Multimodality Imaging in the Diagnosis of Unroofed Coronary Sinus in the Adult

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INTRODUCTION

The population of adults with congenital heart disease (CHD) has grown rapidly over the last few decades and is projected to keep increasing for the next several decades.[1] An estimated 10% of all patients with CHD are initially diagnosed in adulthood[2] and thus likely to present to a noncongenital adult echocardiographic laboratory. The echocardiographer therefore must be adept in the identification of these previously unrecognized patients, as the impact of untreated CHD can have a significant clinical burden as these patients age.

Atrial-level shunts are among the most common type of CHD in adulthood, which is when they are often diagnosed. Most echocardiographers are familiar with the appearance of secundum-type atrial septal defects (ASDs), as they comprise approximately 75% of all atrial shunts.[3] However, the rarer atrial shunts, including unroofed coronary sinus (CS) which represents fewer than 1% of these lesions, may not be as recognizable.[4] Thus, we present a series of 3 adult patients who presented to the adult echocardiography laboratory with previously undiagnosed unroofed CS to illustrate the typical presentation and imaging findings of this uncommon form of CHD. Echocardiography is critical in the initial diagnosis of unroofed CS, and despite its rarity, this shunt can be identified by the astute adult echocardiographer when they are familiar with the patterns of presentation.

Case Presentation 1

A 64-year-old female patient with a history of unspecified CHD, diagnosed in the third decade of life, presented to a primary care clinic with 1 year of progressive exertional dyspnea and nonexertional chest pain. Physical examination revealed normal vital signs including an oxygen saturation of 100% at rest and auscultatory findings of a fixed split S2 heart sound with a II/VI midsystolic murmur heard at the right upper sternal border. Transthoracic echocardiogram (TTE) revealed a large-sized left-to-right interatrial shunt on subcostal 4-chamber view (Figure 1A). A markedly dilated CS was seen on parasternal right ventricular inflow view (Figure 1B, Video 1) with a suspicion for a deficient anterior roof on apical 4-chamber view (Figure 1C). Also noted on apical 4-chamber view was moderate dilation of the right atrium (RA) and right ventricle (RV) with intact septum primum (SP) and normal apical displacement of the tricuspid valve (TV) relative to the mitral valve (MV; Figure 1D).

Subsequent imaging in the adult congenital lab with TTE included a superior-inferior subcostal 4-chamber sweep demonstrating an unrestrictive communication between the left atrium (LA) and the dilated CS with shunting into the RA, confirming the diagnosis of unroofed CS (Video 2). Cardiac computed tomography (CCT) confirmed the diagnosis of a completely unroofed CS into the LA with right heart dilation (Figure 2, Video 3). No left superior vena cava (LSVC) was present, and there was no evidence of pulmonary hypertension (PH) by TTE.

The patient was referred for surgical correction. Intraoperative transesophageal echocardiography (TEE) confirmed the findings of an intact SP and septum secundum (Figure 3A) as well as a completely unroofed CS with left-to-right shunting through a severely dilated CS ostium (Figure 3B and C, Videos 4 and 5). The CS ostium was closed with a pericardial patch with resolution of interatrial shunting (Figure 3D); the CS-LA connection was left patent, resulting in a small residual right-to-left shunt. Upon 6-month follow-up the patient reported resolution of exertional dyspnea, and physical examination revealed a normal oxygen saturation of 97%.

Case Presentation 2

A 47-year-old female patient with a history of a childhood murmur leading to diagnosis of an unspecified intracardiac defect with no cardiology follow-up in adulthood presented to the emergency department with subacute progressive exertional dyspnea, recurrent right-sided pleural effusion, ascites, and pedal edema. Vital signs were notable for hypertension to 145/98 mm Hg, tachycardia with a regular heart rate at 103 bpm, mild tachypnea at 20 breaths per minute, and oxygen saturation of 90% at rest. A mild RV heave and fixed split S2 sound as well as absent breath sounds over the lower third of the posterior right lung field were noted. The patient underwent thoracentesis and intravenous diuresis as an inpatient. A TTE demonstrated dilation of the RA and RV, grade 1 diastolic dysfunction, and LA dilation. Imaging was suggestive of an interatrial shunt diagnosed as an ostium primum ASD. Referrals to the adult CHD team and a congenital TTE were obtained.

An atrial-level shunt was visualized on a modified subcostal view and was demonstrated to occur through a dilated CS rather than a defect in the SP (Figure 4A-C), raising a strong suspicion for unroofed CS. Also noted was a probable LSVC (Figure 4A). The TV continuous-wave Doppler revealed an RV-RA gradient of 57 mm Hg, consistent with significant PH (Figure 4D).

The TEE confirmed the presence of a dilated CS, completely unroofed into the LA with predominant left-to-right interatrial shunting. Agitated saline contrast study performed through a left upper extremity revealed a copious number of bubbles appearing in the LA and LV before the RA and RV, confirming the presence of a persistent LSVC to the LA (Video 6). Subsequent CCT clearly demonstrated a right

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VIDEO HIGHLIGHTS

Video 1: Case 1, 2D TTE, parasternal RV inflow view without (*left*) and with (*right*) color-flow Doppler, demonstrates a severely dilated CS with large volume, predominantly left-to-right shunt.

Video 2: Case 1, 2D TTE. Superior-inferior sweep performed on a subcostal 4-chamber, inverted (congenital) view zoomed in on the atria (LA in near field), without (*left*) and with (*right*) color-flow Doppler at a reduced Nyquist limit demonstrates the intact SP and septum secundum, the dilated CS with a predominantly left-to-right shunt, and the CS opening into the RA.

Video 3: Case 1, CCT, axial stack inferior to superior demonstrates coronary veins draining to the dilated CS that connects to the LA directly through a large (unroofed) posterior connection and drains through the CS ostium into the RA. The SP and septum secundum are intact, and the RA and RV are markedly dilated due to the significant left-to-right shunting.

