

## A Rare Cause of Hypoxemia

### Uma Causa Rara de Hipoxemia

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### Abstract

An unroofed coronary sinus is a very rare cardiac anomaly that is commonly associated with a persistent left superior vena cava. Its diagnosis is often difficult, but clinical signs and echocardiographic findings of an interatrial shunt should raise its suspicion. The authors describe the case of a child with chronic hypoxemia whose diagnostic work-up revealed an unroofed coronary sinus and persistent superior left vena cava.

### Introduction

Unroofed coronary sinus (UCS), characterized by defective formation of the coronary sinus (CS), is a congenital heart disease (CHD) that was first described in 1965 by Raghbi et al.<sup>1</sup> Classified as an atrial septal defect (ASD), it is, in fact, not a true defect of the atrial septum; rather, it is characterized by interatrial shunting across the ostium of the CS.<sup>2</sup> The rarest type of ASD,<sup>3</sup> it accounts for less than 1% of all lesions associated with interatrial shunting.<sup>1,2</sup> Seventy-five percent of cases of UCS are associated with persistent left superior vena cava (LSVC),<sup>1</sup> the most commonly reported congenital systemic venous connection anomaly.<sup>4,5</sup> The incidence of persistent LSVC is 0.3–0.5% among healthy individuals, and as many as 10% of patients with CHD.<sup>5</sup> The complex of UCS and persistent LSVC occurs more frequently in congenitally malformed hearts, particularly with heterotaxy syndromes.<sup>6</sup> A UCS is considered an “occult” type of ASD<sup>1</sup> that might be difficult to diagnose because of its nonspecific clinical features.<sup>2,7</sup> Affected patients may be completely asymptomatic for a long time or show signs of right-sided heart failure induced by chronic right ventricular volume overload.<sup>3,7</sup> Even in the current era of multimodality imaging, saline contrast echocardiography (SCE) is an established simple, inexpensive, and effective diagnostic tool in such these cases.<sup>6</sup>

### Case Report

A 7-year-old child with chronic hypoxemia was referred to our department for a cardiovascular evaluation. Her past medical history was unremarkable.

The physical examination was normal except for a mildly reduced peripheral oxygen saturation (90% on room air).

### Keywords

Contrast echocardiography; Coronary sinus; Hypoxemia.

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A 15-lead surface electrocardiogram showed normal sinus rhythm with no evidence of cardiac chamber enlargement or hypertrophy.

Transthoracic echocardiography (TTE) revealed a dilated CS, suggesting the presence of an LSVC. The right atrium and right ventricle were also mildly dilated. An SCE was performed to confirm our findings. Agitated saline was given via the left cubital vein. Microbubbles first appeared in the CS and then filled the left atrium, confirming the diagnosis of UCS with a persistent LSVC. The patient underwent cardiac catheterization (VÍdeo 1), that revealed a bidirectional shunt through the UCS between the left and right atria with a normal pulmonary artery pressure.

### Discussion

A UCS is a rare congenital cardiac anomaly in which there is partial or complete absence of the roof of the CS, resulting in a communication between the CS and the left atrium.<sup>2</sup> It is often associated with a persistent LSVC<sup>2,3</sup> and other cardiac abnormalities such as atrial isomerism, ASD, atrioventricular septal defect, mitral stenosis, mitral atresia, tricuspid atresia, and tetralogy of Fallot.<sup>1</sup> The clinical presentation of UCS depends on CS defect size and the degree of shunting across it.<sup>3,7</sup> Mild desaturation as well as an increased risk of paradoxical cerebral embolism and brain abscess occur as a result of the right-to-left shunt.<sup>8,9</sup> The left-to-right shunt causes chronic right-sided volume overload that may lead to symptoms of right-sided heart failure.<sup>3,9</sup> The diagnosis of UCS should be considered in the evaluation of an unknown cardiac murmur, right-sided heart enlargement, transient cyanosis or hypoxia, or paradoxical embolism.<sup>3</sup> However, its diagnosis remains challenging.<sup>6</sup> Our patient presented with signs of bidirectional shunt (hypoxemia and enlarged right heart chambers) that were confirmed by cardiac catheterization.

Although TTE is the most widely used non-invasive technique, using it to diagnose UCS might be difficult because of its limited ability to evaluate the posterior structures of the heart.<sup>7,9</sup> A dilated CS on TTE should raise the suspicion of a persistent LSVC, but the CS diameter alone does not help the differentiation between those with and those without UCS.<sup>9</sup> SCE is an inexpensive, easily available, and easily reproducible diagnostic modality that may identify an unusual cause of cyanosis in appropriate clinical context if it is performed and interpreted carefully.<sup>6</sup> In a normal subject, agitated saline microbubbles do not cross the pulmonary capillaries; thus, in the absence of a right-to-left shunt, microbubbles remain confined to the right side of the heart (“negative contrast echocardiogram”);<sup>6</sup> in this case report, the appearance of microbubbles in the left atrium and ventricle led to the diagnosis of a UCS with right-to-left shunt (“positive contrast echocardiogram”) (VÍdeo 2).<sup>6</sup>

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Importantly, however, when a persistent LSVC with UCS is suspected, the agitated saline should be injected into the left arm;<sup>10</sup> otherwise, this diagnosis may be missed.

