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Clinical Presentation and course.

History: A 2 month old male, born by c-section at 39 weeks without complications during pregnancy or birth, was noted to have a systolic murmur at his general pediatrician's office. At time of presentation, he was asymptomatic, without reported cyanosis or shortness of breathing. He was healthy appearing and was growing appropriately.

Physical Exam: He appeared to be a well nourished appearing male. On auscultation he had normal heart sounds S1 and S2 with a grade III/VI systolic regurgitant type murmur, loudest intensity at the apex without radiation to axilla and back. He had normal pulses in the radial and femoral arteries.

A 24-Holter monitor was placed at age 4 months, 11 months of age and 1 ½ years of age, which all demonstrated sinus rhythm without ectopic beats or arrhythmias. Echocardiograms were also performed at 6 month intervals.

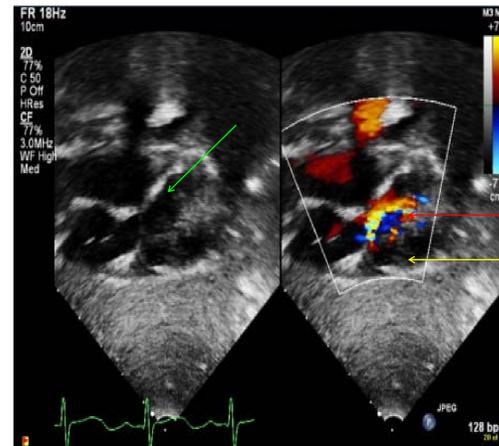


Figure 1. Subcostal view. Yellow arrow showing portion of LV. Red arrow showing vena contracta and green arrow pointing to dilated coronary sinus.

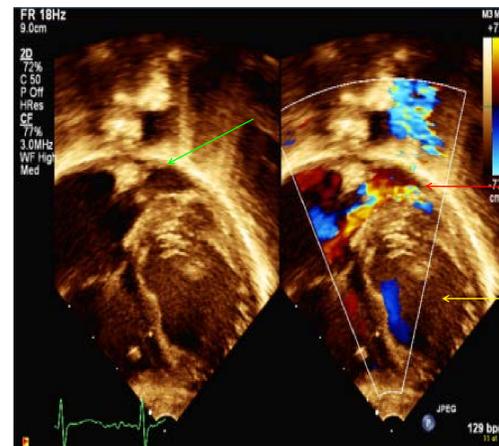


Figure 2. Apical view. Yellow arrow showing normal LV. Red arrow showing vena contracta and green arrow pointing to dilated coronary sinus.

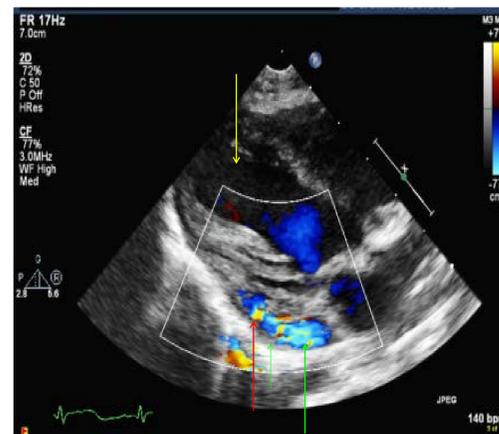


Figure 3. Yellow arrow showing normal LV. Red arrow showing neck of the vena contracta and green arrow pointing to dilated coronary sinus.

Echocardiogram.

A complete 2-Dimensional sector scan, color flow Doppler and Spectral Doppler echocardiogram was performed showing a high-velocity turbulent flow jet into the CS originating from the postero-inferior aspect of the LV. The waist of the jet was 2mm in diameter and its velocity was just over 4m/s, further supporting that it is a small fistula originating from the LV. The flow from the CS, which was dilated, then took an almost 90 degree turn and was seen entering the right atrium in an unobstructed fashion. [Fig. 1-3]. The size of the vena contracta and the estimated pressure gradients have remained stable over the last 2 years.

Cardiac Catheterization.

A cardiac catheterization was performed at the age of 3 months. The study showed a single right superior vena cava entering the right atrium in a normal fashion. The inferior vena cava entered the right atrium in a normal fashion. The pulmonary arteries were normal in appearance. On left ventricular injection, there was flow from the LV into a smooth-walled posterior and inferior chamber in the region of the atrioventricular sulcus which appeared to be the coronary sinus [Fig. 4-5]. There is a small amount of contrast seen to enter the right atrium from the coronary sinus. The pulmonary to systemic flow ratio [Qp:Qs] was 1.1:1. There was a left sided aortic arch with normal brachiocephalic branching pattern. The coronary arterial anatomy was normal without any coronary artery fistula.