Video 4: Case 1, 2D TEE, lower esophageal rightward rotated long-axis (103°) view without (*left*) and with (*right*) color-flow Doppler demonstrates the unroofed CS (*asterisks*) contiguous with the LA, opening into the RA.

Video 5: Case 2, 2D TEE, midesophageal long-axis (83°) view, left-to-right sweep with color-flow Doppler demonstrates a completely unroofed CS with left-to-right shunt from the LA to the RA.

Video 6: Case 2, 2D TEE, midesophageal rightward rotated apical 4-chamber (0°) view with agitated saline contrast from the left upper extremity demonstrates bubbles appearing first in the LA and LV, before appearing in the severely dilated right heart, confirming the diagnosis of an LSVC draining into an unroofed CS.

Video 7: Case 3, 2D TTE, zoomed apical 4-chamber view without (*left*) and with (*right*) color-flow Doppler, demonstrates abnormal interatrial flow into the RA, coming through a dilated CS.

Video 8: Case 3, cardiovascular magnetic resonance white-blood cine steady-state free precession axial view demonstrates the dilated CS with no separation from the LA and a flow signal artifact suggesting flow into the RA in the region of the CS ostium with an intact SP and septum secundum.

Video 9: Case 3, intraoperative TEE with three-dimensional reconstruction, modified surgeon's view of the RA without (*left*) and with (*right*) color-flow Doppler demonstrates the interatrial septum en face, the SVC and IVC, the AV and TV, the fossa ovalis, and the dilated CS ostium with flow entering the RA.

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superior vena cava to the RA (Figure 5A) and LSVC to a completely unroofed and massively dilated CS (Figure 5B-D). Hemodynamic cardiac catheterization revealed a shunt fraction of 1.8:1 and confirmed moderate PH with mean pulmonary artery pressure of 38 mm Hg and mildly elevated pulmonary vascular resistance of 2.4 Woods units.

The patient underwent surgical patch closure of the CS ostium with autologous pericardium. Additionally, the LSVC was transected at the level of the heart and rerouted to the RA appendage using an 18 mm graft. The postoperative course was notable for atrial fibrillation requiring multiple cardioversions and a pericardial effusion that required pericardiocentesis. The patient was ultimately discharged and reported resolution of lower-extremity edema on short-term follow-up with an oxygen saturation of 94%. A TTE 4 months after surgery revealed interval reduction in RA and RV size.

Case Presentation 3

A 41-year-old female patient presented to a pulmonology clinic after incidental diagnosis of cardiomegaly on a chest x-ray performed prior to knee surgery for a benign giant cell tumor. History was notable for lifelong exertional dyspnea with vigorous activity and inability to keep up with peers during physical activity. This had led to a diagnosis of asthma at age 10 years, but prescription for bronchodilator therapy did not result in symptomatic improvement. Pulmonary function testing with bronchodilator challenge showed no evidence of asthma, and a TTE was ordered. An ostium primum ASD was suspected, and the patient was referred to adult congenital cardiology for additional evaluation. Vital signs were normal, but physical examination revealed an RV heave and fixed split S2.

The TTE images were suspicious for unroofed CS with findings of mild right heart dilation and abnormal color Doppler flow on apical 4-chamber view entering the RA through the CS (Video 7). A congenital TTE protocol was performed. Parasternal long-axis view revealed the so-called broken ring sign consistent with complete unroofing of the CS into the LA (Figure 6).

A cardiovascular magnetic resonance imaging scan revealed a completely unroofed CS with left-to-right shunting (Video 8). Right heart dilation was confirmed, as was a shunt fraction of 1.4:1 and absence of an LSVC. In light of a symptomatic atrial-level shunt with right heart dilation and absence of PH, the patient underwent surgical repair. Intraoperative TEE images confirmed the diagnosis, and postclosure imaging showed complete closure of the CS ostium with a pericardial patch (Figure 7, Video 9). On short-term follow-up, exercise capacity was improved and resting saturation was 96%.

DISCUSSION

The CS is the largest coronary venous structure and represents the confluence of the entire coronary venous drainage. As with the other venous structures leading to the heart, the CS derives its embryologic origin from the sinus venosus, which is the primary venous inflow to the primordial atrium. The sinus venosus occupies a posterior position relative to the primordial atrium and divides into the right and left horns. The left horn of the sinus venosus separates from the leftward aspect of the primordial atrium by the fourth week of gestation via an invagination that eventually serves to separate the CS from the pulmonary venous (left) atrium, and the left horn regresses by the 10th week of gestation to form the CS.[5,6] Complete or partial deficiency of this invagination leads to a residual complete or partial persistence of communication between the CS and LA, giving rise to unroofed CS.

While commonly classified as such, unroofed CS is not a true ASD since the interatrial septum is intact and the interatrial shunt occurs exclusively through the CS ostium. Appreciation of this distinction is key to making the diagnosis on echocardiography. The communication between the CS and the LA results in the CS functioning as a

