

Unroofed Coronary Sinus Without Persistent Left Superior Vena Cava Presenting as an Atrial Septal Defect: A Case Report

Syeda Maah Amir

syedamaah.amir@scholar.aku.edu

Aga Khan University Hospital

Laiba Khalid

Aga Khan University Hospital

Saleem Akhtar

Aga Khan University Hospital

Shahid Ahmed Sami

Aga Khan University Hospital

Waris Ahmad

Aga Khan University Hospital

Abdul Hadi Shahid

Aga Khan University Hospital

Case Report

Keywords: Unroofed coronary sinus, Atrial septal defect, Left-to-right shunt, Aortic regurgitation, Case report

Posted Date: August 6th, 2026

DOI: <https://doi.org/10.21203/rs.3.rs-10301556/v1>

License:   This work is licensed under a Creative Commons Attribution 4.0 International License.

[Read Full License](#)

Additional Declarations: No competing interests reported.

Abstract

Background: Unroofed coronary sinus (URCS) is an exceptionally rare congenital cardiac anomaly, accounting for less than 1% of atrial septal defects (ASD). It results from partial or complete absence of the common wall between the coronary sinus and the left atrium, producing a left-to-right shunt. URCS is usually associated with a persistent left superior vena cava (PLSVC); the absence of PLSVC, as in the case described here, is a rarer variant. Because presentation is often nonspecific, diagnosis is frequently delayed and requires advanced multimodality cardiac imaging.

Case presentation: A 50-year-old male ex-smoker presented with a three-month history of palpitations and exertional breathlessness, with a prior syncopal episode. Transthoracic echocardiography demonstrated a 20–22 mm coronary sinus-type ASD due to unroofing of the superior wall of the coronary sinus, left-to-right shunting, severe right atrial and ventricular dilatation, and a bicuspid aortic valve with moderate regurgitation; left ventricular ejection fraction was preserved at 55%. Chest radiography showed cardiomegaly, hilar congestion, and dorsal scoliosis. The patient underwent aortic valve replacement with a 23 mm mechanical bileaflet prosthesis and ASD closure using an autologous fenestrated pericardial patch, with an uneventful early recovery. Two months postoperatively he developed atrial flutter, which was successfully terminated with electrical cardioversion, restoring sinus rhythm.

Conclusions: This case, classified as Type II URCS (unroofed coronary sinus without PLSVC), illustrates the diagnostic value of transthoracic echocardiography in an entity that is easily overlooked, and supports individualised surgical planning, including fenestrated patch closure in patients at risk of elevated right-sided pressures, together with structured long-term follow-up for late arrhythmic complications.

Background

Unroofed coronary sinus (URCS) is a rare congenital cardiac anomaly, accounting for less than 1% of all atrial septal defects (ASD) [1]. It results from partial or complete absence of the common wall separating the coronary sinus from the left atrium, producing direct communication between the two chambers and a resultant left-to-right shunt. URCS is frequently associated with a persistent left superior vena cava (PLSVC) and has been categorised into four morphological subtypes according to the extent of unroofing [2]. Owing to its rarity and often nonspecific symptomatology, URCS is challenging to diagnose and typically requires advanced imaging for confirmation [3]. We report a 50-year-old male with an unroofed coronary sinus without PLSVC, an uncommon variant of an already rare anomaly.

Case presentation

A 50-year-old male ex-smoker presented to the outpatient clinic with a three-month history of palpitations and exertional shortness of breath. He reported a single episode of syncope three months earlier. Clinical examination and an initial metabolic panel were unremarkable.

Transthoracic echocardiography was performed to further investigate his symptoms and revealed a dilated coronary sinus with an atrial septal defect measuring 20–22 mm, showing left-to-right flow due to unroofing of the superior wall of the coronary sinus and a resultant left atrium-to-coronary sinus shunt. The study also demonstrated severe right atrial and right ventricular dilatation, a bicuspid aortic valve with moderate aortic regurgitation, and a left ventricular ejection fraction of 55%. Chest radiography showed cardiomegaly with hilar congestion and dorsal scoliosis with an S-shaped curvature.

Based on the clinical and imaging findings, the patient was diagnosed with an unroofed coronary sinus without PLSVC, an associated ASD, and moderate aortic regurgitation secondary to a calcified bicuspid aortic valve. He was advised to proceed with aortic valve replacement and ASD repair and was admitted for surgical management.

