Post Mitral Valve Replacement: Paravalvular Leak or Unroofed Coronary Sinus?

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INTRODUCTION

Unroofed coronary sinus (CS) is a rare congenital cardiac anomaly in which the CS communicates with the left atrium because of a partial or complete absence of its roof. It may be accompanied by other anomalies, such as a persistent left superior vena cava (PLSVC) draining to the CS.

We present the case of a 37-year-old woman with severe mitral regurgitation found to have a partial unroofed CS.

CASE PRESENTATION

The patient presented with symptoms of congestive heart failure due to severe mitral regurgitation and was brought to the operating room for mitral valve replacement. She had a history of mitral valve repair at the age of 4 years. Intraoperative transesophageal echocardiography (TEE) revealed severe central mitral regurgitation caused by a cleft in the anterior leaflet at the A2 position, with a vena contracta width of 1.07 cm, an effective regurgitant orifice area of 48 mm^2, systolic flow reversal in the pulmonary veins, and left atrial enlargement to 7 cm (Figure 1). There was biventricular dilation with preserved systolic function. The right atrium (5.5 cm) and CS were dilated (1.5 × 1.6 cm; Figure 2), but there was no evidence of a PLSVC.

A 6-cm right anterior thoracotomy was made in the fourth intercostal space. Cardiopulmonary bypass (CPB) was initiated via the right axillary artery and the right femoral vein, and mitral valve replacement was performed under cold fibrillatory arrest without cardioplegia. The valve was excised and replaced with a 29-mm St. Jude mechanical valve (St. Jude Medical, St. Paul, MN), with preservation of all posterior chords. During CPB, the surgeon noted venous blood within the left atrium, which was speculated to be from a patent foramen ovale not noted during prebypass TEE. After separation from CPB, postbypass TEE showed a bileaflet mechanical mitral prosthesis in the antianatomic position, with normal leaflet motion in both the two-dimensional midesophageal and three-dimensional views. Initially, there appeared to be a small paravalvular leak in addition to the normal washing jets (Figure 3), but further echocardiographic examination showed that the color flow originated from an unroofed CS with predominantly right-to-left shunting (Figure 4). The size of the defect measured less than 5 mm in diameter with no noticeable systemic oxygen desaturation from the right-to-left shunt. The decision was made that this unroofed CS was clinically insignificant, and therefore no further surgical intervention was undertaken. A review of recorded two-dimensional and three-dimensional prebypass images indicated the presence of the unroofed CS at the left atrioventricular groove (Figures 5 and 6, Video 1) but flow was not appreciated on the prebypass examination with color flow Doppler.

DISCUSSION

The CS is located posteriorly in the left atrioventricular groove and opens into the right atrium through its orifice in the posterior and inferior right atrial walls superior to the tricuspid valve.[1] The mean diameter of the CS is reported to be 7.05 ± 1.90 mm; a diameter >11 mm measured 3 cm proximal to the ostium is considered dilated and should therefore prompt careful imaging of the entire CS roof.[2] Using TEE, the long axis of the CS is imaged by turning slightly right and advancing or retroflexing the probe from the midesophageal four-chamber view. It is seen in short axis in the midesophageal two-chamber view just superior to the atrioventricular groove. The modified midesophageal bicaval view shows the orifice of the CS and is useful for guiding placement of percutaneous CS catheters during minimally invasive procedures.[3]

Unroofed CS, first reported by Raghib *et al.*[4] in 1965, is a rare congenital cardiac anomaly in which the CS communicates with the left atrium because of a partial or complete absence of its roof. Because it forms an interatrial communication, it is often grouped with true atrial septal defects. It may be accompanied by PLSVC draining to the CS. In a series published by Quaegebeur *et al.*,[5] 75% of patients with unroofed CS were found to have a PLSVC. The ostium of the CS is then frequently enlarged because of the increased blood flow.

Unroofed CS was classified by Ootaki *et al.*[6] into four morphologic types according to the partial or complete absence of the CS roof, location of the defect, and the presence or absence of a PLSVC: type 1, completely unroofed with PLSVC; type 2, completely unroofed without PLSVC; type 3, partially unroofed midportion of the CS; and type 4, partially unroofed terminal portion of the CS.

Unroofed CS has nonspecific clinical features, making diagnosis difficult. This anomaly should be suspected in a patient with PLSVC and events suggestive of intracardiac shunting, such as cerebral emboli or abscess or unexplained arterial oxygen desaturation. Symptoms are related to the size of the defect and the resulting interatrial shunt, which may cause right heart failure.

Management depends on the morphology of the lesion and the degree of interatrial shunting. In general, small defects with diameters of <5 mm and no evidence of right ventricular volume overload do not require closure unless they cause paradoxical embolism.