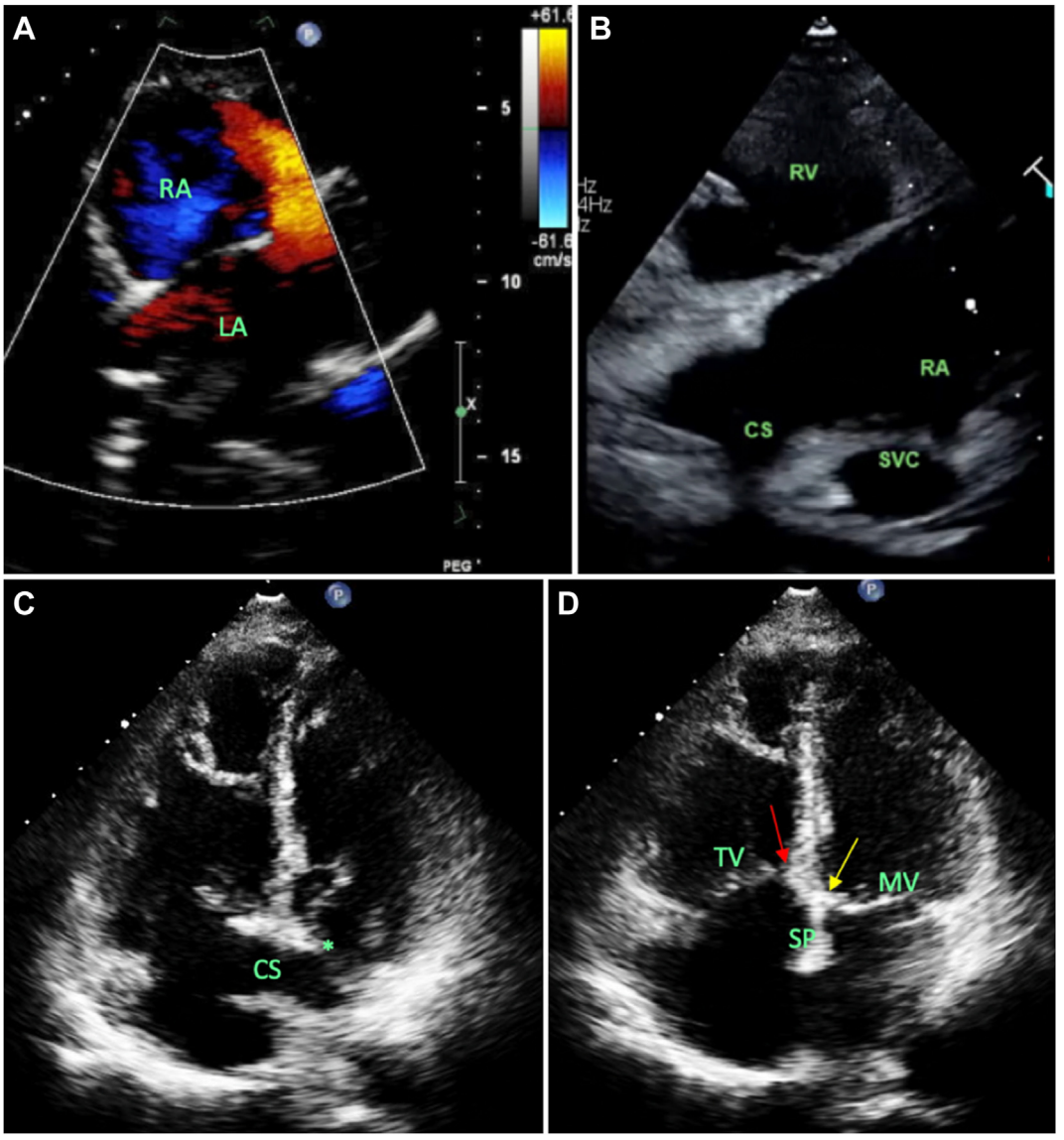

Figure 1 Case 1, 2D TTE. **(A)** Subcostal zoomed 4-chamber view with color-flow Doppler demonstrates a large, continuous low-velocity left-to-right shunt near the crux of the heart at the region of the basal septum. The septum secundum is intact. **(B)** Parasternal RV inflow view demonstrates a severely dilated CS and CS ostium. **(C)** Apical 4-chamber view with posterior angulation demonstrates a severely dilated CS with a defect adjacent to the MV (*) and communication with the LA. **(D)** Apical 4-chamber view demonstrates moderate RA and RV dilation, an intact SP, and appropriate apical displacement of the septal insertion of the TV (*red arrow*) relative to the MV (*yellow arrow*).

conduit between the atria. Like other atrial-level shunts, the shunt direction is predominantly left to right from the LA to CS and ultimately to the RA, due to the lower diastolic compliance of the RV relative to the left ventricle (LV). This can result in right heart dilation, PH, and functional limitation. Left-sided diastolic dysfunction and/or left heart dilation is not part of the pathophysiologic manifestation. However, the development of diastolic dysfunction of the left heart chambers can considerably increase left-to-right shunt fraction through any anatomically stable atrial-level shunt that may have previously gone undetected for several decades. This can then precipitate onset of symptoms and lead to diagnosis later in adulthood. This physiology characterizes the patient in case 2, in the context of preexisting and presumably progressive systemic PH.

Over 60% of patients of unroofed CS are associated with an LSVC, an association first described by Raghib *et al.* in 1965 and thus known eponymously as Raghib syndrome.[7,8] The connection of the LSVC to the unroofed CS is known to cause paradoxical emboli, stroke, and brain abscess due to the obligatory right-to-left shunt component unique to this syndrome compared with the isolated unroofed CS.[9,10] The presence of LSVC has important implications for surgical management and features in the classification by Kirklin and Barratt-Boyes (Table 1).[11]

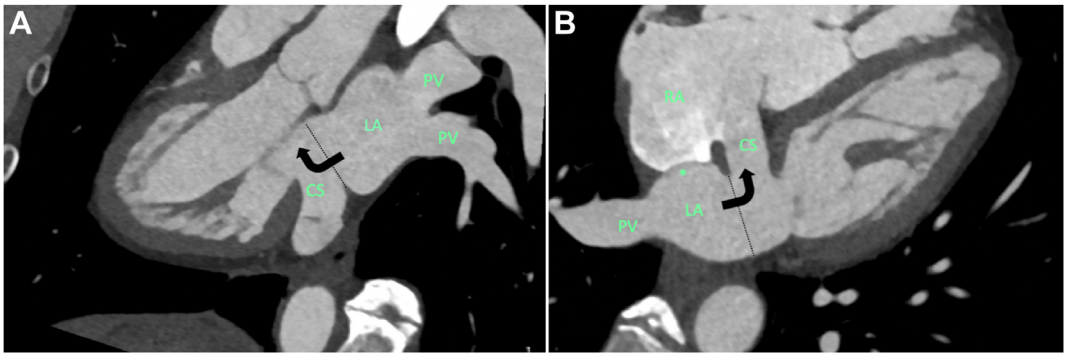

Figure 2 Case 1, CCT oblique sagittal LV outflow tract view **(A)** and oblique axial 4-chamber view **(B)**, demonstrates the pulmonary veins (PV) connecting to the LA and the dilated CS with direct communication with the LA and an intact atrial septum (*). The *line* represents the area where there is deficiency (unroofing) of the CS resulting in a significant left-to-right shunt (*arrows*).