Under general anaesthesia, cardiopulmonary bypass was established via median sternotomy with ascending aortic and bicaval cannulation. Intraoperatively, the severely calcified aortic valve was resected and replaced with a 23 mm mechanical bileaflet prosthesis. The ASD was closed using an autologous fenestrated pericardial patch (Figure 1). The patient had an uneventful postoperative recovery and was discharged home in stable condition.

Two months after discharge, he presented to the Emergency Department with a one-month history of palpitations. An electrocardiogram demonstrated atrial flutter. He underwent electrical cardioversion under sedation, with successful restoration of sinus rhythm. He was discharged with advice to follow up in the outpatient clinic for continued surveillance.

Discussion and Conclusions

The coronary sinus is a venous structure situated along the left posterior atrioventricular groove, adjacent to the circumflex coronary artery and inferior to the left atrium, that normally drains coronary venous blood into the right atrium. Under physiological conditions there is no direct communication between the coronary sinus and the left atrium. When the common wall between these structures fails to form — a defect termed "unroofing" — a left-to-right shunt develops between the left atrium and the coronary sinus [4].

Coronary sinus defects are regarded as a subtype of occult ASD. Kirklin and Barratt-Boyes described four morphological subtypes: type I, complete unroofing with an associated PLSVC; type II, complete unroofing without PLSVC; type III, partial unroofing of the mid-portion; and type IV, partial unroofing of the terminal portion [5].

URCS accounts for less than 1% of all ASDs [6]. In the largest reported surgical series, Quaegebeur and colleagues found that 75% of patients with URCS had an associated PLSVC (consistent with type I disease), while the remaining quarter lacked this association [7]; this distribution has been replicated in

subsequent multimodality imaging cohorts [8]. Our patient, classified as type II URCS without PLSVC, therefore represents an uncommon variant of an already rare anomaly.

The clinical presentation of URCS is largely determined by the size of the defect and the magnitude of the left-to-right shunt. Symptoms range from an asymptomatic course or nonspecific complaints to more pronounced manifestations such as dyspnoea, fatigue, palpitations, syncope, and peripheral oedema [9]. Cardiac findings may be subtle, occasionally including systemic desaturation, and examination may reveal a systolic murmur or an irregular rhythm; tachyarrhythmias are more commonly encountered when diagnosis is delayed [10]. Because symptoms are frequently nonspecific, the diagnosis is easily missed, which increases the complexity of subsequent management [10].

Transthoracic echocardiography (TTE) is typically the first-line imaging modality for suspected URCS, although its diagnostic yield can be limited by difficulty visualising the coronary sinus and other posterior cardiac structures. Real-time three-dimensional TTE has been reported to improve visualisation of surgically unroofed coronary sinus anatomy and may be a useful adjunct where two-dimensional imaging is inconclusive [11]. When transoesophageal echocardiography (TEE) is used, the high four-chamber and mid-oesophageal long-axis views are considered optimal for assessing the coronary sinus [12]. Computed tomography angiography is increasingly used and offers high diagnostic accuracy [9]. In our patient, the diagnosis was established using TTE supplemented by electrocardiography and chest radiography.

Management of URCS is guided by the severity of the shunt and associated symptoms, and generally involves surgical closure of the defect responsible for the left-to-right shunt together with correction of any associated cardiac abnormalities [13]. In this case, the patient also had a significant ASD requiring closure. Pericardial or Dacron patch closure is the preferred surgical technique for ASD repair [13]; we elected to combine aortic valve replacement with ASD closure using an autologous fenestrated pericardial patch. In patients at risk of pulmonary hypertension, complete closure of the defect can precipitate acute right heart failure, and a fenestrated patch is therefore used instead, functioning as a controlled "pop-off" mechanism that permits transient right-to-left shunting to maintain cardiac output during periods of elevated right-sided pressure, such as exertion [14]. Given that our patient was in his sixth decade and therefore at increased risk of pulmonary hypertension, a fenestrated patch was considered an appropriate strategy [15]. Regular postoperative follow-up is recommended to detect late arrhythmic or haemodynamic complications, as occurred in this patient with the development of atrial flutter two months after surgery.

Conclusions: URCS without PLSVC is a rare and structurally complex congenital cardiac anomaly that frequently presents with nonspecific symptoms, and a high index of clinical suspicion supported by targeted echocardiographic assessment is required for timely diagnosis. This case highlights the diagnostic utility of transthoracic echocardiography and the value of an individualised surgical strategy, including fenestrated patch closure where indicated, together with structured long-term follow-up to detect late arrhythmic or haemodynamic sequelae.

