Unroofed Coronary Sinus Discovered Incidentally during Cardiac Surgery: Systematic Approach to Diagnosis by Transesophageal Echocardiography

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INTRODUCTION

First described in 1965 by Raghib *et al,*[1] unroofed coronary sinus (URCS), also known as the coronary sinus (CS) type of atrial septal defect (ASD), is a rare cardiac anomaly in which a communication occurs between the CS and the left atrium (LA) because of partial or complete absence of the common wall that separates the two. The defect or fenestration in the CS wall allows shunting of oxygenated left atrial blood via the CS ostium to the right atrium (RA), similar to an ASD. This anomaly is often associated with persistent left superior vena cava (PLSVC) connection to the CS.[1] The atrial-level left-to-right shunt through the CS causes dilatation of right-sided cardiac chambers and CS. While there has been one previous report of an URCS discovered incidentally at the time of coronary bypass surgery,[2] in that case the defect was not repaired and a comprehensive description of the method of diagnosis was not made. We report a case of URCS found incidentally and repaired at the time of coronary bypass surgery. In this report we provide a systematic two-dimensional (2D) and three-dimensional (3D) transesophageal echocardiographic (TEE) approach to this challenging diagnosis.

CASE PRESENTATION

A 61-year-old man with a history of type II diabetes mellitus, hypertension, smoking, and dyslipidemia developed progressively worsening angina pectoris and dyspnea on exertion. Coronary angiography revealed an 80% ostial stenosis of both the left anterior descending artery and the ramus intermedius not amenable to percutaneous intervention. His preoperative transthoracic echocardiogram (TTE) from an outside facility reported a normal left ventricular ejection fraction of 60% and no valve disease and was notable only for elevated right ventricular systolic pressure of 50 mm Hg. He was referred to our institution for coronary artery bypass surgery.

His blood pressure was 112/63 mm Hg. Routine intraoperative TEE was performed, which showed markedly dilated RA and right ventricle (Figure 1) suggestive of an atrial-level left-to-right shunt, but the precise location was elusive. Comprehensive systematic examination was done to rule out all causes of right heart dilation. There was no tricuspid (TR) or pulmonic regurgitation. There was a small ostium secundum ASD with trivial left-to-right shunt (Figure 1) that could not account for the significant dilatation of the right-sided chambers. There was no evidence of sinus venosus ASD of superior (SVC) or inferior vena cava (IVC) type (Figure 2), and pulmonary veins were connected normally (Figure 3). Severe dilation of CS was noted (Figures 4-6). Persistent left SVC was excluded by negative saline contrast imaging from left arm vein injection. Both the SVC and the IVC were connected normally (Figure 2), and coronary arteriovenous fistula to CS was excluded by lack of high-velocity continuous flow in CS (Figure 4). We performed midesophageal imaging with retroflexion of the probe and obtained the view of the CS. Color flow imaging and pulsed Doppler showed low-velocity early and late diastolic flow after atrial contraction typical of ASD shunt flow (Figure 4). Midesophageal views showed a dilated CS and its communication with the LA (Figure 5). Color flow imaging showed a left-to-right shunt in blue color, and saline contrast TEE confirmed small right-to-left shunt through the ostium of the URCS (Figure 5, Videos 1-5). Three-dimensional TEE in the mitral zoomed mode showed a severely dilated CS behind the LA (Figure 6). Real-time 3D TEE in four-chamber orientation showed a large 1 cm defect between the LA and CS or URCS (Figure 7, Video 6). Saline contrast real-time 3D TEE showed a small right-to-left shunt via the URCS (Figure 7, Video 7). A diagnosis of Xie type IIB URCS type of ASD[3] was made (Figure 8).

Revascularization surgery consisted of left internal mammary graft to the left anterior descending and a saphenous venous graft to the ramus intermedius on cardiopulmonary bypass using bicaval venous cannulation. Antegrade cardioplegia was used for myocardial protection. It was decided to surgically repair the ASD (see discussion). Surgical inspection confirmed that there was no evidence of PLSVC. Open inspection of the RA showed a small ostium secundum ASD remote from the ostium of the CS; this was repaired using 4-0 prolene suture. A probe was placed in the ostium of the CS, and

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VIDEO HIGHLIGHTS

Video 1: Midesophageal TEE view showing a severely dilated CS.

Video 2: Color flow imaging shows a high-volume flow through the ostium of the dilated CS into the RA due to left-to-right shunt via the URCS type of ASD.

Video 3: TEE midesophageal two-chamber view with a dilated CS behind the LA with communication between LA and CS or URCS.

Video 4: TEE midesophageal two-chamber view with color flow imaging showing left-to-right shunt from LA to CS in *blue color* via URCS.

Video 5: Biplane imaging of CS from midesophageal four- and two-chamber views with saline shows right-to-left shunt from CS to LA via URCS.

