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Case Report

Partially Unroofed Coronary Sinus

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Abstract

Unroofed coronary sinus is a rare congenital cardiac abnormality characterized by the partial or complete absence of the roof of the coronary sinus. Unroofed coronary sinus causes a shunt between the inferior left atrium and coronary sinus, which can lead to pulmonary hypertension and heart failure. Unroofed coronary sinus is typically associated with other congenital defects, most commonly a persistent left superior vena cava. We present the cardiac magnetic resonance imaging findings of a 6-year-old child with a rare unroofed coronary sinus requiring surgical closure.

Keywords: congenital heart disease, magnetic resonance imaging, unroofed coronary sinus

INTRODUCTION

Unroofed coronary sinus (URCS) is a rare congenital cardiac defect characterized by an abnormal connection between the coronary sinus and the left atrium due to the partial or complete absence of the roof of the coronary sinus.[1] Unroofed coronary sinus is the rarest form of atrial septal defect (ASD). These defects may cause nonspecific clinical symptoms; however, accurate diagnosis is important as URCS has also been associated with the risk of pulmonary hypertension, brain abscess, or cerebral emboli.[1,2] We herein discuss the imaging of a child who required URCS closure at 6 years of age.

CASE PRESENTATION

A 5-month-old Caucasian male infant with the diagnosis of an ASD, ventricular septal defect (VSD), and patent ductus arteriosus (PDA) had been referred for surgical repair due to failure to thrive with congestive heart failure symptoms. On post-operative day 2 after ASD/VSD closure and PDA ligation, he was found to be hypoxic with oxygen saturation in the low 80s on high fraction of inspired oxygen. Transthoracic echocardiogram

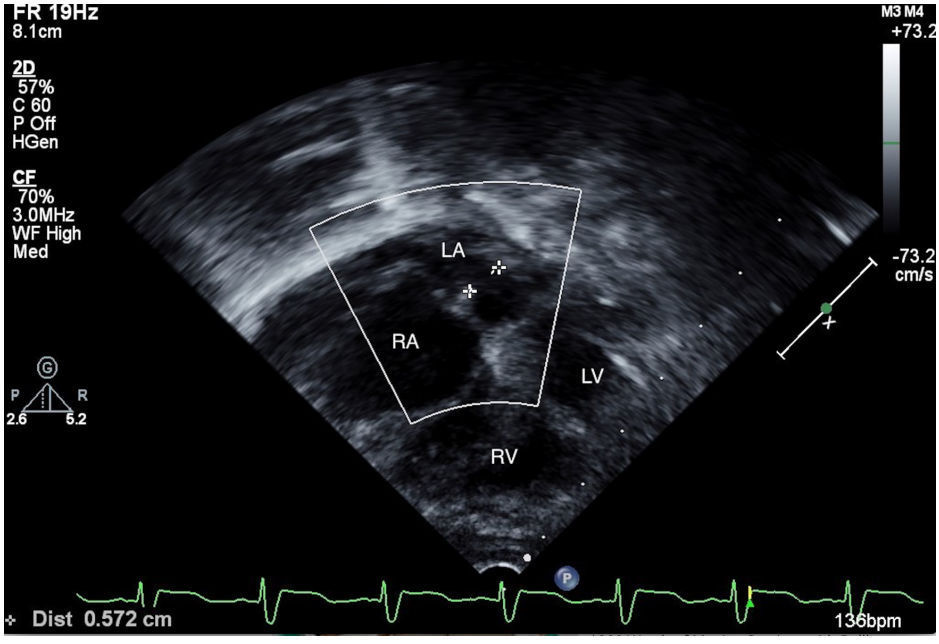

Figure 1. Transthoracic echocardiography demonstrating unroofed coronary sinus between (+) markers measuring 5.7 mm. LA, left atrium; RA, right atrium; LV, left ventricle; RV, right ventricle.

