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 posterior chords. During CPB, the surgeon noted venous blood draining to the CS. In a series published by Quaegebeur et al.4 in 1965, 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.
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.

**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

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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.
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.

REFERENCES

1. Adatia I, Gittenberger-de Groot AC. Unroofed coronary sinus and coronary sinus orifice atresia. Implications for management of complex congenital heart disease. J Am Coll Cardiol 1995;25:948-53.

2. Shah SS, Teague SD, Lu JC, Dorfman AL, Kazerooni EA, Agarwal PP. Imaging of the coronary sinus: normal anatomy and congenital abnormalities. Radiographics 2012;32:991-1008.

3. Sun T, Fei HW, Huang HL, Chen OD, Zheng ZC, Zhang CJ, et al. Transeophageal echocardiography for coronary sinus imaging in partially unroofed coronary sinus. Echocardiography 2014;31:74-82.

4. Raghib G, Ruttenberg HD, Anderson RC, Amplatz K, Adams P Jr, Edwards JE. Termination of left superior vena cava in left atrium, atrial septal defect and absence of coronary sinus. Circulation 1965;31:906-18.

5. Quaegebeur J, Kirklin JW, Pacifico AD, Barger LM Jr. Surgical experience with unroofed coronary sinus. Ann Thorac Surg 1979;27:418-25.

6. 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.

7. Warnes CA, Williams RC, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. Circulation 2008;118:2345-95.