A 44-year-old woman visited our clinic because of sudden onset pain in both lower extremities. After admission, computed tomography (CT) was performed and revealed occlusion of both popliteal arteries and renal infarction in the left kidney. Transthoracic echocardiography (TTE) revealed rheumatic mitral stenosis (Figure 1A). In the apical view of TTE and on transesophageal echocardiography, we identified a huge dilated coronary sinus (CS) (Figure 1B) and detected large defect between the dilated CS and left atrium (LA) (Figure 1C, Movie 1). On contrast

Shin-Jae Kim, MD, PhD, Soe Hee Ann, MD, Yong-Giun Kim, MD, Gyung-Min Park, MD, PhD, Ki-Bum Won, MD, PhD, and Sang-Gon Lee, MD, PhD

Cardiovascular Center, Ulsan University Hospital, Ulsan, Korea

A Case of Unroofed Coronary Sinus with Coronary Sinus Orifice Atresia: Use of Multimodality Imaging

Figure 1. Transthoracic echocardiography revealed mitral stenosis and unroofed coronary sinus. (A) Doming of the anterior mitral leaflet and limited motion of the posterior mitral leaflet were demonstrated. (B) In the apical 4-chamber view with tilting, a huge dilated CS was identified. (C) With counterclockwise rotation of the probe, a 22-mm large defect between the dilated CS and the LA was detected. (D) Agitated saline was injected via the left antecubital vein. On contrast echocardiography, the RA was filled with air bubbles, but not the dilated CS, and then the right ventricle was filled with air bubbles later. This finding showed no persistent left superior vena cava. There was no negative jet in the RA, which raised the possibility of CS orifice atresia. CS: coronary sinus, LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle.
Echocardiography with agitated saline, the right atrium (RA) was filled with air bubbles, but not the dilated CS, and then the right ventricle was filled with air bubbles later (Figure 1D). This finding showed no persistent left superior vena cava (PLSVC), which was supported by the CT imaging finding (Figure 2A). On CT, a huge dilated CS was observed, showing no communication with the RA, which suggested CS orifice atresia (Figure 2B). On real-time three-dimensional (3D) echocardiography in the en face view of the LA, an ovoid-shaped defect was found between the CS and the LA, posterior to the mitral valve (Figure 3A). After horizontal rotation of the image, no communication between the CS and the RA was visible, which suggested CS orifice atresia (Figure 3B). The patient was discharged after stabilization.

Unroofed coronary sinus (UCS) is a spectrum of cardiac anomalies in which part or all of the common wall between the CS and the LA is absent. Seventy-five percent of the UCS is associated with PLSVC. Atresia of the RA orifice of the CS is rare. Most cases of CS orifice atresia are associated with an alternative exit for coronary venous blood return, such as a small PLSVC, large thebesian vein, or CS canal defect, like in the present case. Myocardial ischemia is unlikely as long as an alternate exit exists for CS blood. Multimodality imaging such as, TTE, transesophageal echocardiography, contrast echocardiography, 3D echocardiography, CT, or magnetic resonance imaging is needed to diagnose this anomaly.
like in the present case. In case of operation, the flow of CS blood should be retained. Deaths due to myocardial ischemia after ligation of a PLSVC were reported.  

**SUPPLEMENTARY MATERIAL**

**Movie 1**
On transesophageal echocardiography in 110° view, a defect was found between the dilated coronary sinus and the left atrium, and very slow flow through the defect was observed with spontaneous echo contrast.

Click here to view

**REFERENCES**

PubMed | Crossref

PubMed | Crossref

PubMed | Crossref

PubMed