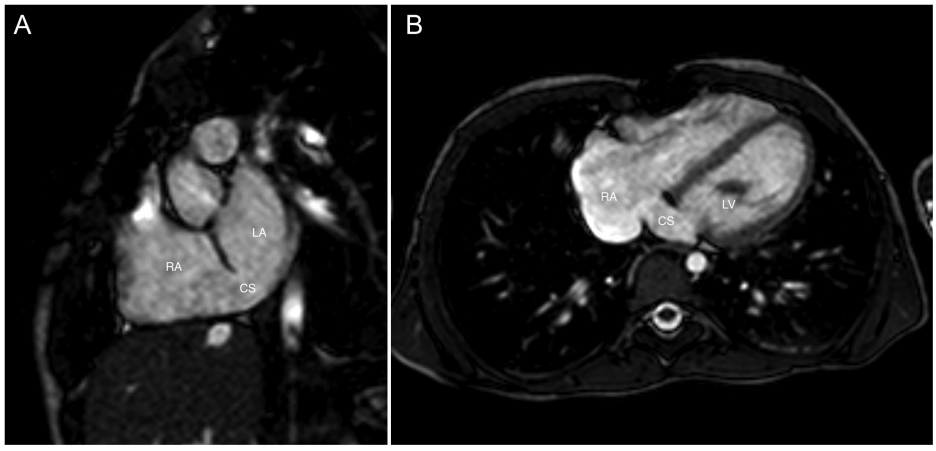

Figure 2. Cine balanced turbo field echo MR images in short axis view at the level of atrioventricular groove (A) and four-chamber (B) views show a dilated coronary sinus ostium with defect in the wall between the coronary sinus and left atrium. There is no PLSVC. LA, left atrium; RA, right atrium; CS, coronary sinus; PLSVC, persistent left superior vena cava.

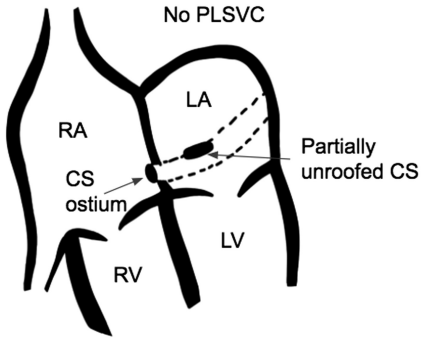

Figure 3. Schematic drawing of this case showing a type 3 partially unroofed mid-portion of the coronary sinus with no PLSVC. CS, coronary sinus; RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; PLSVC, persistent left superior vena cava.

revealed a URCS with bidirectional shunting (Figure 1). Interventional cardiology performed right and left heart catheterization and angiography, which demonstrated unroofing of the mid-portion of the coronary sinus with drainage into the left atrium. There was no evidence of a persistent left superior vena cava (PLSVC).

At 6 years of age, the patient was referred to cardiovascular surgery for URCS closure due to a gradual increase in right ventricular pressure seen on sequential echocardiograms. Cardiac magnetic resonance imaging (MRI) was performed to further evaluate the previous findings for surgical planning. Cardiac spin-echo and cine MRI confirmed the previous findings of URCS with the absence of mid-portion of common wall between the coronary sinus and left atrium (Figures 2 and 3). Dilated coronary sinus ostium was measured at 10 mm. There was mild dilatation of the right atrium and no significant dilatation of the right ventricle. Flow assessment by phase contrast velocity mapping measured Qp : Qs of 1.3 : 1. The patient underwent URCS closure with no intraoperative complications.

MAIN POINTS

- A rare case of unroofed coronary sinus requiring surgical closure was presented in this report.
- The importance of magnetic resonance imaging in diagnosis of unroofed coronary sinus was provided.
- Use of magnetic resonance imaging in surgical planning of unroofed coronary sinus was recommended.

DISCUSSION

Unroofed coronary sinus is a rare congenital cardiac anomaly characterized by a connection between the coronary sinus and the left atrium. It is the rarest form of ASD, representing less than 1% of all the ASDs.[1] The coronary sinus normally drains the cardiac veins into the right atrium (Figure 4). Unroofed coronary sinus is due to incomplete formation of the left atriovenous fold during embryonic development.[4] Isolated URCS, as in this patient, is rare as this defect has a strong association with a PLSVC, occurring in 75% of cases.[1,3]

Kirklin and Barratt-Boyes defined 4 types of URCS: type 1, completely unroofed with PLSVC; type 2, completely unroofed without PLSVC;