Video 6: Real-time 3D TEE from four-chamber orientation showing the communication of URCS between the LA and CS.

Video 7: Real-time 3D TEE from four-chamber orientation with saline contrast imaging shows right-to-left shunt from the URCS to LA.

Video 8: Real-time 3D TEE from four-chamber orientation after surgical patch repair shows intact patch.

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a URCS type of defect was found in the mid portion of the CS in the LA. An incision was made in the atrial septum close to the ostium of the CS, and the CS was unroofed back to the original defect and then repaired using a piece of autologous pericardium (Figure 9). Repeat postoperative TEE and real-time 3D TEE showed intact pericardial patch and no shunt by color Doppler and saline contrast TEE (Figures 10 and 11, Video 8). The patient had an uneventful recovery and was doing well at last follow-up 4 years later. A postoperative TTE with Doppler showed decreased size of right heart chambers and normal right atrial and pulmonary artery pressures (Figure 12).

DISCUSSION

The initial finding at the time of routine TEE that prompted further investigation was severe dilation of the right-sided cardiac chambers and normal dimensions of the left-sided chambers. The differential diagnosis includes TR or pulmonic valve regurgitation, a left-to-right shunt at the atrial level,[4] or partial, anomalous pulmonary venous connection.[5] Our approach began with the evaluation of the tricuspid and pulmonic valves, both of which were found to be competent. Next, we systematically excluded a defect in the atrial septum of sinus venosus type[4] and showed normal pulmonary venous connections.[5] The finding of a severely dilated CS can be caused by PLSVC connected to the CS,[6] anomalous pulmonary venous connection to the CS,[7] interruption of IVC with left hemiazygos vein connection to the CS,[8] coronary arteriovenous fistula to CS,[9] and, finally, URCS.[1-3,10-14] We began by excluding a PLSVC by negative saline contrast imaging from the left arm and exclusion of a coronary fistula to the CS by lack of high-velocity continuous flow in the CS. Normal connection of the IVC to the RA excluded left hemiazygos connection to the CS; therefore the remaining possibility was that of an URCS. Using a combination of 2D and 3D TEE, we were able to get excellent imaging of the dilated CS, ostium of URCS, and shunt flow using color Doppler, pulsed Doppler, and saline contrast techniques (Figures 5-7, Videos 1-7). This imaging sequence is useful for excluding other causes of dilated right heart and should arrive at the correct diagnosis regardless of the pathology involved. Cardiac computed tomography and magnetic resonance imaging have been used[10] to diagnose URCS syndrome. Our case, however, presented from outside for coronary bypass surgery. This report demonstrates that 2D and 3D TTE and TEE are powerful tools that can be utilized at bedside to arrive at a precise diagnosis.[11,12]

The indication for repair concomitant to the coronary bypass operation was to prevent further dilatation of right-sided chambers and to prevent possible future paradoxical embolization.[15] The TR velocity was incomplete, but outside TTE reported pulmonary arterial systolic pressure (PASP) of 50 mm Hg. Since there was dilation of right heart chambers (Figure 1), large left-to-right shunt on color Doppler (Figure 4, Video 2), trivial right-to-left shunt by saline contrast study (Figures 5, 7, Videos 5, 7), and PASP <2/3 of systemic pressure, it was decided to surgically repair the ASD. Follow-up TTE showed normalization of PASP (Figure 12).

Kirklin and Barratt-Boyes[13] classified the anatomical variants of URCS as follows: type I, completely URCS with a PLVCS; type II, completely URCS without PLVCS; type III, partially URCS in the mid-portion; and type IV, partially URCS in the terminal portion. To include all the variations of URCS syndrome, classification was modified by Xie and colleagues.[3] These different variants of URSC are illustrated in Figure 8. Our patient had a type IIB variant. The site of unroofing of CS and communication with the LA was higher than expected in our case, but the defect was confirmed on direct inspection during surgery. For patients with URCS without PLSVC (types II-IIIB) there are two basic options for surgical repair[13,14]: atrial septostomy and patching of the defect in the CS (Figure 9) or simply patching the ostium of the CS in the RA and allowing the trivial CS effluent to return to the LA. In type IB there is complete unroofing of the CS and the ostium functions as ASD. An easier option is to patch the ostium of the CS or perform a more technically demanding surgery of constructing a baffle to roof the CS using a piece of pericardium.[14] For patients with URCS with PLSVC (types I-IIIA), the first consideration is whether the PLSVC can be safely ligated. When there is a large, patent innominate vein bridging the PLSVC with the right-sided SVC and low pressure in the PLSVC, then ligation should be well tolerated and the patient is converted to the B-type classifications with the above-described strategies available. If the PLSVC cannot be safely ligated in the type IA variant, a baffle must be constructed using a piece of pericardium redirecting the PLSVC flow into the RA via the ostium of the CS.[14] In the type IIA and IIIA variants, when the PLSVC cannot be safely ligated, an atrial septostomy is made and the defect in the CS is patched.

