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CASE REPORT

Unroofed Coronary Sinus Syndrome Presenting with Early Pulmonary Hypertension

Maria E Franco-Fuenmayor, MD[1*], Numra A Aleem, MD[1], Jorge D Salazar, MD[2] and Ashraf M Aly, MD[3]

[1]*Department of Pediatrics, University of Texas Medical Branch, USA*

[2]*Division of Pediatric Cardiovascular Surgery, University of Texas McGovern Medical School and Children's Memorial Hermann Hospital, USA*

[3]*Division of Pediatric Cardiology, Department of Pediatrics, University of Texas Medical Branch, USA*

***Corresponding author:** *Maria E Franco-Fuenmayor, Department of Pediatrics, University of Texas Medical Branch, 301 University Blvd. RB6 Room 3.230, Galveston, TX, 77555, USA*

Abstract

Isolated partial or complete absence of the coronary sinus roof hemodynamically behaves like an atrial septal defect and rarely causes pulmonary hypertension. We report a case of unroofed coronary sinus (URCS) presenting with post-operative cyanosis due to pulmonary hypertensive crisis following surgical repair of a ventricular septal defect at 6 months of age. He had progressively decreased stamina with gradual increase in pulmonary pressure requiring surgical closure of the URCS by 6 years of age with resolution of symptoms and normalization of the RV pressure.

Keywords

Coronary sinus defect, Pulmonary hypertension, Congenital heart disease, Congenital heart surgery

Introduction

URCS is a rare congenital heart defect (CHD) commonly associated with persistent left superior vena cava (LSVC), VSD, cortriatriatum, tetralogy of Fallot, total anomalous pulmonary venous connections and heterotaxy [1]. Clinical presentation depends on the associated CHD, though when isolated it behaves like an ASD. This defect increases the risk for paradoxical emboli, brain abscess or atrial arrhythmias [2]. As with other types of ASD, it is unlikely for URCS to present with early pulmonary hypertension.

Case

A two-week-old male infant was referred for evaluation of a heart murmur. An echocardiogram (echo) showed a large membranous VSD and a dilated CS without persistent LSVC. At 5 weeks, he developed congestive heart failure managed initially medically but required surgical repair at 6 months of age. Post-operatively, he developed cyanosis and underwent cardiac catheterization that revealed a right to left shunt across a large defect in the CS roof "Qp/Qs ratio of 0.6 and mildly elevated right atrial (RA) and right ventricular (RV) end-diastolic pressures (8 mmHg) and elevated pulmonary artery pressure to 60 mmHg. The pulmonary resistance was also increased to 3.8 Woods units. The pulmonary pressure gradually decreased and the URCS was monitored clinically [3]. The pulmonary hypertension was initially managed with inhaled nitric oxide and supplemental oxygen and later with oral Sildenafil for 3 months. He did relatively well except for some growth delay, progressive exercise intolerance and snoring with a gradual increase in RV pressure. He underwent tonsillectomy and adenoidectomy to exclude upper airway obstruction as the cause at 5 years of age. Sequential echocardiograms demonstrated gradual increases in RV pressure. An echo done at 6 years revealed a left to right shunt across a large size defect (14 mm) between the CS and LA (Figure 1 and Figure 2). A 30 mmHg TR jet ve-