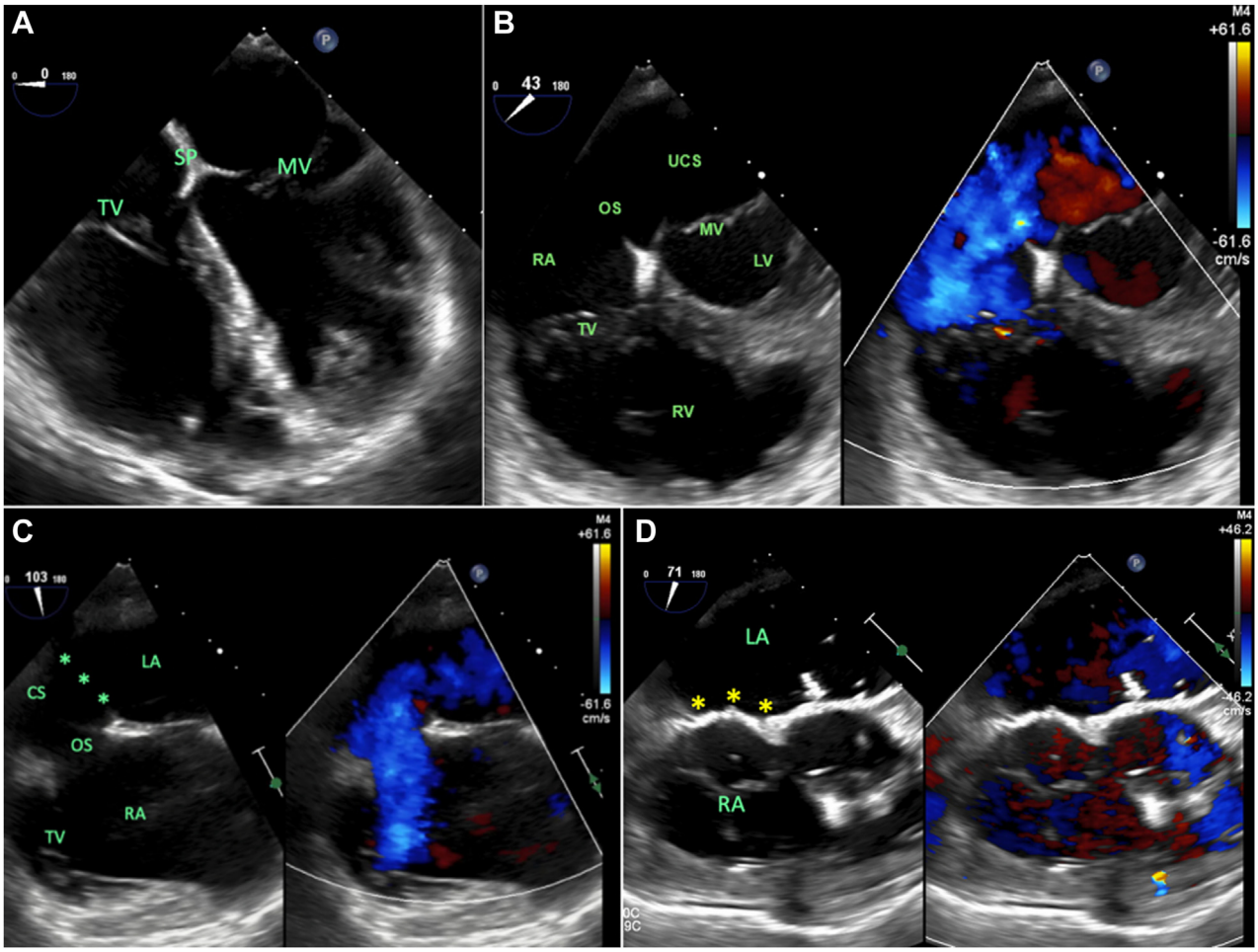

Figure 3 Case 1, TEE. **(A)** Two-dimensional midesophageal 4-chamber (0°) view demonstrates intact SP and normal apical displacement of the TV. **(B)** Two-dimensional off-axis lower esophageal (43°) view with slight retroflexion without (*left*) and with (*right*) color-flow Doppler demonstrates that the CS is contiguous with the LV inlet and MV indicative of complete unroofing. Shunting is noted from the LA into the RA through the CS ostium (OS). **(C)** Two-dimensional TEE, lower esophageal rightward rotated long-axis (103°) view without (*left*) and with (*right*) color-flow Doppler, demonstrates the unroofed CS (*asterisks*) contiguous with the LA, opening into the RA. **(D)** Intraoperative 2D TEE, lower esophageal rightward rotated long-axis (71°) view without (*left*) and with (*right*) color-flow Doppler, demonstrates successful surgical patch closure of the CS OS (*asterisks*) with no residual shunting.