CONCLUSION

We have described a rare case of URCS without PLSVC found incidentally at the time of myocardial revascularization surgery. This case demonstrates the power of 2D and 3D TEE echocardiography in narrowing the differential diagnosis of isolated right-sided chamber dilatation by systematically excluding progressively more complex etiologies. Using this approach, the correct diagnosis should

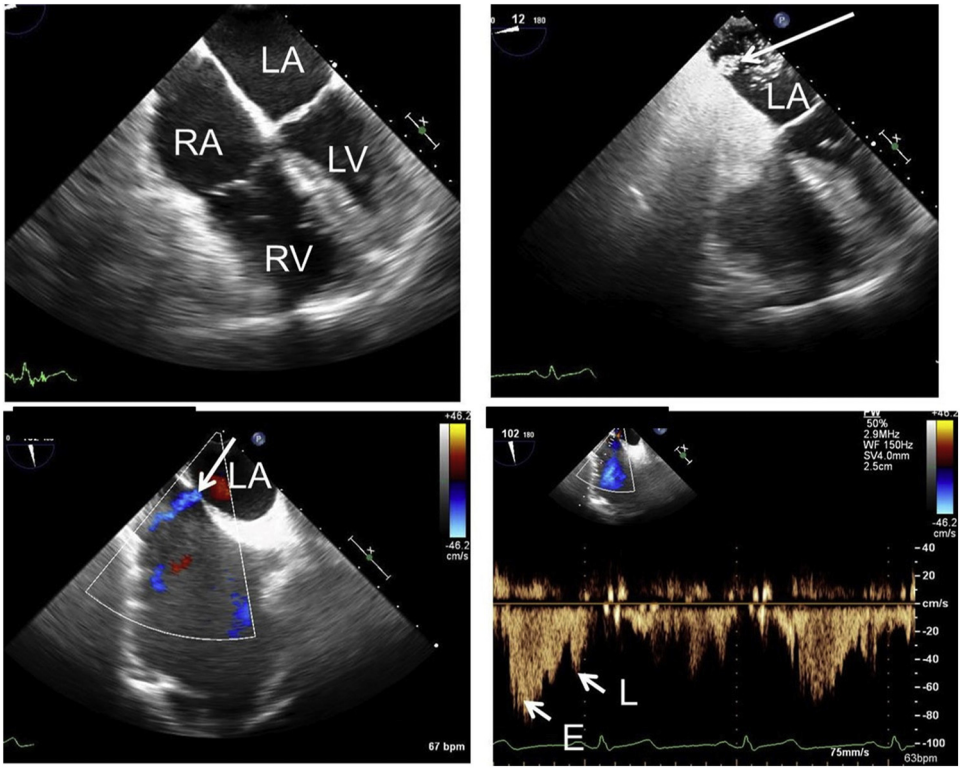

Figure 1 Midesophageal TEE images. *Top left*: Four-chamber view showing dilated RA and right ventricle (RV) and normal sized LA and left ventricle (LV). *Bottom left*: Atrial septal TEE view shows a small ostium secundum ASD (*arrow*). *Top right*: Four-chamber TEE with saline shows a small right-to-left shunt (*arrow*). *Bottom right*: TEE atrial septal view and pulsed Doppler through the left-to-right shunt shows typical early (E) and late (L) peaking flow pattern of ASD left-to-right shunt.

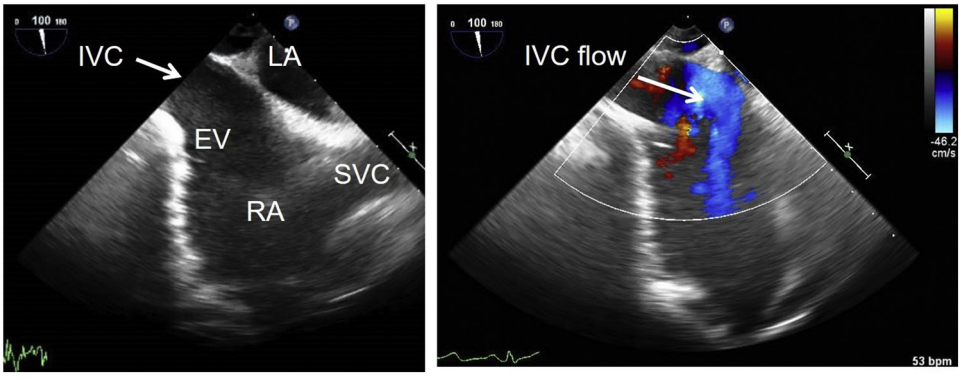

Figure 2 *Left panel*: Midesophageal TEE bicaval view with intact atrial septum at the level of the cava-atrial junction. *Right panel*: Normal flow from IVC. *EV*, Eustachian valve.