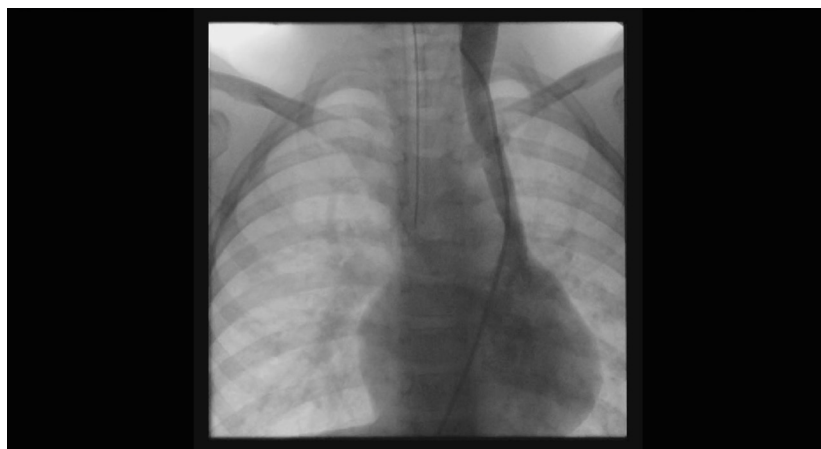
### Conclusion

Unexplained arterial oxygen desaturation and signs of a significant left-to-right shunt (enlarged right heart chambers)

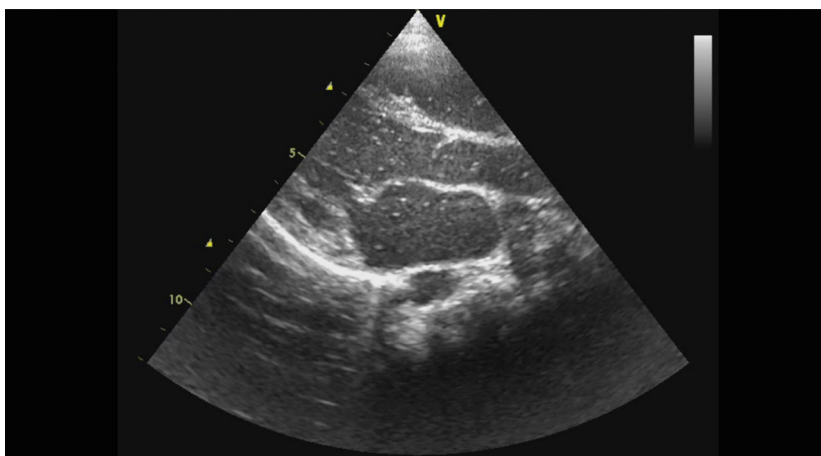
in the presence of a dilated CS should raise the suspicion of a persistent LSVC with UCS. The diagnosis can be easily confirmed with SCE with contrast injected into the left arm.

### Conflict of interest

The authors have declared that they have no conflict of interest.



**Video 1** – Catheter venography was performed in the internal jugular vein showing venous return through persistent left vena cava superior draining into the coronary sinus, with early left atrium and ventricle opacification, confirming the diagnosis of unroofed coronary sinus.



**Video 2** – Saline contrast echocardiography was performed with the injection in the left cubital vein, during which the microbubbles first appeared in the coronary sinus and then filled the left atrium, confirming the diagnosis of unroofed coronary sinus.

### References

1. Cintează E, Filip C, Duică G, Nicolae G, Nicolescu A, Bălgrădean M, Unroofed coronary sinus: update on diagnosis and treatment. Rom J Morphol Embryol. 2019;60(1):33–40.
2. Murli L, Ranjit MS, Shah P. Unroofed coronary sinus: An unusual interatrial communication and a rare childhood entity. Ann Pediatr Cardiol. 2019;12(1):64-5.

3. Thangarooan M, Truong Q, Kalra M, Yared K, Abbara S. Rare Case of an Unroofed Coronary Sinus Diagnosis by Multidetector Computed Tomography. *Circulation*. 2009; 119(16):e518–e520.
4. Goyal I S, Punnam S, Verma G, Ruberg F. Persistent left superior vena cava: a case report and review of literature. *Cardiovasc Ultrasound*. 2008;6:50.
5. Kurtoglua E, Cakinb O, Akcayc S, Akturkd E, Korkmaza H. Persistent Left Superior Vena Cava Draining into the Coronary Sinus: A Case Report. *Cardiol Res*. 2011;2(5):249–52.
6. Gupta S, Shetkar S, Ramakrishnan S, Kothari SS. Saline Contrast Echocardiography in the Era of Multimodality Imaging – Importance of “*Bubbling It Right*”. *Echocardiography*. 2015;32(11):1707-19.
7. Bonardi M, Valentini A, Camporotondo R. Unroofed coronary sinus and persistent left superior vena cava: a case report. *J Ultrasound*. 2012;15(3):179–82.
8. Kong P, Ahmad F. Unroofed coronary sinus and persistent left superior vena cava. *Eur J Echocardiogr*. 2007;8(5):398-401.
9. Pérez Matos A, Planken R, Bouma B, Groenik M, Backx A, Winter R, et al. Unroofed coronary sinus newly diagnosed in adult patients after corrected congenital heart disease. *Neth Heart J*. 2014;22(5):240–5.
10. Thaiyananthan N, Jacono F, Patel S, Kern J, Stoller J. Right to left anatomic shunt associated with a persistent left superior vena cava. The importance of injection site in demonstrating the shunt. *Chest*. 2009;136:617–20.