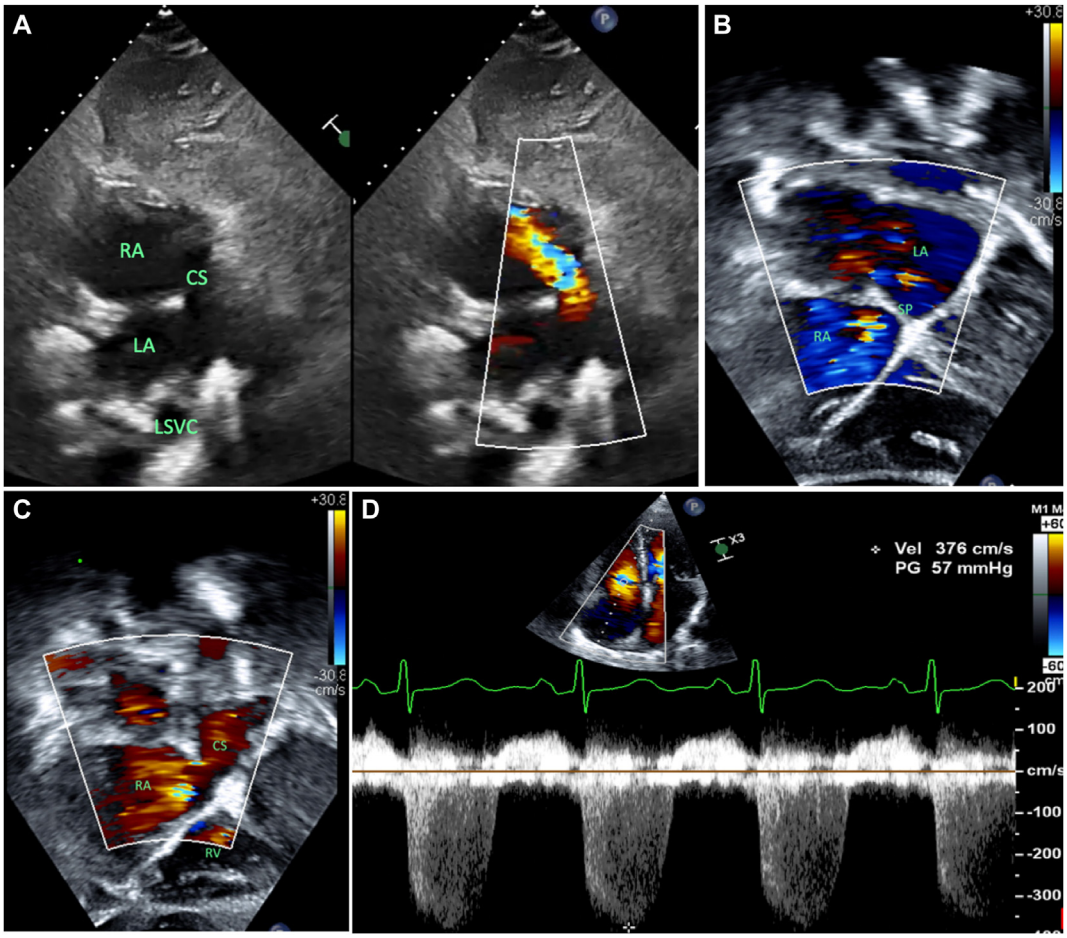

Figure 4 Case 2, 2D TTE. **(A)** Modified subcostal view with inferior angulation without (*left*) and with (*right*) color-flow Doppler demonstrates shunting predominantly from the LA to the RA via a dilated CS. Also noted is an LSVC. **(B)** Inverted subcostal 4-chamber view with color-flow Doppler, demonstrates an intact SP and septum secundum. **(C)** Inverted subcostal 4-chamber inferior angulation with color-flow Doppler view demonstrates a dilated CS with left-to-right flow through the CS into the RA. **(D)** Apical 4-chamber view with color-flow Doppler-guided continuous-wave Doppler across the TV demonstrates a peak tricuspid regurgitant velocity of 3.76 m/sec (57 mm Hg) indicating significant PH.

We present 2 patients with type II (cases 1 and 3) and 1 patient with type I (case 2) unroofed CS. In all patients, TTE was either strongly suspicious or diagnostic. The shared imaging features were

- RA and RV dilation.
- Marked dilation of the CS accompanied by significant continuous flow left to right at the CS ostium into the RA. Even in the presence of an isolated LSVC, one does not see this amount of flow into the dilated CS without concomitant unroofed CS.
- Visualization of the left-to-right interatrial shunt in the subcostal long-axis (4-chamber) view by color-flow Doppler despite an intact SP and septum secundum.
- An intact atrioventricular canal septum and appropriate apical displacement of the TV compared with the MV, ruling out an ostium primum ASD (Figure 4).

The last point deserves emphasis, especially since 2 of the 3 patients were originally misdiagnosed with ostium primum ASD. Despite the name, ostium primum defects are defects in the embryologic endocardial cushions that form the atrioventricular canal septum, rather than true defects in the SP. They are invariably accompanied by defective septation of the common atrioventricular valve to the MV and TV, resulting in the septal leaflets of the MV and TV inserting at the same level, as well as a "cleft" in the left atrioventricular (mitral) valve.

Additional two-dimensional (2D) imaging of the CS can reveal a complete continuity between the CS and the LA as seen in Figure 1; however, it can be extremely difficult to diagnose unroofed CS based on 2D imaging alone. An apical 4-chamber view provides the ideal angulation for 2D imaging of the roof of the CS (perpendicular to the probe); however, even with careful gain settings it can be difficult to distinguish unroofing from acoustic dropout. The same imaging should be attempted on the parasternal long-axis view, where a completely unroofed CS can produce the broken ring sign (Figure 7A). In terms of color-flow Doppler interrogation, the angulation of these views is ideally parallel with the probe; however, completely unroofed CS will not have an easily defined color jet and only with a very low Nyquist setting, appropriate gain, and high suspicion will one be able to rely on these anatomic and physiologic features alone.

