of LSVC to the RA, right superior vena cava, or pulmonary artery have been reported, each with advantages and drawbacks.[4] With complex intra-atrial baffling, complications such as pulmonary vein stenosis, LSVC tunnel obstruction and supra-mitral stenosis are not uncommon.[4–6] In the case of direct LSVC to LA drainage, percutaneous closure or surgical ligation (without reconnection) of the distal segment of the LSVC is an alternative.[3,7–10] The caveat with this approach is that venous hypertension can develop with a risk of cerebral injury when venous collateralization is inadequate, particularly in patients without a bridging innominate vein.[9]

In our patient, the lesion is described as an LSVC defect, as opposed to unroofed CS because the defect is located at the vertical portion of the LSVC as it descends to join the CS. The CS typically has a transverse course in the sulcus between the LA and ventricle.[11] Further, the CS is a product of the tributaries of the greater and smaller cardiac veins.[11] However, at the site of the LSVC defect in this case, the venous structure does not receive any cardiac veins. The unique anatomic variant in our patient was amenable to re-routing and preservation of LSVC return to the RA by transcatheter placement of a covered stent; without sacrificing the systemic venous pathway by device occlusion of the LSVC in previous reports.[4,7,8,10,12] This strategy also avoids the risks of a re-do sternotomy, cardiopulmonary bypass, and attendant surgical morbidities. This concept is similar to the recently reported covered stent correction of sinus venosus atrial septal defect with encouraging early outcomes.[13] In our case, the need for substantial expansion of the distal stent due to significant size mismatch of the LSVC and CS caused concerns for stent embolization, incomplete defect coverage (due to stent foreshortening), and pulmonary venous obstruction. The use of 3D modelling and simulation was crucial to provide thorough assessment of the lesion and impact of balloon/stent deployment on surrounding structures, allowing alteration of techniques or management decisions to minimize periprocedural complications.

Our choice of balloon expandable covered stent over a self-expandable stent graft was based on several considerations. Firstly, balloon expandable stent grafts are typically deployed in thicker wall aortic pathologies and expansion is partly dependent on constraint of the vessel wall. In our case, stent expansion in a vein with a large area of deficient vessel wall may be unpredictable and protrusion towards the unconstraint portion/defect may result in pulmonary venous

