Images in Cardiovascular Medicine

Supracristal Ventricular Septal Defect

Lane R. Miller, MD
Margit Nemeth, MD
Scott D. Flamm, MD
Chung Sung, MD
Raymond F. Stainback, MD

Ventricular septal defects (VSD) are the most common congenital cardiac malformation. Epidemiologic data report that the prevalence of this anomaly may be as high as 3.3% to 3.8% of live births.[1,2,3] A VSD can occur in isolation or in association with other cardiac malformations. These congenital malformations are usually detected in childhood, and a substantial number undergo spontaneous closure before adulthood. Anatomic classification distinguishes between ventricular septal defects in accordance with whether they arise above or below the crista supraventricularis. Several classifications of VSDs exist, and there are many synonymous names for the various defects. The major classifications have their basis in anatomic position. Those located below the crista supraventricularis are called subaortic, perimembranous, and muscular defects. Those located above the crista supraventricularis are called supracristal (conus) defects.[4]

In the United States, the supracristal VSD comprises only 2% to 3% of all ventricular septal defects. This defect is much more common in patients of Eastern Asian descent. Many ventricular septal defects close spontaneously during childhood; however, a substantial portion of the larger defects eventually cause significant aortic insufficiency secondary to prolapse of the aortic valve. When this occurs, surgical repair of the defect and suspension of the aortic valve is indicated.[5]

Echocardiography is used to evaluate young patients with heart murmurs. Valvular disease, obstructive septal hypertrophy, and septal defects are easily identified through use of spectral and pulsed-wave Doppler techniques. Due to the complex 3-dimensional anatomy of the ventricular septum, several imaging planes must be used in order to correctly classify the VSD. Determining the origin of the VSD jet in relationship to the aortic valve annulus is helpful in distinguishing among the VSD types. If the aortic valve is thought of as the face of a clock (parasternal short-axis view, aortic valve level), the most commonly seen perimembranous VSD will arise from the 10 to 11 o'clock position. The subaortic VSD is seen at the 11 to 1 o'clock position, and the least commonly seen supracristal defect arises at the 1 to 2 o'clock position.

We present images obtained from a 27-year-old Chinese male who was referred for evaluation of a prominent, high-frequency, pansystolic murmur. He had no shortness of breath, chest discomfort, or physical activity limitations. An echocardiogram was obtained, along with cardiac magnetic resonance imaging. Both techniques (see figures) illustrate the location of this uncommon defect.

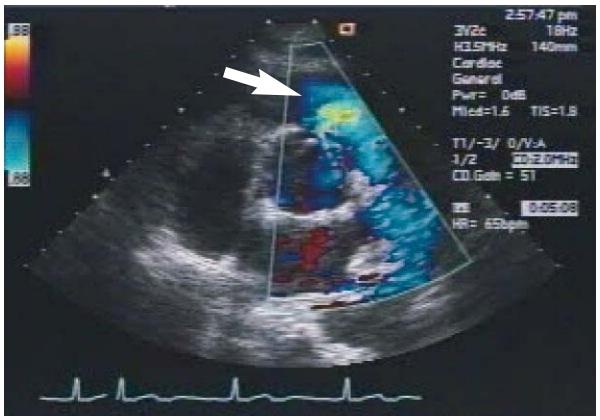

Fig. 1 *Echocardiography (in a parasternal short-axis view with color-flow Doppler) shows an abnormal jet lesion in the right ventricular outflow tract (arrow) near the aortic valve's 1 o'clock position—supracristal VSD. The lesion is small and "restrictive."*

Real-time motion image is available at texasheart.org/education/thijournal/miller331.cfm.

Section Editor:
Raymond F. Stainback, MD, Department of Adult Cardiology, Texas Heart Institute and St. Luke's Episcopal Hospital, 6624 Fannin Street, Suite 2480, Houston, TX 77030

From: *Departments of Cardiology (Drs. Miller, Nemeth, Stainback, and Sung) and Radiology (Dr. Flamm), Texas Heart Institute at St. Luke's Episcopal Hospital, Houston, Texas 77030*

Dr. Miller is now at BCS Heart, College Station, Texas.

Address for reprints:
Raymond F. Stainback, MD, FACC, FASE, Department of Cardiology, St. Luke's Episcopal Hospital, Houston, TX 77030

E-mail: rstainback@sleh.com

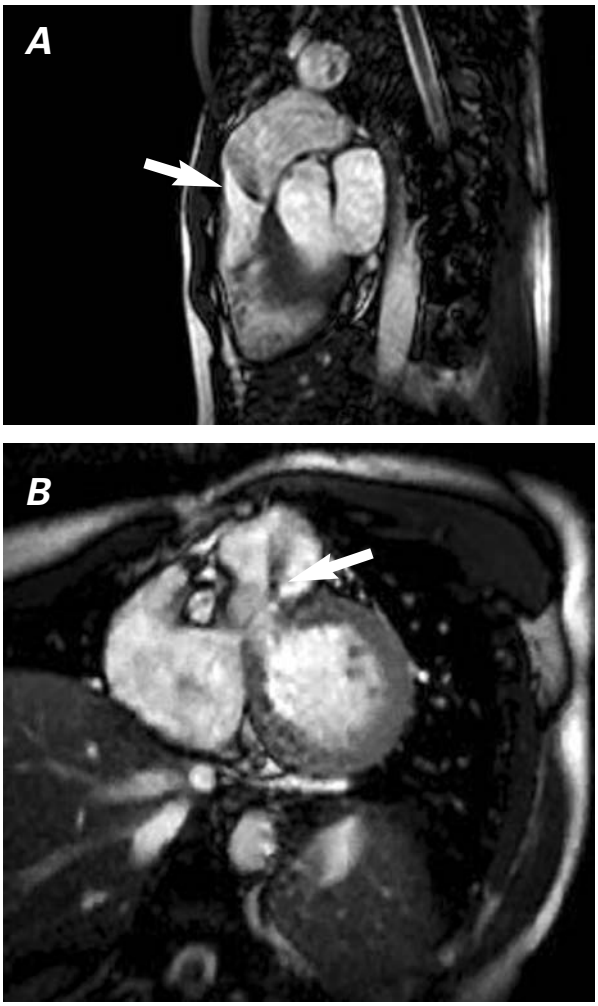

Fig. 2 *Magnetic resonance imaging of the defect in 2 different image planes (**A**, **B**) shows the surrounding anatomic relationships in planes not possible by standard echocardiographic windows and delineates the complex curvilinear anatomy of the basal ventricular septum.*

AoV = aortic valve; arrow = ventricular septal defect jet; RVOT = right ventricular outflow tract

Real-time motion images are available at texasheart.org/education/thijournal/miller331.cfm.

References

1. Weyman AE. 2nd ed. Principles and practice of echocardiography. Philadelphia: Lea & Febiger; 1994. p. 946-52.
2. Ferencz C, Rubin JD, McCarter RJ, Brenner JI, Neill CA, Perry LW, et al. Congenital heart disease: prevalence at livebirth. The Baltimore-Washington Infant Study. Am J Epidemiol 1985;121:31-6.
3. Martin GR, Perry LW, Ferencz C. Increased prevalence of ventricular septal defect: epidemic or improved diagnosis. Pediatrics 1989;83:200-3.
4. Feigenbaum H. Echocardiography. 5th ed. Philadelphia: Lea & Febiger; 1994. p. 384-91.
5. Snider AR, Serwer GA, Ritter S. Echocardiography in pediatric heart disease. Chicago: Year Book Medical Publishers; 1990. p. 261-3.