Patient perspective

The patient reported significant improvement in his symptoms following surgical intervention and cardioversion. He expressed satisfaction with the information provided about his diagnosis and treatment plan and felt well supported throughout his clinical course. He remains compliant with scheduled follow-up appointments and understands the importance of long-term cardiac surveillance for detection of late haemodynamic or arrhythmic complications.

Abbreviations

ASD
Atrial septal defect
URCS
Unroofed coronary sinus
PLSVC
Persistent left superior vena cava
LSVC
Left superior vena cava
CS
Coronary sinus
LA
Left atrium
RA
Right atrium
RV
Right ventricle
TTE
Transthoracic echocardiography
TEE
Transoesophageal echocardiography
CTA
Computed tomography angiography
CPB
Cardiopulmonary bypass
ECG
Electrocardiogram
ER
Emergency room.

Declarations

Consent for publication: Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

Availability of data and materials: The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interests: The authors declare that they have no competing interests.

Funding: No funding was received for this case report.

Authors' contributions: SMA and LK were major contributors in writing the manuscript and reviewing the literature. SA and WA performed the surgical management and provided clinical data. SAS contributed to the design of the report and critically revised the manuscript. AHS collected and interpreted the patient data. All authors read and approved the final manuscript.

Acknowledgements: Not applicable.

References

1. Aljizeeri A, Dennie CJ, Chan K-L, Beauchesne LM. Unroofed coronary sinus atrial septal defect: diagnosis by multimodality cardiac imaging. *Echocardiography*. 2014. <https://doi.org/10.1111/echo.12588>
2. Xu B, Betancor J, Cremer PC, Renapurkar R, Pettersson GB, Asher CR, et al. Multimodality cardiovascular imaging of unroofed coronary sinus defects. *JACC Cardiovasc Imaging*. 2018;11:1027–30. <https://doi.org/10.1016/j.jcmg.2018.03.001>
3. Chaturvedi A, Dubinsky TJ, Maki JH. MR findings of a rare defect, coronary sinus ASD. *Int J Cardiovasc Imaging*. 2012;28:429–30. <https://doi.org/10.1007/s10554-010-9670-8>
4. Cintează EE, Filip C, Duică G, Nicolae G, Nicolescu AM, Bălgrădean M. Unroofed coronary sinus: update on diagnosis and treatment. *Rom J Morphol Embryol*. 2019;60:33–40.
5. 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. [https://doi.org/10.1016/S0022-5223\(03\)01019-5](https://doi.org/10.1016/S0022-5223(03)01019-5)
6. Sugimori H, Nakao T, Ikegaya Y, Iwahashi D, Tsuda S, Kume N, et al. Coronary sinus atrial septal defects in adults over the past 20 years at new Tokyo hospital: case series. *J Cardiothorac Surg*. 2021;16:1–9. <https://doi.org/10.1186/s13019-021-01522-x>
7. Quaegebeur J, Kirklin JW, Pacifico AD, Barger LM Jr. Surgical experience with unroofed coronary sinus. *Ann Thorac Surg*. 1979;27:418–25. PMID: 454016.
8. Kim H, Choe YH, Park SW, Jun TG, Kang IS, Yang JH, et al. Partially unroofed coronary sinus: MDCT and MRI findings. *AJR Am J Roentgenol*. 2010;195:W331–6. <https://doi.org/10.2214/AJR.09.3689>

9. Thangaroopan M, Truong QA, Kalra MK, Yared K, Abbata S. Rare case of an unroofed coronary sinus: diagnosis by multidetector computed tomography. *Circulation*. 2009;119:e518–20. <https://doi.org/10.1161/CIRCULATIONAHA.107.707018>
10. Sun L, Juan YH, Chen J, Zhuang J, Xie J, Li H, et al. Evaluation of unroofed coronary sinus syndrome using cardiovascular CT angiography: an observational study. *AJR Am J Roentgenol*. 2018;211:314–20. <https://doi.org/10.2214/AJR.17.19128>
11. Singh A, Nanda NC, Romp RL, Kirklin JK. Assessment of surgically unroofed coronary sinus by live/real time three-dimensional transthoracic echocardiography. *Echocardiography*. 2007;24:74–6. <https://doi.org/10.1111/j.1540-8175.2006.00359.x>
12. Pojar M, Timbilla S, Cerny S, Karalko M, Vojacek J. Case report: robotic repair of unroofed coronary sinus. *Front Cardiovasc Med*. 2022;9:974089. <https://doi.org/10.3389/fcvm.2022.974089>
13. Geva T, Martins JD, Wald RM. Atrial septal defects. *Lancet*. 2014;383:1921–32. [https://doi.org/10.1016/S0140-6736\(13\)62145-5](https://doi.org/10.1016/S0140-6736(13)62145-5)
14. Qureshi AM, Kenny D. Atrial septal defect closure in patients. *JACC Cardiovasc Interv*. 2020;13:2035–7. <https://doi.org/10.1016/j.jcin.2020.05.023>
15. Berra G, Noble S, Soccal PM, Beghetti M, Lador F. Pulmonary hypertension in the elderly: a different disease? *Breathe*. 2016;12:43–9. <https://doi.org/10.1183/20734735.003416>