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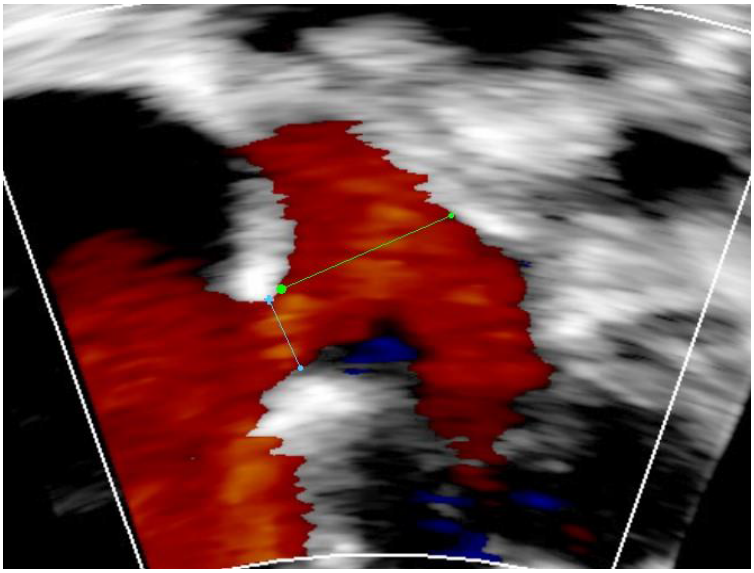

Figure 1: A 4-chamber color Doppler echo showing flow through large URCS (14 mm) and a dilated CS orifice (7 mm) into RA.

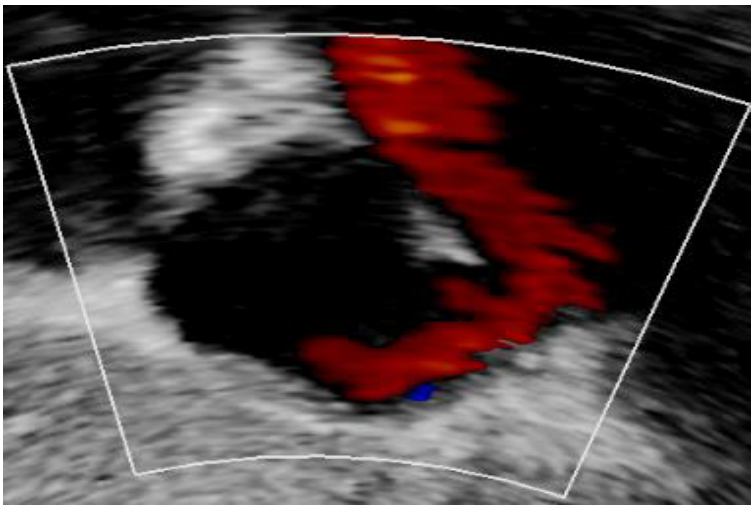

Figure 2: A subcostal color Doppler echo showing flow from LA through URCS to RA.

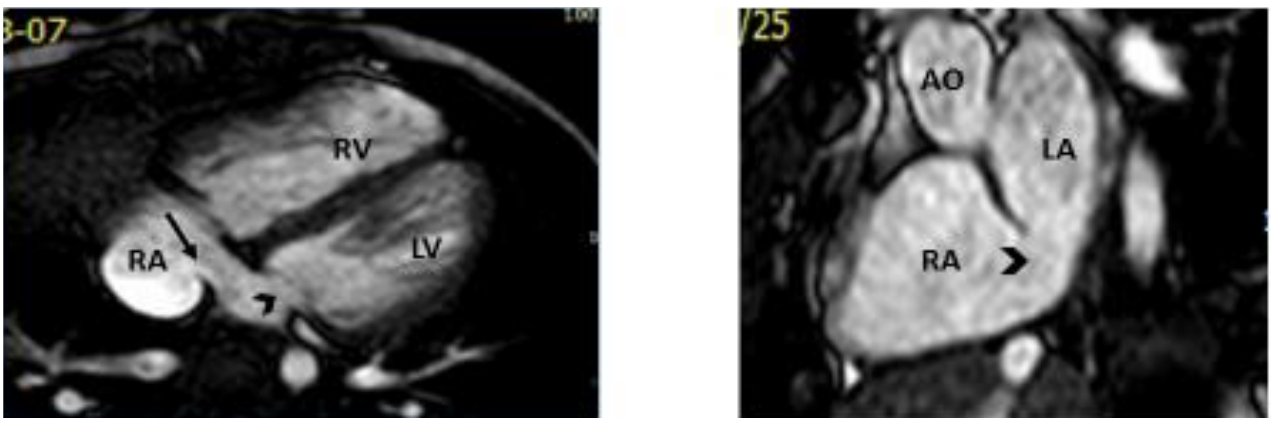

Figure 3: Steady-state free precession images showing CSASD (arrow heads) with absence of common wall that separates the CS and LA. Dilated ostium of CS (arrow) towards RA.