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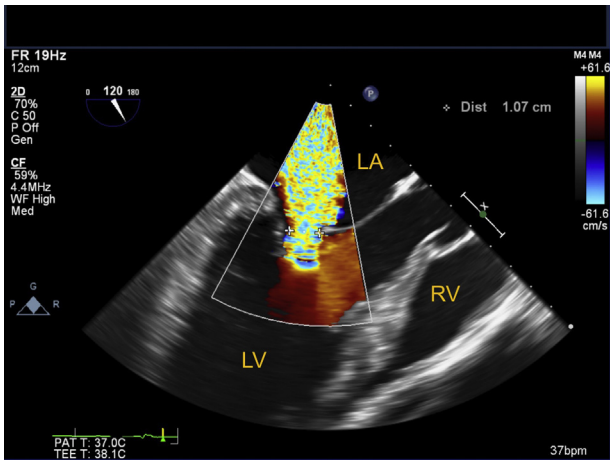

Figure 1 Midesophageal long-axis view showing severe mitral regurgitation with a vena contracta width of 1.07 cm. *LA*, Left atrium; *LV*, left ventricle; *RV*, right ventricle.

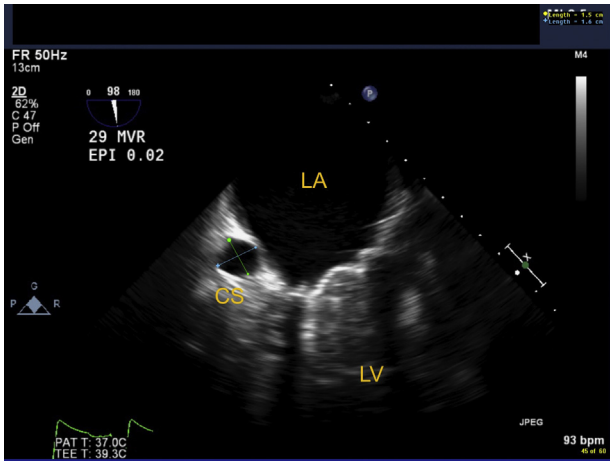

Figure 2 Midesophageal two-chamber view showing dilated CS. *LA*, Left atrium; *LV*, left ventricle.

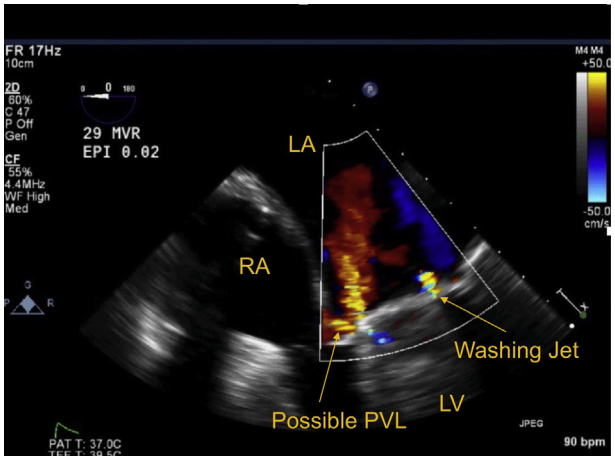

Figure 3 Midesophageal four-chamber view showing normal washing jet and a regurgitant jet outside the sewing ring, possibly indicating a paravalvular leak. *LA*, Left atrium; *LV*, left ventricle; *PVL*, paravalvular leak; *RA*, right atrium.

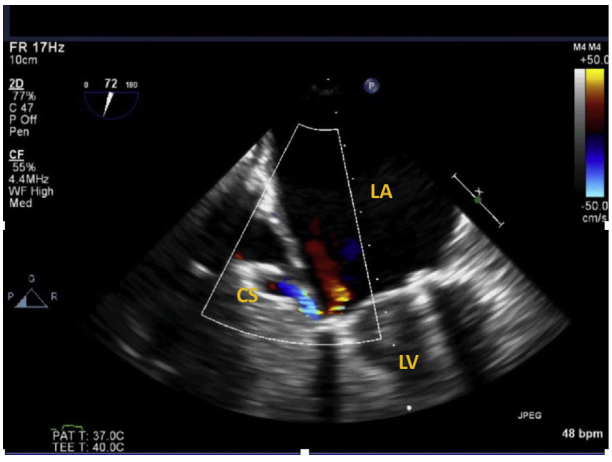

Figure 4 Midesophageal commissural view demonstrating direct communication from unroofed CS to left atrium (LA). *LV*, Left ventricle.