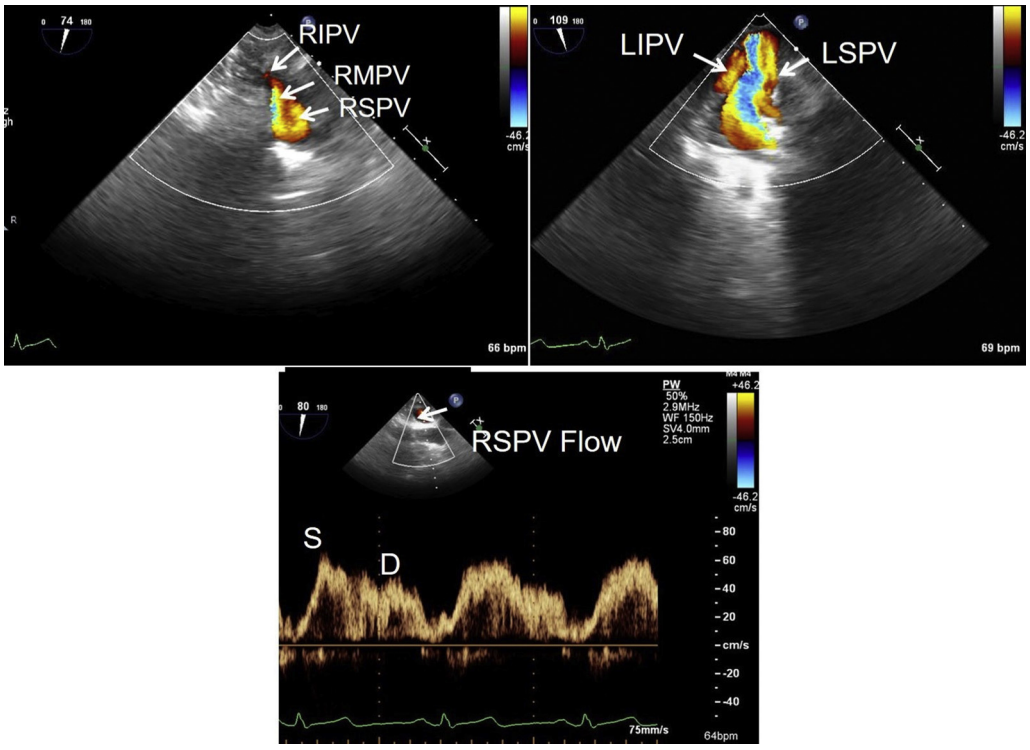

Figure 3 TEE images of normal connection of pulmonary veins, flow and pulsed Doppler pattern. *Top left panel*: Right superior pulmonary vein (RSPV), right middle pulmonary vein (RMPV), and a small right inferior pulmonary vein (RIPV). *Top right panel*: Larger left superior (LSPV) and a smaller left inferior pulmonary vein (LIPV). *Bottom panel*: Normal RSPV pulsed Doppler flow with a larger systolic (S) and a smaller diastolic (D) flow velocity.