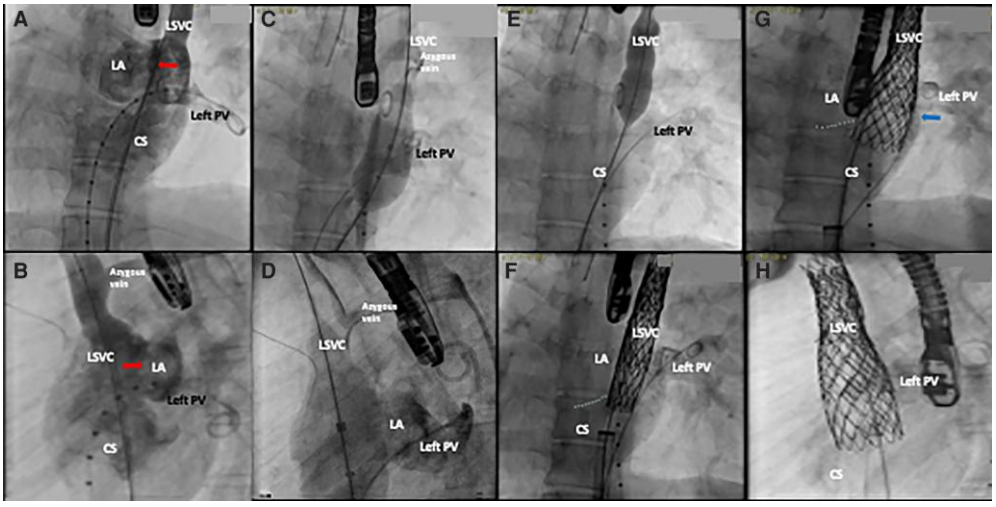

Figure 2 Angiography depicting procedural steps and technical considerations for safe deployment of stents. Baseline (*A*) anteroposterior and (*B*) lateral angiogram of the left superior vena cava. The arrow indicate left superior vena cava posterior wall defect with immediate filling of left atrium. The pigtail catheter traverses the coronary sinus to the left pulmonary vein through the defect. (*C*) Anteroposterior and (*D*) lateral left superior vena cava angiogram following test occlusion of the defect with simultaneous pulmonary vein angiography showed unobstructed pulmonary venous return and complete seal of defect. (*E*) A semi-compliant balloon is used to determine anchorage point for first stent. (*F*) After deploying the anchoring superior stent, subsequent stents were the positioned to overlap within the prior stent to cover the entire defect. The distal tip of the delivery sheath is positioned in the coronary sinus during deployment to serve as a 'buttress' if required. The dotted line indicates the roof of the coronary sinus. (*G* and *H*) The distal portion of the stent was dilated until adequate seal was achieved. Contrast injection in the left pulmonary vein showed unobstructed flow into left atrium and minimal residual flow around the edge of the stent into the coronary sinus (arrow).

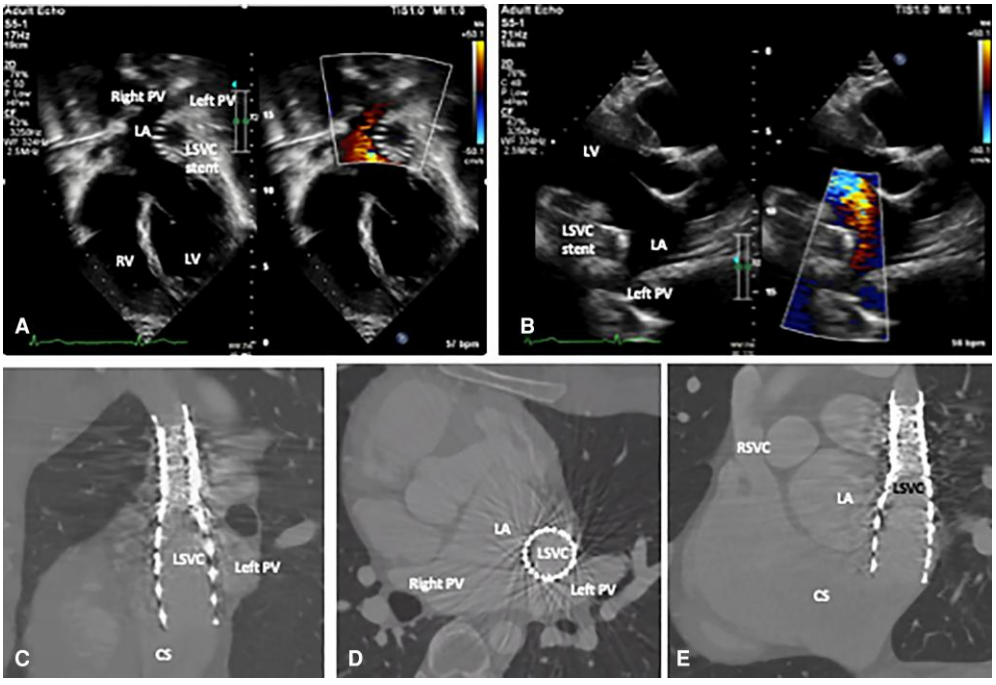

Figure 3 Follow-up echocardiography and computed tomography imaging at 3months follow up. Echocardiographic (*A*) apical four-chamber view and (*B*) parasternal short-axis view showing unobstructed pulmonary vein flow into the left atrium around the stent. Bottom panel shows computed tomography multiplanar reformat in (*C*) sagittal, (*D*) axial, (*E*) coronal view of the left superior vena cava stent completely sealing the defect and extending minimally into the coronary sinus. The computed tomography axial plane (*C*) showed widely patent pulmonary venous drainage into the left atrium posterior to the stented left superior vena cava.

obstruction. Secondly, the anchoring pins for active fixation of self-expandable stent grafts have been reported to cause vessel perforation to the SVC and surrounding structures such as the aorta.[14] In contrast, balloon expandable stents provide reliable expansion to an expected diameter and can be gradually expanded in our case until elimination of shunt is achieved. Currently, the 10 zig covered CP stent can be custom-made in lengths of 6–11 cm but only dilatable to 34 mm in diameter with moderate shortening at diameters >28 mm. The covered CP stent used in this case was manufactured in 12 zigs to provide sufficient radial strength at predicted maximal expansion to 40 mm. There is also theoretically less foreshortening of the 12-zig stent at larger diameters, but the increased stent profile required a larger delivery sheath size of 22F (personal communication with the manufacturer).