RV: Right ventricle; LV: Left ventricle; LA: Left atrium; RA: Right atrium; Ao: Aorta.

locity suggested an estimated RV pressure of about 35 mmHg. A cardiac MRI confirmed the findings (Figure 3).

He underwent successful surgical repair of the URCS at 6 years of age. Intra-operatively, three openings (4

mm each) were found at the roof of the CS communicating with the LA. The "re-roofing" procedure involved baffling of the 3 coronary venous ostia from the LA to the CS opening into the RA. The baffle material used was a novel pre-shaped, curved pericardial patch (30° Cardiocel, Admeus) that facilitates baffling without obstruction or patch shrinkage. A follow-up echo showed no residual defects and normal RV pressure. He gradually regained stamina and weight.

Discussion

The atrial septum develops in the 5th week of gestation. The coronary sinus is a venous channel that drains the coronary venous blood into the RA. It runs in the atrioventricular groove behind the LA. An URCS occurs when there is a lack of separation from the inferior wall of the LA and the roof of CS allowing for a communication "unroofing". Isolated URCS is rare. Since its first description in 1965 [2], four morphologic types have been identified. These include: Type I with completely unroofed CS with LSVC, type II with completely unroofed CS without LSVC, type III with partially unroofed CS in the mid portion, and type IV with partially unroofed CS in the terminal portion. Our patient likely has a type III unroofed CS. These defects could be missed with echo and are often diagnosed intra-operatively or at necropsy [4,5]. They should be considered if the CS is dilated especially in the presence of LSVC and right heart dilatation. Although saline contrast study may be helpful, cardiac MRI or CT and in some cases cardiac catheterization may be needed for diagnosis [6]. The pathophysiology of URCS is consistent with a left to right atrial shunt. The clinical presentation depends on the defect size and the presence of other cardiac anomalies. Chronic right heart volume overload and pulmonary over-circulation eventually causes pulmonary hypertension over the span of decades [7].

Pulmonary hypertension in children with congenital heart disease remains a complex condition. The pathophysiology varies amongst pre-tricuspid and post-tricuspid lesions. Pre-tricuspid shunts are low pressure leading to right sided volume overload and pulmonary over circulation without immediate increase in pulmonary pressure. The magnitude of the shunt is dependent on size and relative ventricular compliance. Recent research estimates the development of pulmonary hypertension at 6-17% beyond the fourth decade of life [8]. In contrast, post-tricuspid shunts are high pressure, depend on relative pulmonary and vascular resistances, and may lead to the development of pulmonary hypertension in early childhood.

Our patient did not follow the expected pattern as he developed pulmonary hypertension at an early age. This could be attributed to having residual pulmonary vascular injury from the left to right shunt due to early VSD, which may explain the early postoperative pulmonary hypertensive crisis. It appears that the URCS plays a role as there was a gradual increase in the RV pressure that eventually normalized after intervention.

Conclusion

URCS is rare and thus may be overlooked in transthoracic echo. It should be suspected in patients with dilated CS especially if associated with persistent LSVC and right heart dilatation. Although contrast echo may be helpful, other imaging such as cardiac MRI/CT or even cardiac catheterization may be needed to establish the diagnosis. To our knowledge, this is the first case report of an URCS that presented with pulmonary hypertension in early childhood.

Acknowledgments

We confirm that the manuscript has been read and approved by all named authors and that there are no other persons who satisfied the criteria for authorship.

Disclosures

We confirm that there are no known conflicts of interest nor financial support associated with this work.

Author's Statement

We confirm that written permission was granted by patient's mother.

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