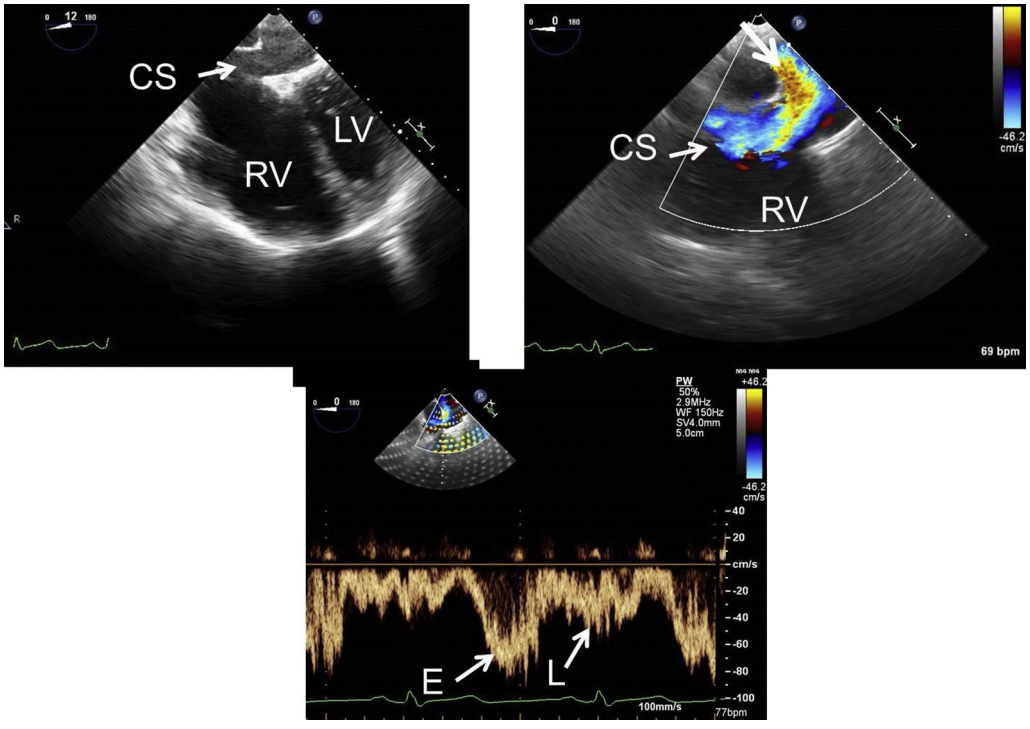

Figure 4 Midesophageal views of CS. *Top left panel*: Severely dilated CS and ostium (arrow). *Top right panel*: High-volume flow through the ostium into RA due to left-to-right shunt via the URCS type of ASD. *Bottom panel*: Pulsed Doppler interrogation of left-to-right shunt with low velocity early (E) and late diastolic (L) flow after atrial contraction, typical of ASD. *LV*, Left ventricle; *RV*, right ventricle.

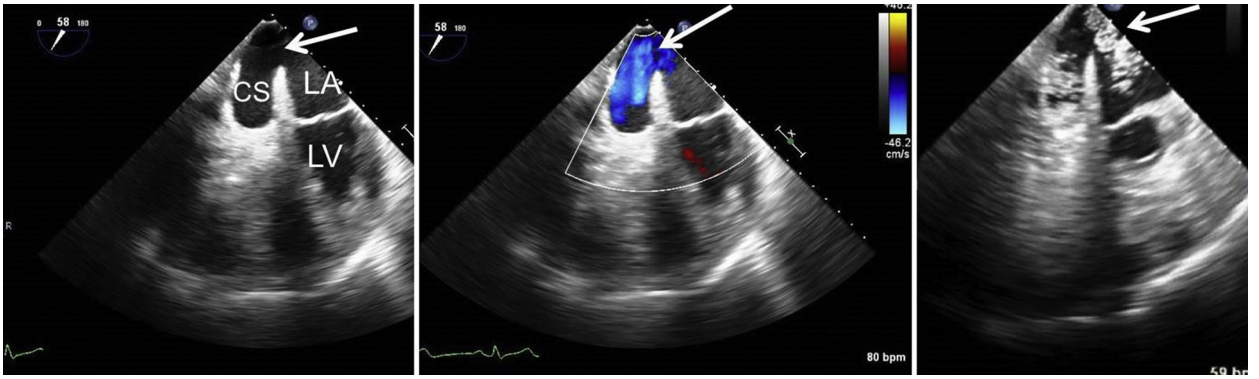

Figure 5 *Left panel*: Midesophageal two-chamber view with a dilated CS behind the LA with communication between LA and CS or URCS (*arrow*). *Middle panel*: Left-to-right shunt from LA to CS (*arrow*) through the URCS. *Right panel*: With saline, right-to-left shunt from CS to LA via URCS (*arrow*). *LV*, Left ventricle.

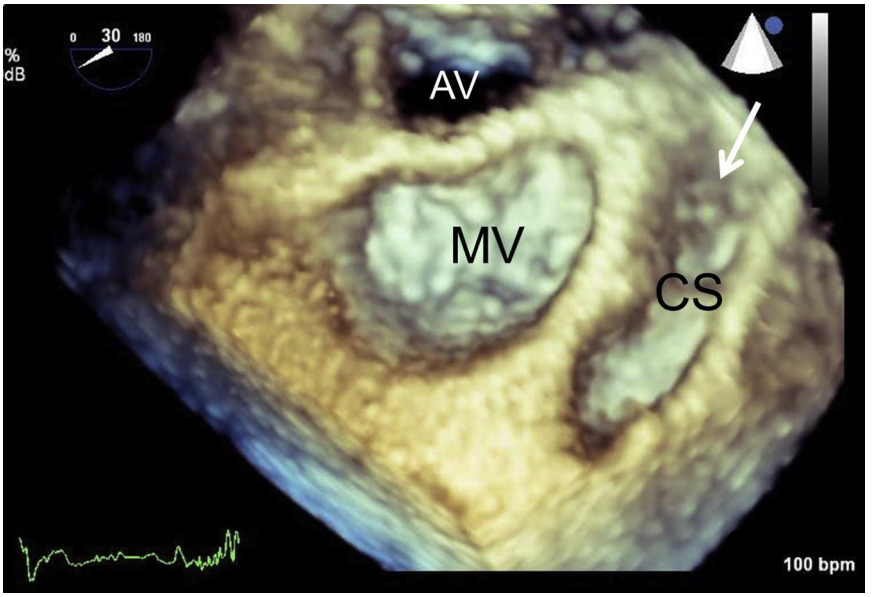

Figure 6 Mitral valve (MV) 3D TEE in zoomed mode showing a severely dilated CS and its ostium opening in RA (*arrow*). *AV*, Aortic valve.

