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# Congenital Left Ventricle-to-Coronary Sinus Fistula: A Rare Isolated Anomaly of the Coronary Sinus

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## INTRODUCTION

Congenital anomalies of the coronary sinus (CS) are very uncommon. They can occur either in isolation or in association with other congenital heart defects.<sup>1</sup>

Among those anomalies, congenital fistulae connecting the left ventricle and the CS represent a very rare entity. Most reported cases of CS fistula generally originate in the right coronary artery or left anterior descending coronary artery system.<sup>2</sup>

We describe an extremely rare case of an isolated high-pressure congenital left ventricle-to-CS fistula as an anomaly of the CS in an otherwise asymptomatic child. The initial clinical presentation and follow-up, imaging studies, and plan for management are presented along with a review of relevant literature.

## CASE PRESENTATION

A 2-month-old male patient, born by cesarean section at 39 weeks without complications during pregnancy or birth, was referred to our cardiology department for a harsh systolic murmur noted by his general pediatrician during a well-child visit. At the time of presentation, he was asymptomatic, without reported shortness of breath or cyanosis. He was healthy, growing, and developing appropriately.

On further examination, the patient had a quiet precordium, and the apex was not displaced. Auscultation of the heart revealed normal heart sounds with a III/VI systolic murmur, best heard at the left lower sternal border. He had normal 2+ peripheral pulsations.

A complete two-dimensional sector scan, color flow Doppler, and spectral Doppler echocardiographic examination was performed, showing a 2-mm patent foramen ovale and high-velocity turbulent flow in the dilated CS. The jet originated from the posterior-inferior aspect of the left ventricular free wall and ended in the lateral aspect of the CS. The waist of the jet measured 2 mm in diameter, and velocity was just over 4 m/sec, suggestive of restrictive flow with a pressure gradient of 65 to 75 mm Hg. The flow from the CS then took an almost 90° turn and was seen entering the right atrium in an unobstructed fashion (Figure 1, Video 1).

Cardiac catheterization was performed at the age of 3 months. The study demonstrated flow from the left ventricle into a smooth-walled posterior and inferior chamber in the region of the atrioventricular sul-

cus, which appeared to be the CS. A small amount of contrast was seen entering the right atrium from the CS. The degree of shunting in this region was minimal by angiography (Figure 2, Video 2). The size and systolic function of both the right and left ventricles were normal. The right and left coronary arteries were also normal in appearance. The posterior descending coronary branch arose from the right coronary artery. No mitral valve regurgitation was seen. The pulmonary-to-systemic flow ratio was 1.1:1.

A 24-hour Holter monitor was placed at the age of 4 months, 11 months, and then annually. All have demonstrated normal sinus rhythm without ectopic beats or arrhythmias. Echocardiographic studies were also performed at 6- to 12-month intervals. The waist measurements of the CS fistula and the estimated pressure gradient stayed stable over 5 years. The sizes of the right atrium, right ventricle, and left ventricle remain within normal limits.

Overall, the patient continues to be asymptomatic, gaining weight appropriately without any signs of heart failure or diastolic dysfunction.

## DISCUSSION

Congenital left ventricle-to-CS fistula is rare. The mechanism of such defect from an embryologic standpoint has not been fully described. In fact, and because the CS arises from the left horn of the sinus venosus, it should have no communication with the left ventricle, which has a quite separate embryologic origin. Acquired fistula between the left ventricle and CS are more commonly documented. Previous repeated mitral valve replacement,<sup>3</sup> myocardial infarction,<sup>4</sup> and rarely catheter ablation<sup>5</sup> were the major etiologies of the occurrence of the acquired shunts.