Conclusion

While a rare entity, one should always remain cognizant of the possibility of LSVC to LA connection in the presence of cyanosis or paradoxical embolism or right heart volume overload without other intracardiac shunts. This case highlights an increasing trend in the management of complex congenital heart lesions with innovative percutaneous techniques. Notably, thorough pre-procedure planning with 3D modelling and simulation is crucial for success of such innovative procedures.

Lead author biography

Sok-Leng Kang is a congenital cardiac interventionist at AlderHey Children's Hospital and Liverpool Heart and Chest Hospital, Liverpool, United Kingdom.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Acknowledgements

The authors would like to thank the adult congenital cardiologists, cardiac catheterization team (Liverpool Heart and Chest Hospital), Dr John Thomson and Dr Jamie Bentham (Leeds General Infirmary) for their collaboration and assistance in this case.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for the submission and publication of this case, including images, has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: None declared.

References

1. Irwin RB, Greaves M, Schmitt M. Left superior vena cava: revisited. *Eur Heart J Cardiovasc Imaging* 2012;**13**:284–291.
2. Azizova A, Onder O, Arslan S, Ardali S, Hazirolan T. Persistent left superior vena cava: clinical importance and differential diagnoses. *Insights Imaging* 2020;**11**:110.
3. Hutyra M, Skala T, Sanak D, Novotny J, Köcher M, Taborsky M. Persistent left superior vena cava connected through the left upper pulmonary vein to the left atrium: an unusual pathway for paradoxical embolization and a rare cause of recurrent transient ischaemic attack. *Eur J Echocardiogr* 2010;**11**:E35.
4. Aguilar JM, Rodriguez-Serrano F, Ferreiro-Marzal A, Esteban-Molina M, Gabucio A, Garcia E, Boni L, Garrido JM. Left superior vena cava draining into the left atrium: clinical entities, diagnosis and surgical treatment. *Arch Cardiovasc Dis* 2019;**112**:135–143.
5. 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**:1800–1802.
6. 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**:280–281.
7. Tomar M. Percutaneous device closure of persistent left superior vena cava connecting to the left atrium with intact coronary sinus: a rare entity. *Images Paediatr Cardiol* 2017;**19**:1–8.
8. Bugami SA, Althobaiti M, Momenah T, Alrahimi J, Kashkari WA. Percutaneous intervention of a persistent left superior vena cava draining into left pulmonary vein and coarctation of the aorta. *Cardiol Res* 2016;**7**:157–160.
9. 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**:1869–1871; discussion 71-2.
10. Troost E, Gewillig M, Budts W. Percutaneous closure of a persistent left superior vena cava connected to the left atrium. *Int J Cardiol* 2006;**106**:365–366.
11. Habib A, Lachman N, Christensen KN, Asirvatham SJ. The anatomy of the coronary sinus venous system for the cardiac electrophysiologist. *Europace* 2009;**11**:v15–v21.
12. Ylanen K, Litwin L, Ojala T, Puntila J, Kalajoki-Helmio T, Pihkala J. Left superior vena cava draining to left atrium with partially anomalous pulmonary venous connection and left-to-right shunt - multimodality imaging and percutaneous treatment. *Postepy Kardiol Interwencyjnej* 2018;**14**:312–313.
13. Rosenthal E, Qureshi SA, Jones M, Butera G, Sivakumar K, Boudjemline Y, Hijazi Z, Almaskary S, Ponder R, Salem M, Walsh K, Kenny D, Hascoet S, Berman D, Thomson J, Vettukattil J, Zahn E. Correction of sinus venosus atrial septal defects with the 10 zig covered Cheatham-platinum stent - an international registry. *Catheter Cardiovasc Interv* 2021;**98**:128–136.
14. Yalamanchi R, Sivaprakasam MC, Janke RVR, Chandrasekharan K, Sadhasivam VS, Showkathali R. Unanticipated complication of transcatheter correction of superior sinus venosus atrial septal defect. *J Cardiol Cases* 2022;**25**:99–102.