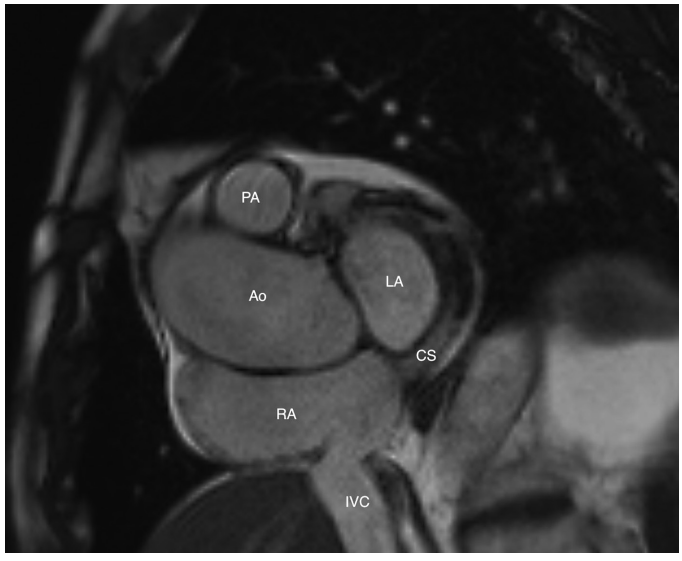

Figure 4. Cardiac MRI demonstrating normal coronary sinus draining into the right atrium. LA, left atrium; RA, right atrium; CS, coronary sinus; Ao, aorta; PA, pulmonary artery; IVC, inferior vena cava; MRI, magnetic resonance imaging.

type 3, partially unroofed mid-portion (as demonstrated in this case); and type 4, partially unroofed terminal portion.[5,6]

The symptoms associated with URCS are determined by the size of the defect and the degree of left-to-right shunting.[1] Clinical symptoms may range from asymptomatic to severe dyspnea with right-sided heart failure due to pulmonary hypertension.[1] Furthermore, because the coronary sinus is connected to the left atrium, there is a risk for paradoxical emboli such as cerebral abscesses.[2] Diagnosis of URCS is challenging because the clinical symptoms are nonspecific.

Prior to the use of advanced cardiac imaging, diagnoses such as URCS would only be possible during surgery or at autopsy. Although echocardiography is typically the first line in evaluating ASDs, the coronary sinus is a posterior structure that can be difficult to visualize.[3] Kim et al found echocardiography could easily visualize a dilated coronary sinus but not URCS in 5 out of 11 patients. Meanwhile, CT and MRI accurately demonstrated that all the patients had a defect in the coronary sinus.[7] Computed tomography and MRI are non-invasive imaging tests that have proven useful and more accurate in the diagnosis of URCS.[3,4,7]

When comparing CT and MR imaging for the diagnosis of URCS, both techniques have their advantages. Computed tomography imaging provides better anatomic information, but MRI shows high contrast resolution visualization of deep structures of the heart without the need for intravenous contrast or ionizing radiation.[3] With phase-contrast velocity-encoded cine technique, MRI also allows for flow measurement of the shunt created by URCS. This technique is performed on an imaging plane perpendicular to the long axes of the aorta and main pulmonary artery, then selecting the area of interest on cine images.[7] The ratio of pulmonary to systemic flow (Qp : Qs) can then be calculated. Although flow information can also be obtained through cardiac catheterization, MRI is non-invasive and carries significantly fewer risks than a surgical procedure.

When evaluating for URCS on CT and MRI, the coronary sinus view, depicted by the cardiac short-axis view in the plane of the atrioventricular groove, provides the best visibility.[7] Both CT and MRI provide important 3D information about the anatomy of the URCS and play an important part in surgical planning.

Management of URCS is dependent on the severity of clinical symptoms. Transcatheter techniques are typically preferred over open-heart surgeries due to their less invasive nature.[1] Surgical repair of URCS is performed by simple closure of the defect. In this case, the patient underwent successful open repair of the URCS.

CONCLUSION

We present a rare case of URCS without PLSVC that required surgical closure in a 6-year-old child. Although echocardiography can aid in the diagnosis of URCS, cardiac MRI offers definitive diagnosis and high-resolution visualization of defects. Cardiac MRI provides important anatomical and flow measurement information that may be used in surgical planning, which can reduce the need for invasive procedures such as catheterization.

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Declaration of Interests: The authors declare that they have no competing interest.

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