Mantini *et al.*<sup>6</sup> analyzed necropsy specimens and classified CS anomalies into various categories. A review of the subject by Shah *et al.*<sup>1</sup> suggested a modification of the Mantini classification in which high-pressure CS fistula could be due not only to a coronary artery-to-CS fistula but also to a left ventricle-to-CS fistula. We are aware of only a single case of isolated congenital left ventricle-to-CS fistula documented in the literature by Gnanapragasam *et al.*<sup>7</sup> in 1989.

The limited number of reported cases of dilated CS due to left ventricular fistula have otherwise been associated with additional cardiac defects, all of which required closure. Percutaneous closure in the cardiac catheterization laboratory is theoretically possible through the CS approach. Fontes Pedra *et al.*<sup>8</sup> found that the use of three-dimensional computed tomography aided in subsequent catheter-based closure of the coronary fistula. Surgical patch closure of this has been successfully described with good short-term results and minor postoperative morbidity.<sup>9</sup>

We suggest in asymptomatic infants with this isolated abnormality that noninvasive monitoring is acceptable as opposed to surgical or catheter-based repair. This monitoring should include periodic echocardiographic examinations with electrocardiography and 24-hour Holter monitoring. Considerations for surgical repair would include evidence of significant left-to-right shunt with signs of right and/or left ventricular volume or pressure overload, arrhythmias, or those