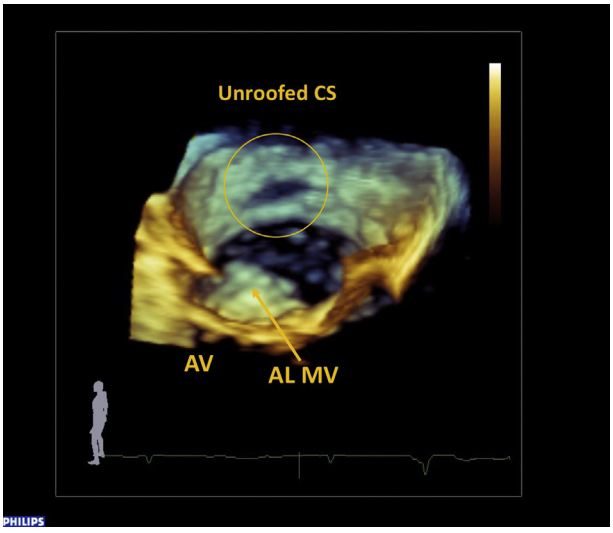

Figure 5 Three-dimensional volume rendering demonstrating the unroofed CS along the posterior atrioventricular groove at the level of P2–P3 leaflets of the mitral valve. *AL MV*, Anterior leaflet of the mitral valve; *AV*, aortic valve.

Larger defects with evidence of right ventricular volume overload on echocardiography usually cause symptoms only after 30 years of age, and closure is often indicated to prevent long-term complications. Unroofed CS should be repaired surgically rather than by a percutaneous approach, in which case patch reconstruction of the CS roof is usually performed.[7]

CONCLUSION

Thorough transesophageal echocardiographic interrogation should be performed before and after CPB. In this case, the unroofed CS was not recognized before CPB, because of the high left atrial pressure and severe mitral regurgitation, obliterating a significant right-to-left shunt from the unroofed CS and making interrogation with color flow Doppler limited. After replacement of the mitral valve and discontinuation of CPB, the unroofed CS was unmasked, from the

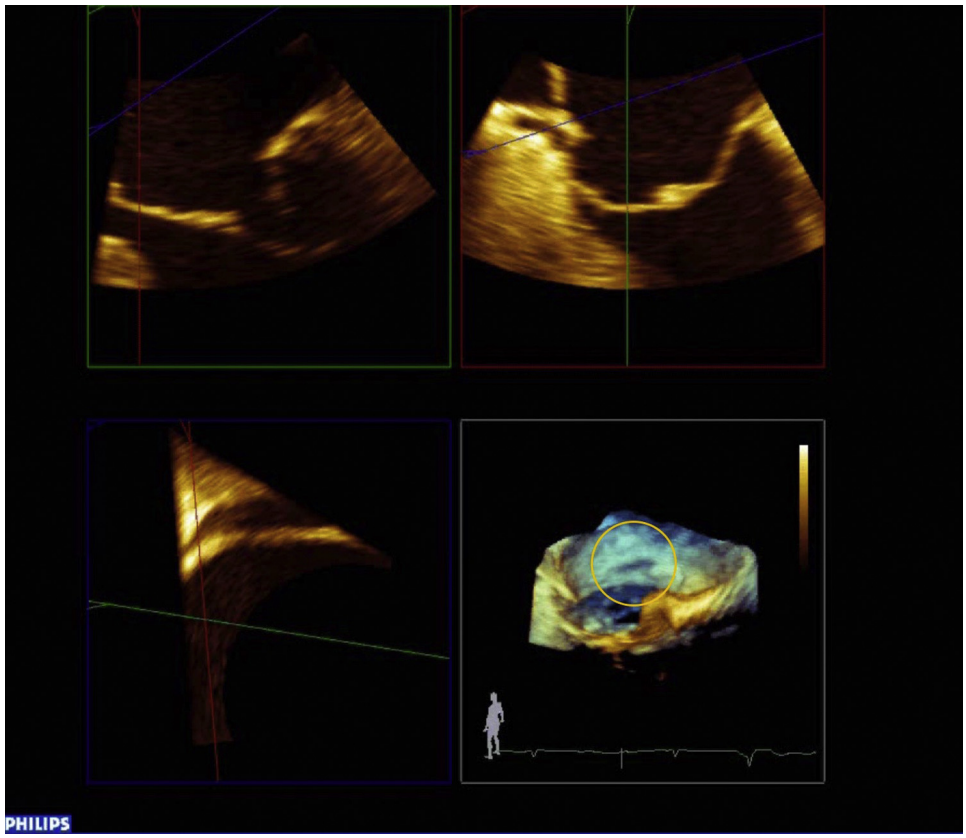

Figure 6 Multiplanar reconstruction views using Philips 3D software showing the unroofed CS in the two-dimensional and three-dimensional views.

decrease in left atrial pressure. Small defects with insignificant shunting and without evidence of right ventricular volume overload can be determined to be clinically insignificant and do not require surgical intervention.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.case.2017.07.009.

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