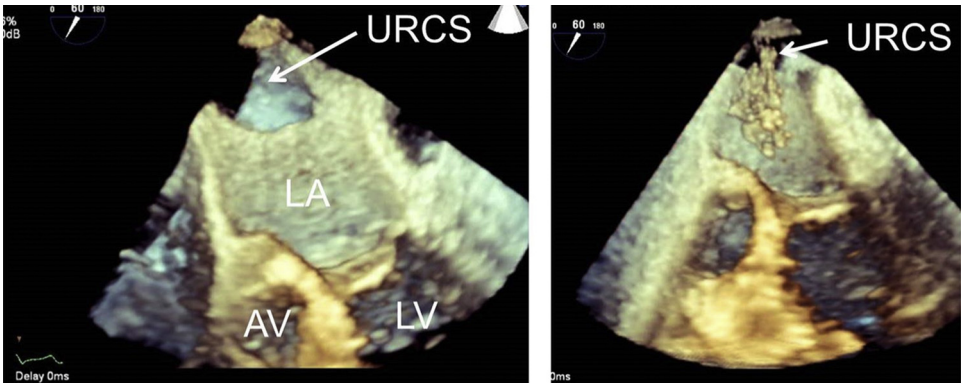

Figure 7 *Left panel*: Real-time 3D TEE from four-chamber orientation showing the communication between LA and CS or URCS (*arrow*). *Right panel*: With saline contrast, right-to-left shunt from URCS to LA (*arrow*). *AV*, Aortic valve; *LV*, left ventricle.