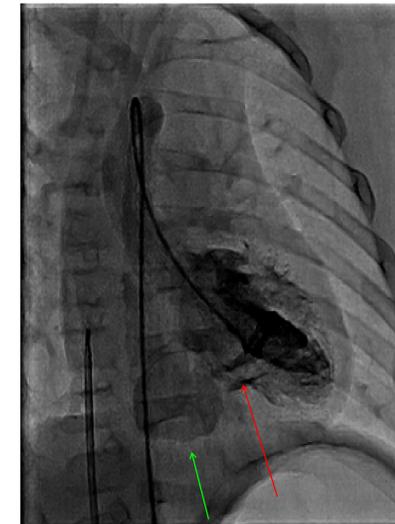


Figure 4. Cardiac catheterization shows the vena contracta (long red arrow) and dilated coronary sinus (green arrow).



Figure 5. Lateral view of cardiac catheterization shows the vena contracta (long red arrow) and dilated coronary sinus (green arrow).

References

- Shah, S.S., et al., *Imaging of the coronary sinus: normal anatomy and congenital abnormalities*. Radiographics, 2012. 32(4): p. 991-1008.
- Gnanapragasam, J.P., A.B. Houston, and S. Lilley, *Congenital fistula between the left ventricle and coronary sinus: elucidation by colour Doppler flow mapping*. British heart journal, 1989. 62(5): p. 406-408.
- Pedra, S.R.F.F., et al., *Left Ventricle to Coronary Sinus Fistula Complicating the Outcome of Transposition of the Great Arteries*. Pediatric Cardiology, 2002. 23(4): p. 466-468.
- McGarry, K.M., J. Stark, and F.J. Macartney, *Congenital fistula between left ventricle and coronary sinus*. British heart journal, 1981. 45(1): p. 101-104.
- Fetter, J.E., et al., *Successful repair of congenital left ventricle-to-coronary sinus fistulas*. The Annals of thoracic surgery, 1994. 57(3): p. 757-758.

Discussion.

An LV to CS fistula has rarely been documented in an otherwise structurally normal heart. In the classification of coronary sinus anomalies [1], the other varieties of anomalies of the CS have been described extensively. According to this classification scheme, this anomaly of the CS would fall into the category of high pressure left to right shunt causing CS enlargement. On reviewing the literature regarding this, only one additional case by Gnanapragasam et al in 1989 described isolated LV to CS fistula without additional structural heart defect in a 20 month old. In this case, initially echo showed a high velocity jet from the LV to an enlarged coronary sinus and subsequently a catheterization was performed to confirm the diagnosis[2].

A limited numbers of reported cases of dilated coronary sinus due to LV fistula have otherwise been associated with additional cardiac abnormalities[3]. McGarry et al. reported coronary artery sinus enlargement secondary to a LV fistula in the setting of transposition of the great arteries with ventricular septal defect and left ventricular outflow obstruction, which required surgical repair along with repair of their fistula [4]. Successful surgical repair of LV to CS fistulas has been reported with good result by patch closure using Gore-Tex[5]. Postoperative repair of the coronary sinus was complicated by supraventricular tachycardia, atrial flutter and fibrillation in 1 case [5] and mortality in one case [4] operated in 1979. The cause of death on post-mortem analysis was poor myocardial protection during ischemic arrest.

Echocardiogram with color flow and Spectral Doppler can establish the diagnosis. If there are associated structural heart defects and surgery is contemplated, additional imaging by CT scan or conventional angiography should be obtained. Percutaneous closure in the cardiac catheterization lab is theoretically possible through the CS approach for a small fistula. Surgical patch closure of this has been successfully described with good short term results and minor post-operative morbidity.

Conclusion.

We present a case of a toddler with structurally normal heart except an isolated LV to coronary sinus fistula. On follow-up echocardiograms the appearance of this fistula has remained stable and on surveillance 24-Holter monitors, he has not had any arrhythmias. He continues to have normal growth and development. We suggest:

- In subjects with this isolated LV to CS fistula, non-invasive monitoring should be considered as opposed to invasive intervention.
- This monitoring should include periodic echocardiograms with EKGs and 24 hour Holter monitor.
- Surgical or catheter based intervention may become necessary if there is evidence of significant left to right shunt, arrhythmias or if there is associated congenital heart disease. Further diagnostic testing, including cardiac catheterization / CT scan / MRI, may be helpful in planning for catheter based or surgical intervention.