Figures

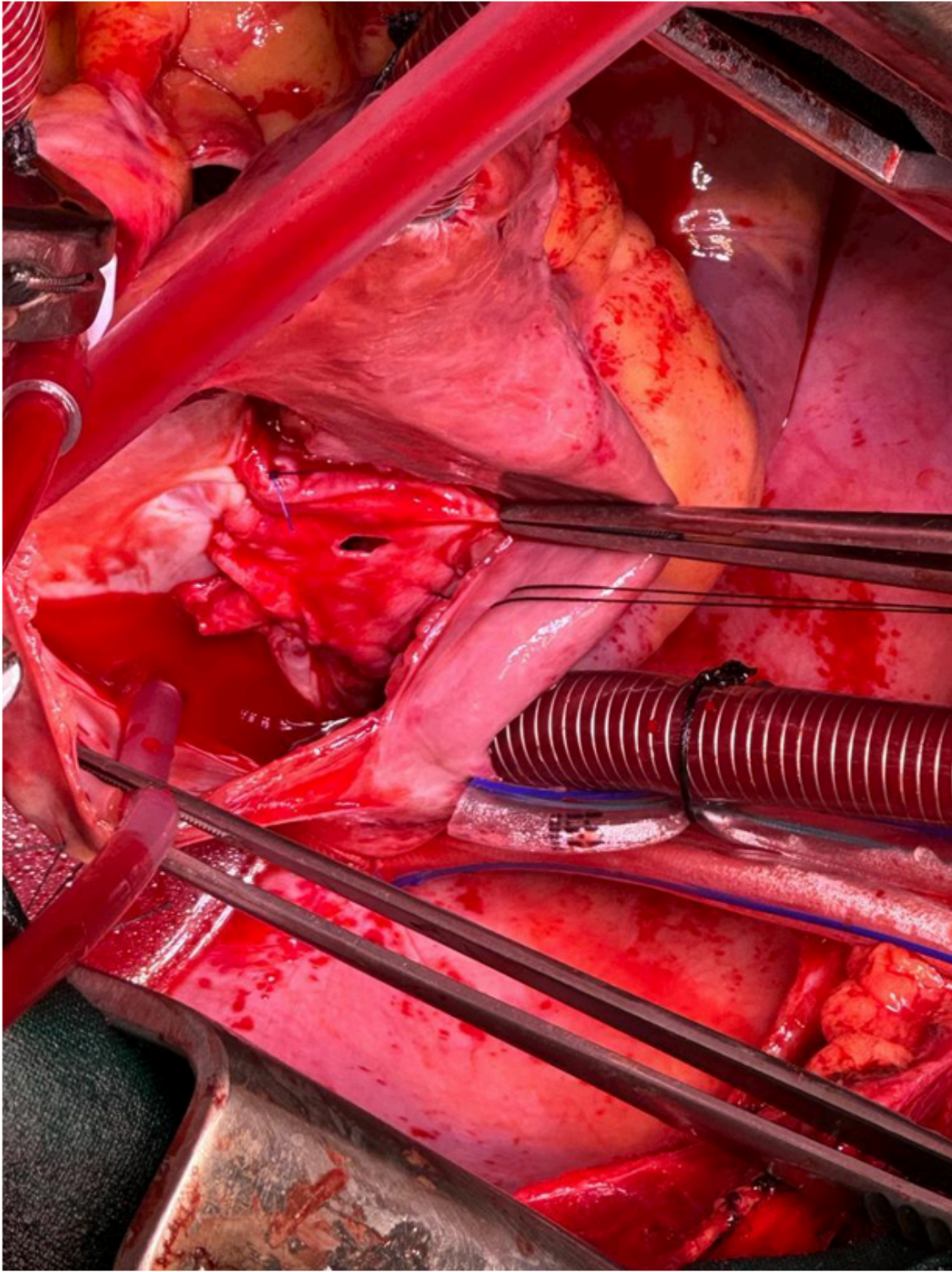


Figure 1

Repair of Coronary Sinus ASD utilizing a fenestrated autologous pericardial patch.