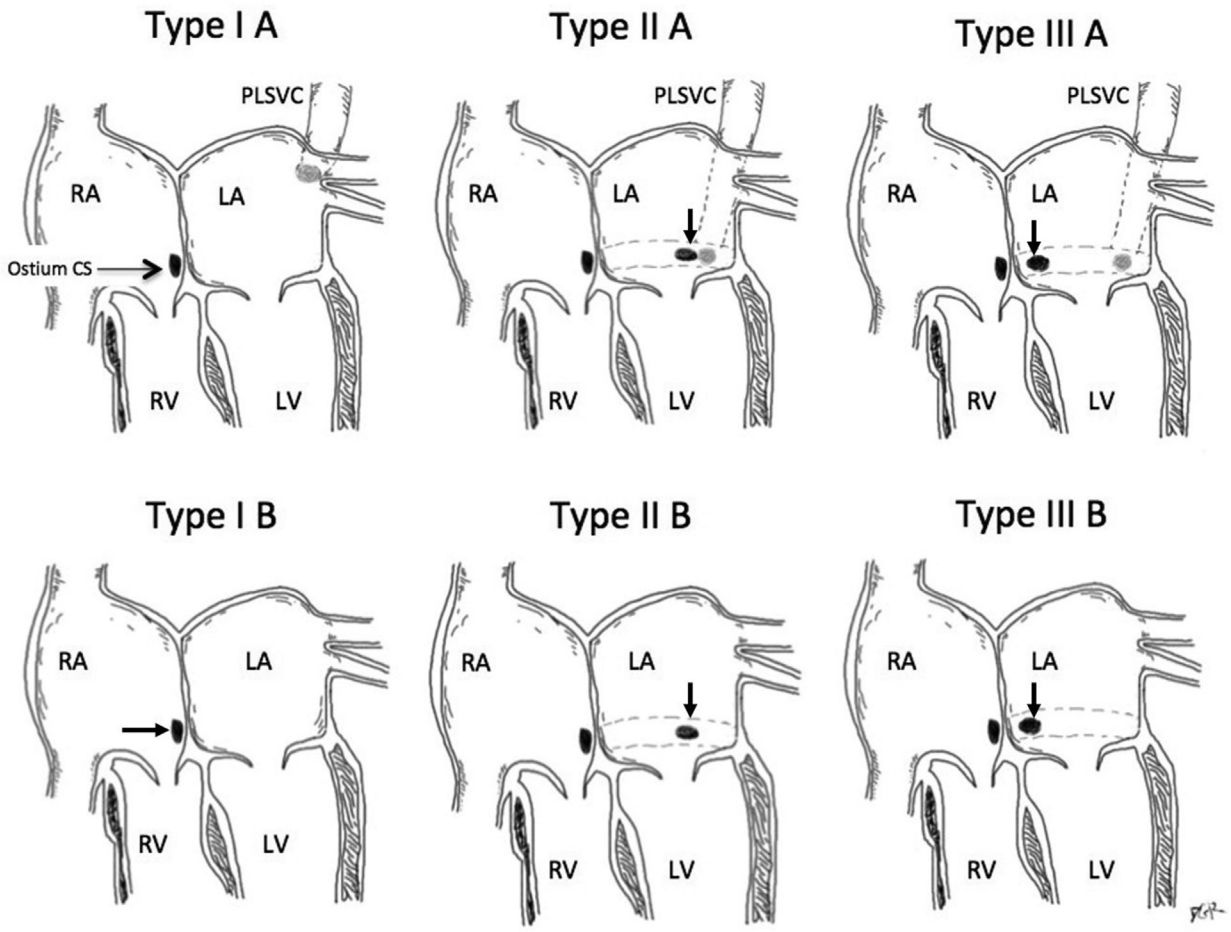

Figure 8 Classification of URCS. Type IA: completely absent CS with PLSVC, which connects to LA between left atrial appendage and left superior pulmonary vein. Type IB: completely absent CS, no PLSVC. Coronary sinus ostium serves as ASD (*arrow*) in both IA and IB. Type IIA: unroofing of the mid portion of CS (*arrow*) with PLSVC. Type IIB: unroofing of the mid portion of CS (*arrow*) without PLSVC. Type IIIA: unroofing of terminal portion of the CS (*arrow*) with PLSVC. Type IIIB: unroofing of the terminal portion of CS (*arrow*) without PLSVC. The position of the CS behind the LA is shown by the *dashed line*. This figure is partially based on illustrations from the article by Quaegebeur *et al.*[14] *LV*, Left ventricle; *RV*, right ventricle.

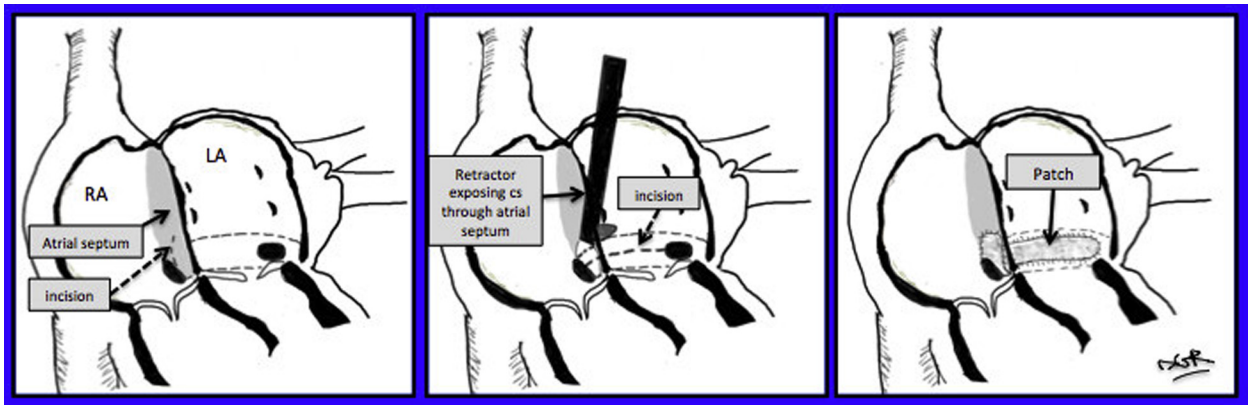

Figure 9 Technique used to repair the URCS. *Left panel*: The anatomy is demonstrated, and an incision was made in the atrial septum near the CS ostium. *Middle panel*: A retractor is used to expose the CS through the atrial septum. The incision used to complete the unroofing is shown. *Right panel*: The patch of autologous pericardium is shown repairing the CS as well as the incision in the atrial septum.

