Aortic Stenosis

Fadi E. Moaikel, BA; Richard B. Towbin, MD; Jeffrey A. Towbin, MD; Jason N. Johnson, MD, MHS; Alexander J. Towbin, MD

Abstract

Pediatric Aortic Stenosis (AS) should be suspected in children with fatigue, dizziness, cyanosis, and chest pain. AS can be associated with other congenital heart conditions. Echocardiogram is the first-line imaging study. Up to 40% of patients require intervention, generally involving repair and/or replacement of the aortic valve.

Keywords: congenital heart defect

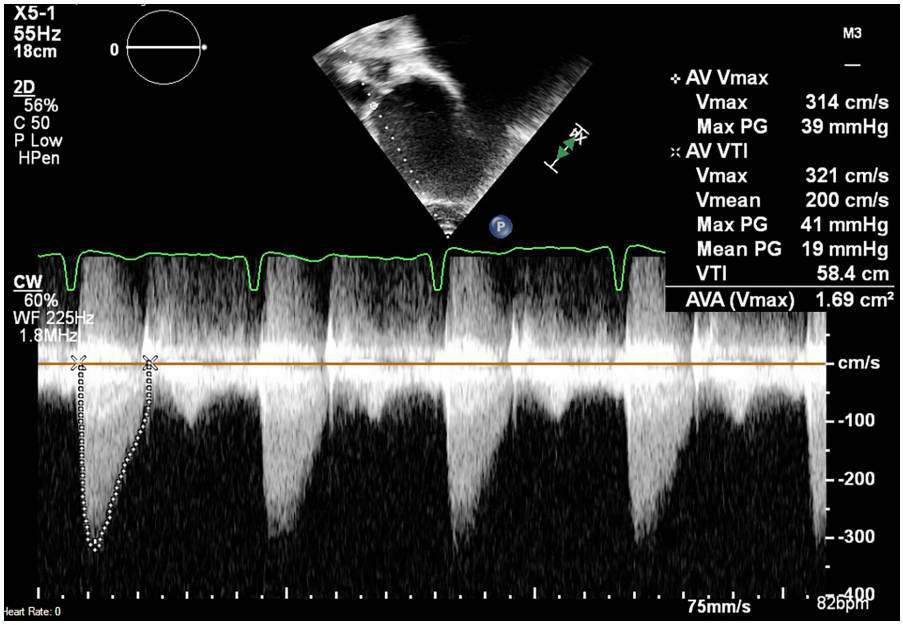
Case Summary

A teenage boy presented with chest pain and shortness of breath during exercise. His physical exam was notable for normal vital signs, but a II/VI systolic ejection murmur was present at the right upper sternal border.

Imaging Findings

Transthoracic echocardiogram (Figure 1) showed a bicuspid aortic valve with stenosis and normal left ventricular systolic function. Cardiac MRI confirmed the presence of a bicuspid aortic valve with moderate aortic stenosis (Figure 2) and moderate insufficiency. There was mild left ventricular chamber dilation and normal systolic function.

Figure 1. Apical 5-chamber transthoracic echocardiogram continuous-wave Doppler across the aortic valve shows a peak velocity of 321 cm/second consistent with moderate aortic valve stenosis.



Diagnosis

Aortic stenosis.

The differential diagnosis is primarily hypertrophic obstructive cardiomyopathy.

Discussion

In pediatric patients, aortic valve stenosis (AS) is a congenital heart defect that creates left ventricular outflow tract obstruction.[1] In neonates, this obstruction leads to an increase in afterload after birth as the low-resistance placental circulation is removed. The loss of the placental circulation, along with the closure of the ductus arteriosus, leads to a drop in systemic cardiac output and the subsequent development of congestive heart failure.[1]

AS is more common in males and accounts for about 3-6% of congenital heart defects. Affected patients present with fatigue, dizziness, cyanosis, and chest pain. Approximately 15-20% of children

Affiliations: Midwestern University Chicago College of Osteopathic Medicine, Downers Grove, Illinois (Moaikel); Department of Radiology, Phoenix Children's Hospital, Phoenix, Arizona (RB Towbin); Department of Cardiology, Le Bonheur Hospital, Memphis, Tennessee (JA Towbin, Johnson); Department of Radiology, Cincinnati Children's Hospital and University of Cincinnati College of Medicine, Cincinnati, Ohio (AJ Towbin).