Intraoperative photograph demonstrating repair of the coronary sinus atrial septal defect (ASD) using an autologous fenestrated pericardial patch. The fenestration permits controlled right-to-left decompression in the event of elevated right-sided pressures.

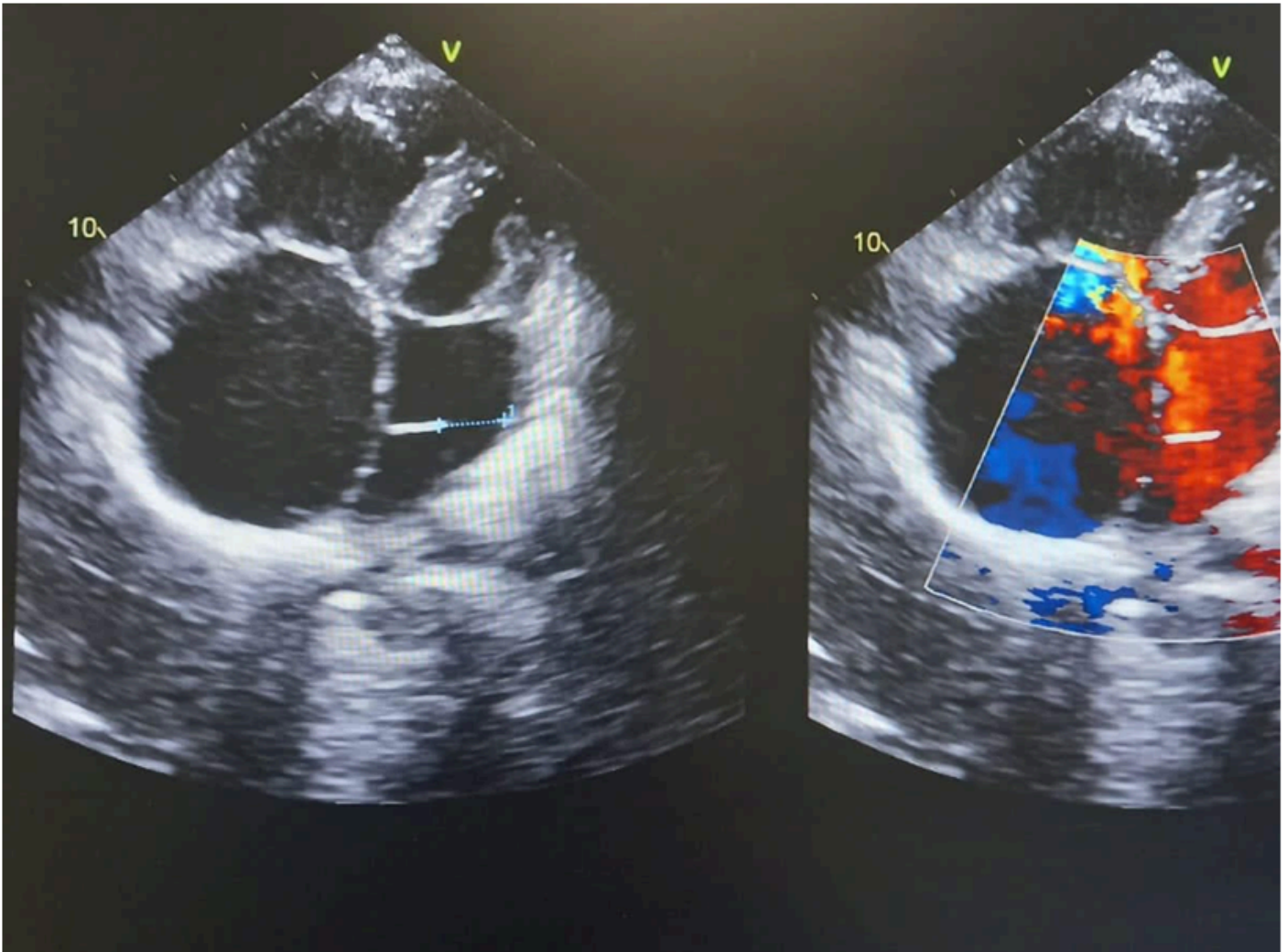


Figure 2

Preoperative transesophageal echocardiography images showing the severely dilated and unroofed coronary sinus.

Preoperative transthoracic echocardiography images demonstrating the severely dilated and unroofed coronary sinus, with evidence of left-to-right shunting across the defect.