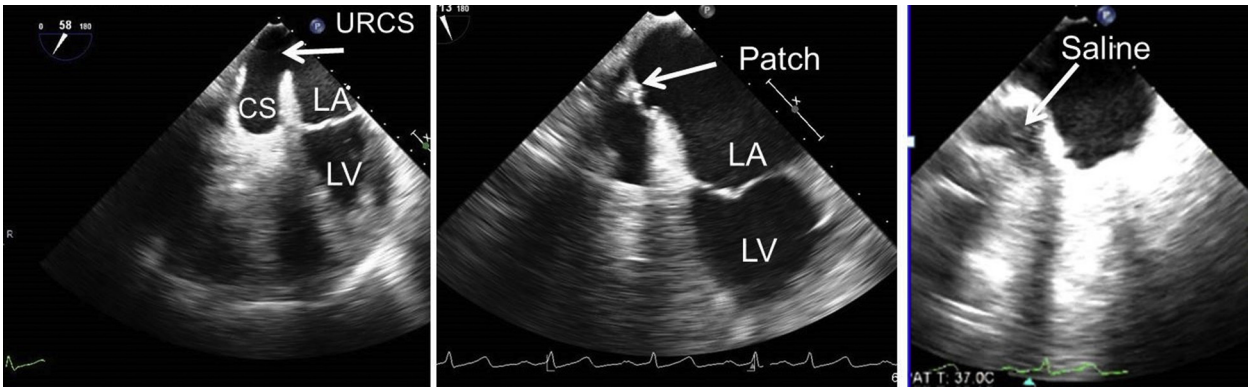

Figure 10 *Left panel*: TEE two-chamber view prior to surgery showing URCS (*arrow*). *Middle panel*: Intact patch after surgical patch repair (*arrow*). *Right panel*: After patch repair and saline contrast, right-to-left shunt is no longer seen. *LV*, Left ventricle.

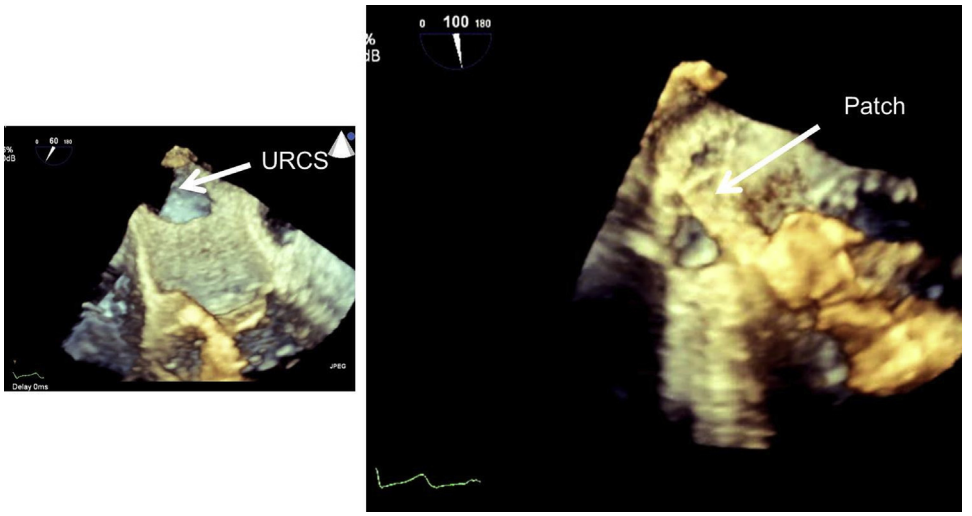

Figure 11 *Left panel*: Real-time 3D TEE from four-chamber orientation showing the communication between LA and CS or URCS (*arrow*). *Right panel*: Intact patch after surgical patch repair (*arrow*).

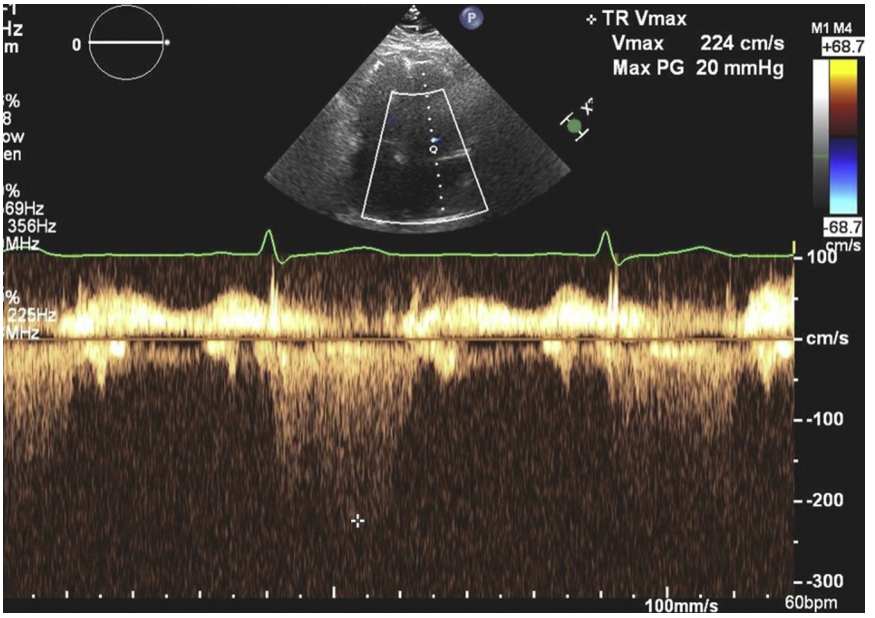

Figure 12 Postoperative study 4 years after surgery showed decreased right heart size, small IVC, estimated RA pressure of 5 mm Hg, trace TR, velocity of 2.2 m/sec, and PASP of 25 mm Hg.

be determined regardless of the etiology allowing for precise surgical planning.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2021.07.014.

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