CONGENITAL HEART DISEASE
NEVER TOO YOUNG OR TOO OLD TO BE DIAGNOSED WITH CONGENITAL HEART DISEASE

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

Ramesh C. Bansal, MD, FASE, Timothy P. Martens, MD, PhD, Hyayong Hu, MD, and David G. Rabkin, MD, Loma Linda, California

INTRODUCTION

First described in 1965 by Raghib et al., 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 ostium of the URCS 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 venous 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 ostium of URCS (Figure 7, Video 7). A diagnosis of Xie type IIb URCS type of ASD 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...
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.
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.

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

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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