The presence of an LSVC considerably increases the volume of the inflow into the CS and thus the pathologic severity of the interatrial shunt. Consequently, patients with the so-called Raghib syndrome

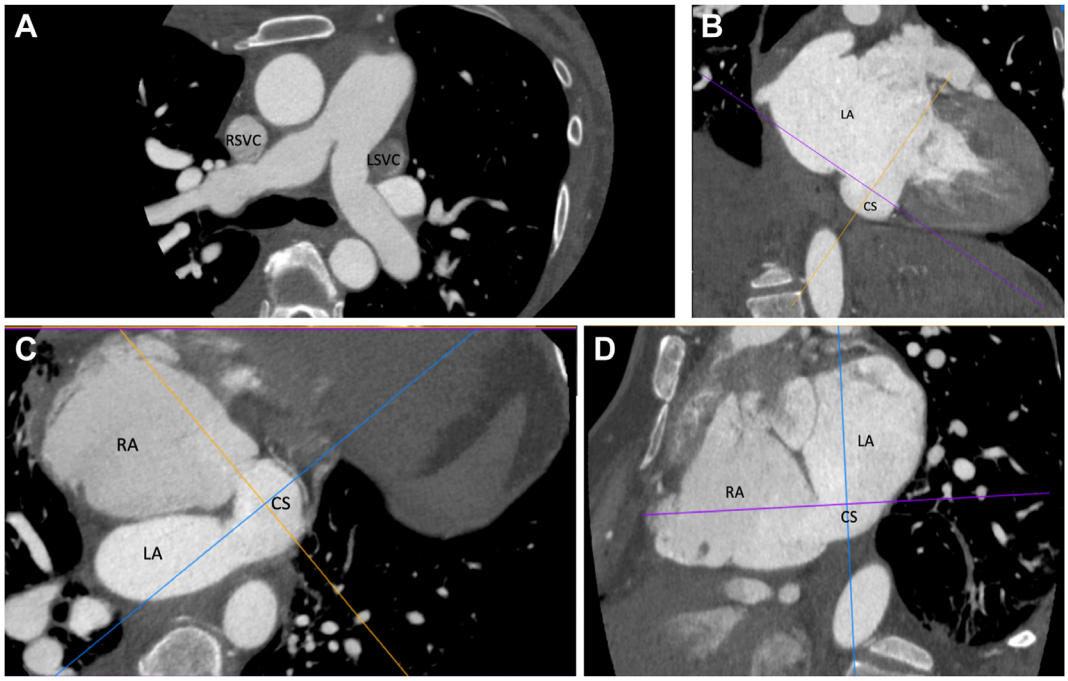

Figure 5 Case 2, CCT. **(A)** Axial image demonstrates a normal RSVC that is opacified with contrast and a nonopacified LSVC. (**B**-**D**) Three oblique orthogonal views generated using three-dimensional multiplanar reconstruction demonstrate the dilated CS in relation to the LA and RA. *RSVC*, Right superior vena cava.

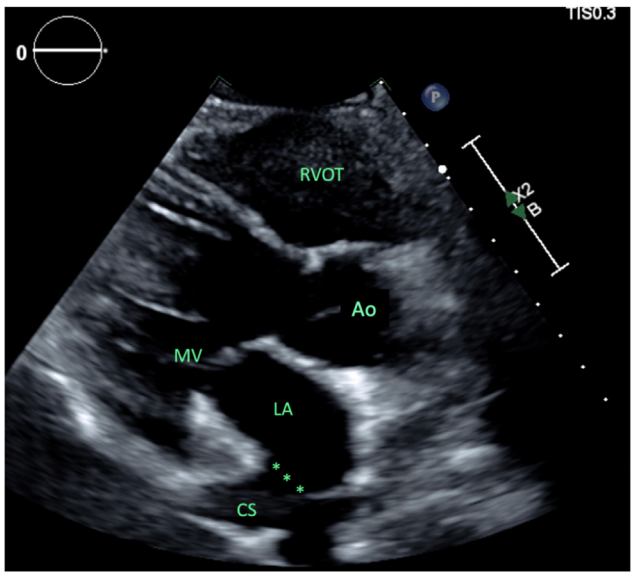

Figure 6 Case 3, 2D TTE, parasternal long-axis view, demonstrates a dilated CS in the atrioventricular groove with complete unroofing (*asterisks*) into the LA, that is, the broken ring sign. *Ao*, Aorta; *RVOT*, right ventricular outflow tract.

can have an even larger diameter of the CS compared to an unroofed CS without LSVC. A high left parasternal view with color-flow Doppler will usually demonstrate the presence of an LSVC and should be performed when a diagnosis of unroofed CS is noted. Conversely, when a dilated CS is seen incidentally and there is increased CS flow to the RA by color-flow Doppler, agitated saline contrast should be administered to diagnose unroofed CS. Agitated saline contrast, when injected via the left upper extremity in a patient with an LSVC draining into an unroofed CS, gives a pathognomonic finding: the saline contrast fills the CS, followed by the LA and then the RA. Due to the direct CS-LA communication, the filling of the LA can be instantaneous with and indistinguishable from filling of the CS, especially in a type I unroofed CS (Video 6). An isolated LSVC with an intact CS would only opacify the RA with agitated saline injection in the left upper extremity. Importantly, a right upper extremity contrast injection is of little value with or without the presence of an LSVC, as any right-to-left saline contrast shunting would be indistinguishable from any other form of atrial-level shunt.

Transesophageal echocardiography is a valuable adjunct to diagnosing unroofed CS, provided that the echocardiographer is aware of its potential pitfalls. A completely unroofed CS can be challenging to diagnose since the CS and LA are in complete continuity and the characteristic tubular structure of the CS does not exist. Identification of the CS itself can hence be challenging. The echocardiographer must be alert to the possibility of the unroofed CS when on a midesophageal 4-chamber view at 0° the SP is intact and the atrioventricular valve insertion points at the septum are discrepant, but a large atrial-level shunt is seen only when the probe is advanced further into the lower esophagus, where the CS would be expected to be usually seen. In the case of an LSVC, however, the massive dilation of the unroofed CS can result in the CS being visible even in a conventional midesophageal view and can lead to the erroneous diagnosis of ostium primum ASD.