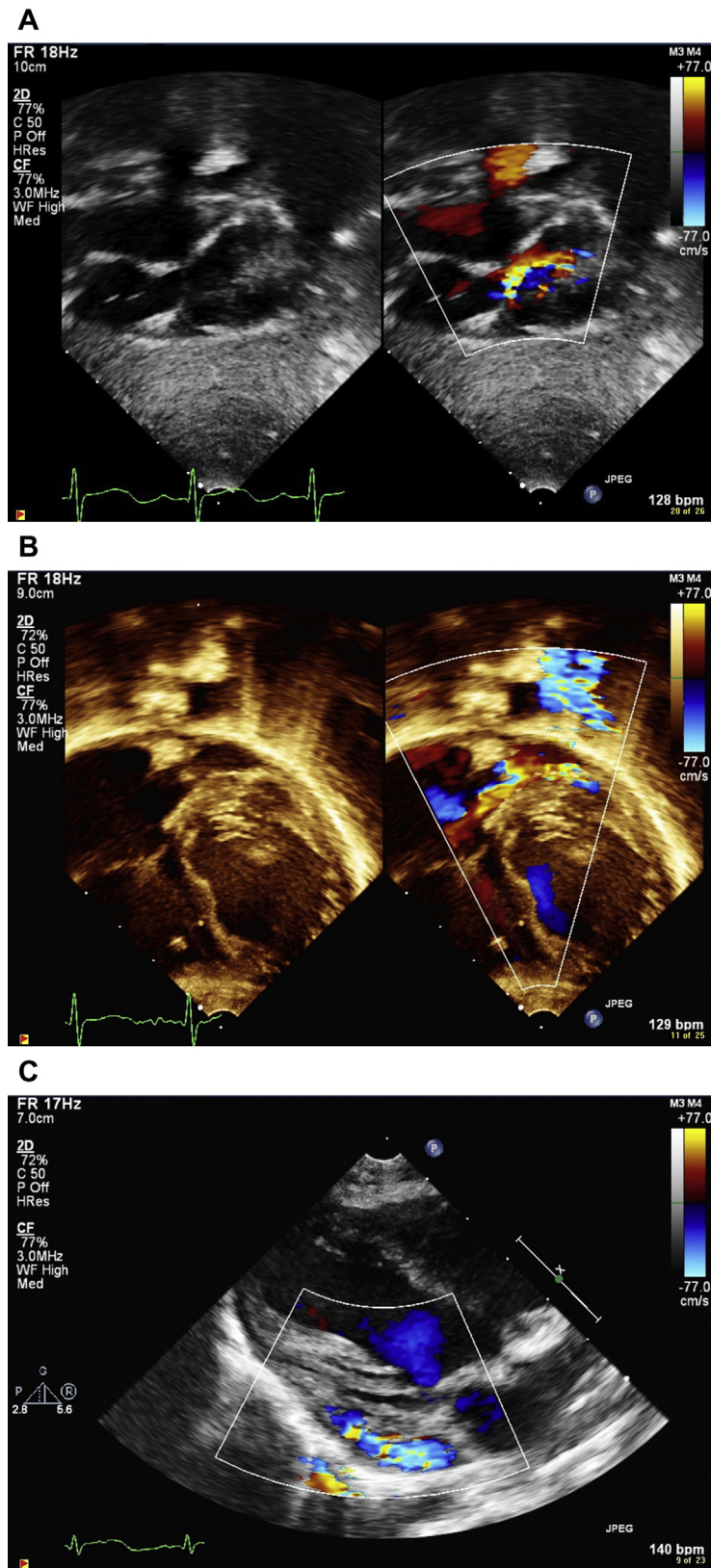
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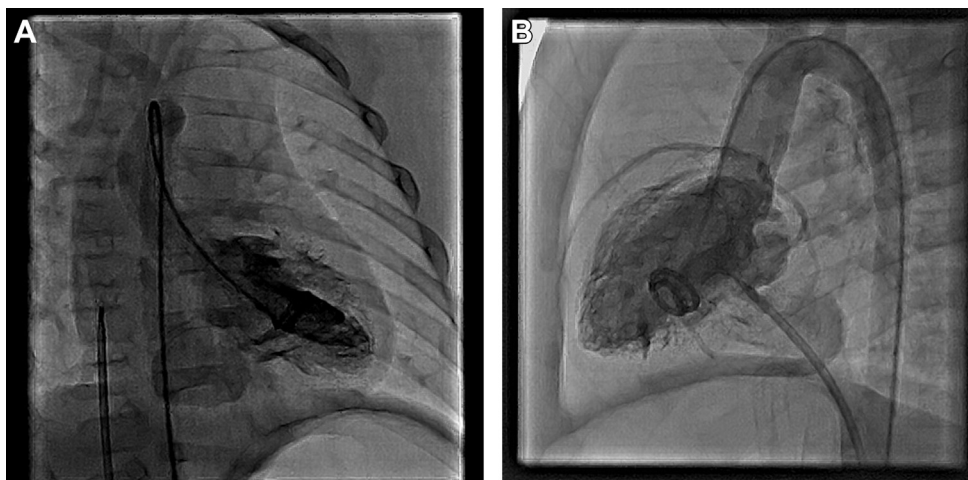
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**Figure 1** Transthoracic echocardiography. **(A)** Subcostal coronal view, two-dimensional and color Doppler showing moderate dilation of the CS with turbulent flow. **(B)** Apical four-chamber view with color Doppler showing the turbulent flow jet coming from the left ventricle to the CS. **(C)** Parasternal long-axis view with color Doppler showing the fistula flow from the posterior-inferior aspect of the left ventricular free wall to the CS. 2D, Two-dimensional; CF, color flow Doppler.



**Figure 2** Cardiac catheterization left ventriculography. **(A)** Anteroposterior projection showing a small amount of contrast entering the right atrium from the CS. **(B)** Lateral projection showing flow from the left ventricle into the CS. Coronary arteries have normal origins and courses.

with associated congenital heart disease. Ultimately, further diagnostic testing, including cardiac catheterization, computed tomography, or magnetic resonance imaging, may be helpful in planning for catheter-based or surgical intervention.

### CONCLUSION

We report a rare isolated congenital lesion in an otherwise structurally normal heart, a restrictive left ventricle-to-CS fistula. Over a 5-year period, our patient with this lesion remains asymptomatic, with normal biventricular size and function. Therefore, we suggest in infants and young children with this isolated abnormality that routine monitoring may be acceptable as opposed to surgical or catheter-based treatment very early on.

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### SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.case.2017.04.007>.

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