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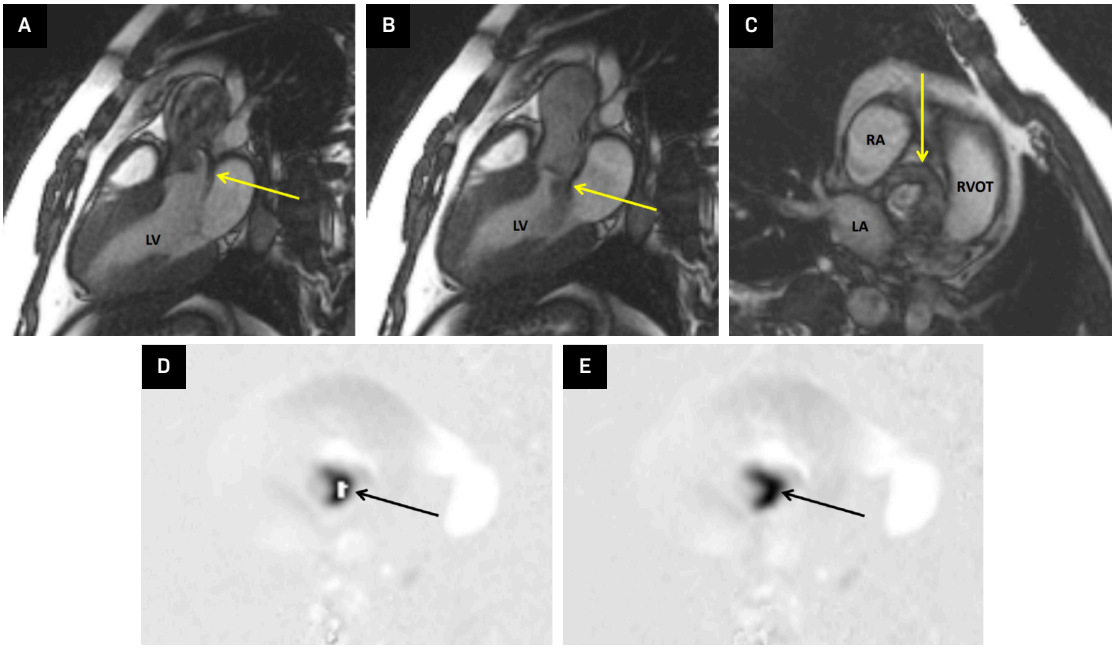
Figure 2. (A) A 3-chamber CMR cine steady-state free precession (SSFP) at peak systole shows systolic doming of the aortic valve with dephasing across the valve consistent with stenosis (yellow arrow). LV, left ventricle. (B) A 3-chamber CMR cine SSFP at end diastole shows aortic valve insufficiency (yellow arrow). LV, left ventricle. (C) Aortic valve en face CMR cine SSFP at peak systole shows a bicuspid aortic valve (yellow arrow). LA, left atrium; RA, right atrium; RVOT, right ventricular outflow tract. (D) Aortic valve en face CMR phase contrast through plane at peak systole shows aliasing across the aortic valve (black arrow). The velocity limit set on the sequence is 300 cm/second, and the aliasing (white pixels in the middle of the black pixels) indicates the velocity across the valve is higher than 300 cm/second. (E) Aortic valve en face CMR phase contrast through plane at peak systole shows resolution of the aliasing across the aortic valve (black arrow). The velocity limit set on the sequence is 400 cm/second, and the resolution of the previously seen aliasing (uniform black pixels) indicates the velocity across the valve is between 300 and 400 cm/second.



with AS have associated congenital heart diseases such as ventricular septal defects and coarctation of the aorta.[2] Several congenital etiologies can cause AS. While supravalvular and subvalvular stenosis can occur in conditions such as Williams syndrome, true valvular stenosis is the most common cause of AS.[3] Of the valvular etiologies, 60% of cases are due to a congenital bicuspid aortic valve. While less common, rheumatic heart disease can cause an acquired AS. When affected in rheumatic heart disease, the aortic valve can become thickened, with shortened cusps and fused commissures.[3]

Transthoracic echocardiography is the preferred imaging modality to assess patients with known or suspected AS. The focus of the echocardiogram is to assess the extent of left ventricular hypertrophy and the ejection fraction during systole.[3] When assessing a bicuspid aortic valve, it is important to identify systolic "doming" of the valve leaflets. When this occurs, the leaflets take a concave shape in respect to the orifice due to restricted excursion.[4] Other important findings include an abnormal number and division of valve commissures and cusp fusion, which may give a false appearance of a tricuspid valve.[4] In assessing the severity of the stenosis, Doppler flow can be used to estimate the degree of stenosis through measurement of the jet velocity and mean gradient. A jet velocity of 2.6-2.9 m/second and mean gradient of <30 mm Hg is indicative of mild AS. Moderate AS is shown with a jet velocity of 3.0-4.0 m/second and a mean gradient of 30-50 mm Hg. Anything greater than this is considered severe AS.[5] While chest radiographs and CT scans can be used to assess the extent of calcification in the aortic valve leaflets, this is a rare finding in the pediatric population and more commonly utilized in adult-onset AS.[3]

Cardiac MRI allows for more detailed dynamic assessment of the left ventricle and aortic valve via improved soft-tissue definition. On cardiac MRI, the radiologist or cardiologist should look for increased left ventricular wall thickness (>12 mm), increased left ventricular mass, and post-stenotic dilatation of the ascending aorta distal to the stenotic valve.[3]

Cardiac catheterization is the gold standard modality used to assess and potentially treat AS in children. During this procedure, the cardiac interventionalist can directly measure pressures across the aortic valve. However, in most instances, cardiac catheterization is only used when there are discrepancies between echocardiogram, when the noninvasive imaging has shown to be inconclusive, or balloon angioplasty is indicated to treat AS.[6]

Currently, there are 2 major interventions indicated for children with AS: balloon aortic valvuloplasty and surgical repair.[7] In a large follow-up study with patients aged 1 month to 23 years, 10-

and 20-year freedom from reintervention was 54% and 27%, respectively. Groups at greatest risk of restenosis are those aged 3 months or younger and those with an immediate post-valvuloplasty mean gradient of >30 mm Hg.[8]

Surgical options include aortic repair, where fibrous excess is removed, or valve replacement for patients with an uncorrectable valve. Those with a bicuspid aortic valve with significant dysfunction and dilation >4.5 cm will likely need a valve replacement.[1] The 10-year freedom from reintervention for patients who underwent a surgical valvuloplasty was about 73%.[9] While the surgical option provides a higher rate of freedom from reintervention, it is associated with greater morbidity and an extended hospital stay compared with balloon valvuloplasty.[1]

Conclusion

Pediatric AS should be suspected in children with fatigue, dizziness, cyanosis, and chest pain. AS can be associated with other congenital heart conditions. Echocardiogram is the first-line imaging study. Up to 40% of patients require intervention, generally involving repair and/or replacement of the aortic valve.

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