A midesophageal view at ~100° to 120° is the most diagnostic, as it demonstrates the CS in cross section and its deficient roof, but this is a rarely performed view and thus can be challenging to interpret. In our experience, while this is the most helpful view for diagnosis on TEE, care must be taken to not mistake the CS for the inferior vena cava

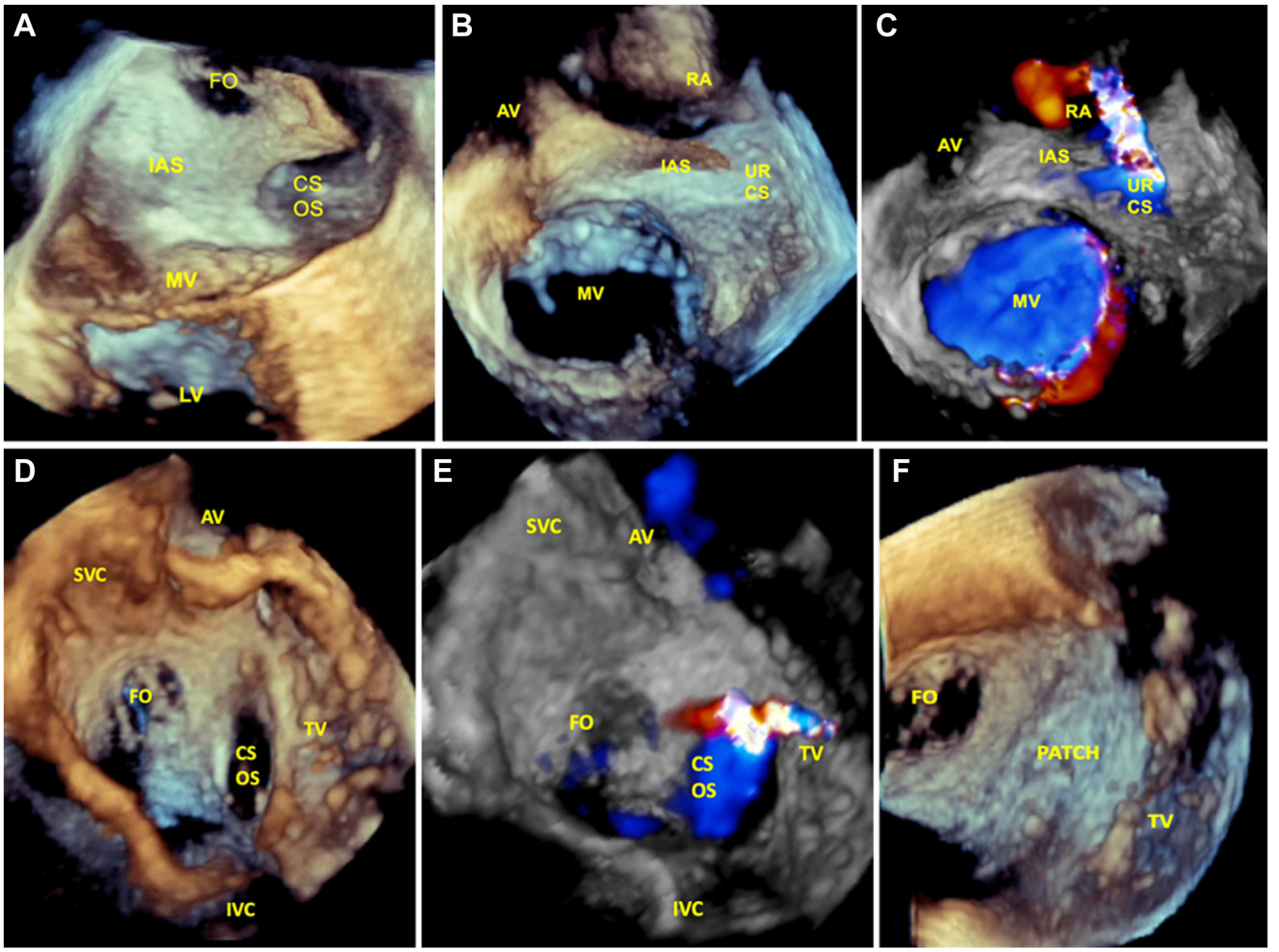

Figure 7 Case 3, intraoperative three-dimensional TEE volume-rendered reconstruction images. **(A)** En face view of the inferior aspect of the interatrial septum (IAS) as visualized from the lateral aspect of the LA. Inferoposteriorly the CS ostium (OS) is seen with lack of a roof separating the CS from the body of the LA. The fossa ovalis (FO), MV, and LV are also seen. **(B, C)** Surgeon's view of the LA without **(B)** and with **(C)** color-flow Doppler demonstrates the large CS running in the atrioventricular groove posterior to the MV and a large right-to-left shunt through the unroofed CS. **(D, E)** Modified surgeon's view of the RA without **(D)** and with **(E)** color-flow Doppler demonstrates the IAS en face, the superior vena cava (SVC) and IVC, the AV and TV, the fossa ovalis (FO), and the dilated CS OS with flow entering the RA. **(F)** Postoperative three-dimensional TEE, modified surgeon's view of the RA, demonstrates that the CS OS has now been closed with a surgical patch.

