A 6-month-old girl was referred for surgical repair of tetralogy of Fallot. A transthoracic echocardiogram revealed a large, unrestrictive ventricular septal defect, mild aortic override, a normal pulmonary valve with mild infundibular obstruction (peak gradient 22 mm Hg), a small patent ductus arteriosus, secundum atrial septal defect, and persistence of the left superior vena cava to the coronary sinus. The preoperative ECG showed sinus rhythm, mean frontal plane axis of 130°, right ventricular hypertrophy, and no signs of preexcitation (Figure 1). A chest x-ray showed cardiomegaly and prominent pulmonary vascularity with mild hyperinflation (Figure 2). Under cardiopulmonary bypass, the patient underwent repair consisting of resection of multiple infundibular muscle bundles, patch closure of the ventricular septal defect, suture closure of atrial septal defect, and ligation of the patent ductus arteriosus. She had an uncomplicated recovery, but a routine postoperative echocardiogram before discharge showed an echolucent mass measuring 16×22 mm, posterior and inferior to the left ventricle, causing a localized wall-motion abnormality (Figure 3 and Movies I and II). Because of concerns about a localized pericardial collection, she underwent surgical exploration with a view to drainage. On direct inspection, however, no pericardial fluid collection was found. Instead, there was a cystic lesion intimately related to the posterior aspect of the left ventricle. It was nonpulsatile and covered by epicardium. Transesophageal echocardiogram confirmed that this was the area in question (Figure 4 and Movie III).

Cardiac MRI was performed with a 1.5-Tesla Siemens Sonata scanner. Cine imaging (Figure 5 and Movie IV) and in-plane phase contrast (Figure 6 and Movies V and VI) revealed flow into the mass from the coronary sinus, and the left superior vena cava to the coronary sinus was demonstrated (Figure 7 and Movie VII). Time-of-flight angiography after gadolinium contrast injection into the left external jugular vein showed first-pass contrast into the mass, indicating a large coronary sinus diverticulum (Figure 8). The patient was discharged on aspirin and on follow-up remained asymptomatic, without arrhythmias or ECG evidence of preexcitation.

Coronary sinus diverticula are rare and mostly detected in adults undergoing retrograde angiography of the coronary sinus during electrophysiological studies for posterior septal accessory pathways. These pathways frequently have short refractory periods that predispose to rapid ventricular response during atrial fibrillation, thus increasing the risk of sudden death. A review of the literature of all right atrial malformations from 1955 to 1998 found 28 patients with coronary sinus diverticula. Cases in children are rare, with only 5 reports, 4 of which were not associated with Wolff-Parkinson-White syndrome. We report a case in a child with congenital heart disease and a left superior vena cava to coronary sinus, without evidence of preexcitation or arrhythmias. The diagnosis was made by cardiac MRI using cine and phase-contrast imaging, with confirmation of the drainage by time-of-flight angiography. In the absence of symptoms or arrhythmias, conservative treatment is recommended.

Disclosures

None.

References

1. Funabashi N, Asano M, Komuro I. Giant coronary sinus diverticulum with persistent left superior vena cava demonstrated by multislice computed tomography. Int J Cardiol. 2006;111:468–469.
Figure 1. Preoperative ECG showing sinus rhythm, axis of 130°, right ventricular hypertrophy, and no signs of preexcitation.

Figure 2. Chest x-ray showing cardiomegaly and prominent pulmonary vascularity with mild hyperinflation. A nasogastric tube is seen.

Figure 3. A, Transthoracic echocardiogram in the parasternal long axis, showing an echolucent collection posterior and inferior to the left ventricle (see Movie I). B, Transthoracic echocardiogram in the subcostal short axis showing an echolucent collection posterior and inferior to the left ventricle (see Movie II). LV indicates left ventricle; LA, left atrium; CSD, coronary sinus diverticulum; and CS, coronary sinus.

Figure 4. Transesophageal echocardiogram in the transgastric view showing the coronary sinus diverticulum posterior to the left ventricle (see Movie III). Abbreviations as in Figure 3.
Figure 5. Cardiac magnetic resonance short-axis oblique cine image of the coronary sinus diverticulum (see Movie IV). RV indicates right ventricle. Other abbreviations as in Figure 3.

Figure 6. A, In-plane magnitude phase image in the coronal plane demonstrating flow into the coronary sinus diverticulum (see Movie V). B, In-plane phase velocity image in the coronal plane demonstrating flow into the coronary sinus diverticulum (see Movie VI). CSD indicates coronary sinus diverticulum; RA, right atrium.
Figure 7. Cardiac magnetic resonance cine image of the left superior vena cava to coronary sinus (see Movie VII). LSVC indicates left superior vena cava; RA, right atrium.

Figure 8. Series of time-resolved magnetic resonance angiograms in the coronal plane after gadolinium injection into the left external jugular vein, showing entry of contrast into the coronary sinus diverticulum on first phase, confirming its connection to the coronary sinus. Abbreviations as in Figures 3 and 6.
Cardiac Magnetic Resonance Imaging of a Coronary Sinus Diverticulum Associated With Congenital Heart Disease
Edythe B.C. Tham, David B. Ross, Michael Giuffre, Jeffrey Smallhorn and Michelle L. Noga

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