Table 1 Classification of unroofed CS as per Kirklin and Barratt-Boyes

Type of unroofed CS	Degree of unroofing	Additional qualifier
I	Complete	LSVC present
II	Complete	LSVC absent
III	Partial	Midportion of CS unroofed
IV	Partial	Terminal portion of CS (near ostium) unroofed

(IVC). Vaskelyte *et al.*[12] have elegantly demonstrated why even an intact CS is very commonly mistaken for the IVC on a conventional midesophageal bicaval view as the CS is also a right-sided inferoposterior venous structure that lies adjacent to the IVC. In the setting of an unroofed CS, not only can the dilation of the CS confound identification of the IVC, but the inferoposterior location of the shunt can be mischaracterized as an inferior sinus venosus defect. Advancing the probe inferiorly to find the intrahepatic IVC and then retracting the probe while following it draining into the RA can conclusively identify the IVC and distinguish it from the CS. This maneuver can also evaluate for override of the IVC, which is typically seen in an inferior sinus venosus defect. Even if this is not performed, however, the echocardiographer should recognize that when a large atrial-level shunt is seen both at the crux of the heart at 0° and in an inferoposterior location at 120°, the shunt can represent neither an ostium primum ASD (which is not inferoposterior) nor an inferior sinus venosus defect (which cannot be seen at 0° adjacent to the atrioventricular valves). Rather, the only diagnosis that would explain the shunt in both views would be an unroofed CS.

Thus, upon suspicion of unroofed CS by TTE, we recommend CCT or cardiovascular magnetic resonance imaging to confirm and characterize the lesion and definitively identify the presence or

Table 2 Unroofed CS echocardiography imaging pearls

TTE	
Apical 4-chamber	• Right heart dilation • CS dilation with continuous left-to-right color-flow Doppler shunt with low Nyquist limit • Intact atrioventricular canal septum • Normal apical displacement of the TV • Estimation of RV systolic pressure with TR
Subcostal 4-chamber/long-axis	• Intact SP and septum secundum • Visualization of CS ostium • Color-flow Doppler (low Nyquist limit) with continuous flow
Saline contrast (bubble study)	• Left arm injection required to identify LSVC • Apical 4-chamber or parasternal long-axis view • Bubbles seen in LA before RA if LSVC drains to unroofed CS
TEE	
100°-120°	• Leftward tilt: unroofed CS in cross section • Rightward tilt: left-to-right shunt from LA to CS
0°	• Intact atrioventricular canal septum • Normal apical displacement of the TV • Estimation of RV systolic pressure with TR
Saline contrast (bubble study)	• Left arm injection required to identify LSVC • 0° midesophageal 4-chamber view • Bubbles seen in LA before RA if LSVC drains to unroofed CS

absence of an LSVC. Typically, TEE will be performed intraoperatively; thus an understanding of the imaging techniques described above is important to confirm postsurgical success, but the performance of an isolated diagnostic preoperative TEE is rarely indicated after cross-sectional imaging is obtained.

A partially unroofed CS has specific challenges in diagnosis by TEE, since the interatrial shunt may only be seen if the probe is advanced both inferiorly to visualize the CS and angled leftward to visualize the roof of the CS (and thus the defect). If focus is given only to the RA and the CS at its ostium, the diagnosis may be missed due to its focality. The 100° to 120° midesophageal view can once again be useful, as the cross section of the CS can be seen and a defect in the roof can be identified by color-flow Doppler.[13,14] Table 2 summarizes some of the key features of unroofed CS as seen by TTE and TEE.

Surgical management is the current mainstay of management in patients with symptomatic shunting and/or significant shunt with evidence of right heart dilation. Successful percutaneous closure of unroofed CS with commercially available septal occluders and covered stents has been reported in isolated case reports, but long-term durability of such procedures is unknown.[15-17] Hemodynamic catheterization is indicated in these patients if there is any suspicion of PH by echocardiography. A pulmonary vascular resistance greater than one-third systemic and/or pulmonary artery systolic pressure greater than one-half systemic imparts significant risk. Such patients should be referred to a specialist multidisciplinary team with adult CHD and PH experts before surgical repair is entertained.[18]

Several surgical approaches have been described in the literature, with the procedure of choice being dependent on the type of unroofed CS and on surgeon preference. For cases not associated with LSVC, options include surgical patch closure of the ostium via right atriotomy, which leaves the LA portion of the CS unroofed, or reroofing the CS through a left atriotomy. The former is preferred at our institution, especially for completely unroofed CS, for its technical ease, reduced risk of atrioventricular block, and shorter pump time. This approach, which was used in all 3 of the patients in this case series, leaves patent the coronary venous connection to the LA, creating a small net right-to-left shunt, which as demonstrated, does not cause significant desaturation or clinical symptoms. A concomitant LSVC can be addressed in several ways: In the presence of a bridging vein connecting to the right superior vena cava, the LSVC can be ligated below the level of the bridging vein without rerouting. In the absence of a bridging vein, the LSVC must be connected to the RA—techniques described include a baffle into the RA via a surgical patch, an intra-atrial tunnel to the RA, or an extracardiac synthetic graft, as was done in case 2 in this series.[8,19,20] No head-to-head comparison data are yet available, but in a large retrospective series of 159 cases of surgical repair of unroofed CS using several of the aforementioned techniques (mostly intra-atrial tunnel and baffle creation), a 3.1% in-house mortality was observed with no long-term deaths in hospital survivors.[8] All patients who died had a considerable burden of other associated CHD lesions.

CONCLUSION

We present 3 unique patients with unroofed CS diagnosed in adulthood with common imaging features to assist the echocardiographer with pattern recognition. Transthoracic echocardiography is an excellent screening and even diagnostic modality for these lesions. Saline contrast injection in a left upper extremity can identify an LSVC associated with Raghib syndrome. While TTE may be diagnostic for unroofed CS, tomographic imaging should be strongly considered to best characterize the lesion, especially for identification of a concomitant LSVC and presence of a bridging innominate vein. Transesophageal echocardiography can be useful for diagnosis but has the potential for misinterpretation and may perhaps be reserved for intraoperative imaging. In patients without contraindication, surgical repair is the cornerstone of management, to eliminate the left-to-right shunt, promote reverse remodeling, and result in significant functional improvement.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2023.12.030.

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