Radiology Corner (Case #3)

Bicuspid Aortic Valve

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Introduction

Bicuspid aortic valve is a common congenital heart defect, often diagnosed incidentally or as a consequence of an associated condition. Patients with this anomaly are at increased risk for a variety of cardiovascular complications and require surveillance for associated valvular and aortic diseases. While echocardiogram is typically the initial diagnostic modality, MRI can be a valuable adjunct for more comprehensive examination of both the aorta and aortic valve.

History

38-year-old man with heart murmur on physical exam. Electrocardiogram (ECG)-gated MR of the chest was performed and included axial (Figure 1A) and sagittal (Figure 1B) black blood double inversion recovery fast spin echo (DIR FSE) and axial (Figure 1C) and sagittal (Figure 1D) bright blood fast steady state free precession images. Oblique sagittal cine bright blood fast gradient echo images (Figure 1E, select systolic images) were performed parallel to the long axis of the aortic root. Oblique axial bright blood cine phase contrast images (Figure 1F, select serial systolic images) through the base of the heart through the aortic valve plane were also performed.

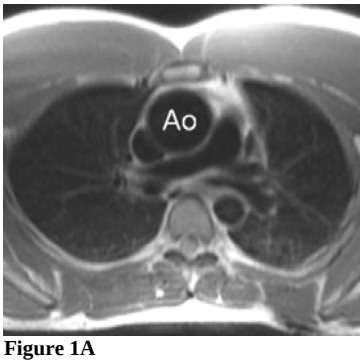

Figure 1A

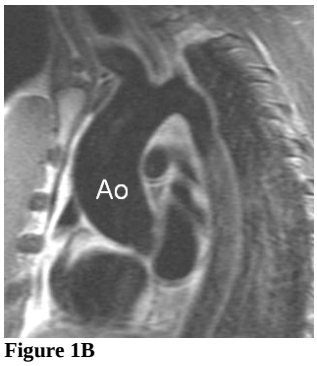

Figure 1B

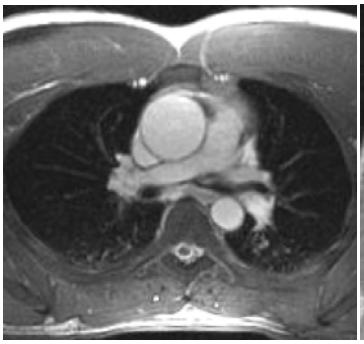

Figure 1C

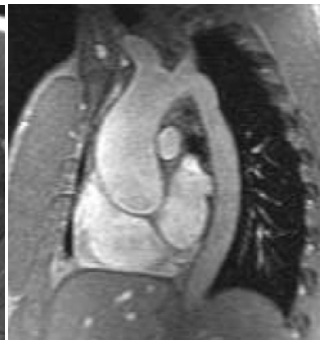

Figure 1D

Figures 1A through 1D. On DIR FSE images (1A, axial; 1B, sagittal), dilation of the ascending aorta to 4.8 cm is well seen. This is confirmed on corresponding bright blood images (1C, axial; 1D, sagittal) performed using a steady state free precession pulse sequence. Ao = ascending aorta

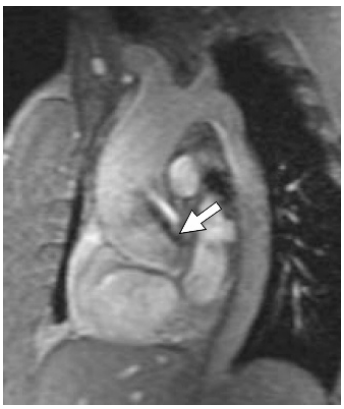

Figure 1E

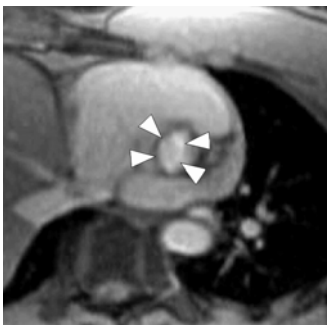

Figure 1F

Figure 1E. Oblique sagittal cine bright blood imaging of the long axis of the aortic root demonstrates a moderate to large flow jet (arrow) on this systolic image consistent with moderate-to-severe aortic stenosis.

Figure 1F. Oblique axial cine bright blood imaging through the valve plane of the aorta, demonstrates the aortic valve to have a lens shaped orifice (arrow heads) consistent with a bicuspid aortic valve.

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Imaging Findings

On axial (Figure 1A) and sagittal (Figure 1B) black blood DIR FSE images, the patient is noted to have a dilated ascending aorta, which measures approximately 48 mm in diameter. Steady state free precession is a relatively newer MR pulse sequence that provides good homogeneity of the bright blood pool signal and confirms the aortic dilatation noted on black blood images (Figures 1C and 1D). On cine bright blood MR using fast gradient echo imaging, a distinctive systolic flow jet (Figure 1E, arrow) is noted which arises from the posterior aortic valve into the ascending aorta. This moderate to large jet is consistent with moderate to severe aortic stenosis. No diastolic jet to suggest aortic insufficiency was seen (not shown). On cine bright blood phase contrast imaging at the level of the aortic valve, the aortic valve orifice is noted to be lens shaped (Figure 1F, arrowheads). Contrast-enhanced three-dimensional MRA (Figure 1G) confirmed the dilated ascending aortic root and absence of an aortic dissection or coarctation.

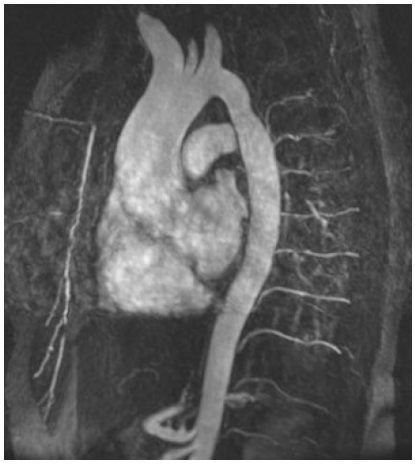

Figure 1G. Contrast-enhanced three-dimensional MRA, performed during the arterial phase of a 0.2 mmol/kg dose of a Gadolinium-chelate contrast bolus injection administered via a right antecubital vein at 2 mL/sec, demonstrates a dilated ascending aorta but no evidence of intimal tear. Note that there is suggestion of an aortic narrowing of the distal arch. However, there was no flow jet seen across the region on cine bright blood imaging (Figures 1D and 1E) or dilated collateral vessels—features that would invariably be present if there was a hemodynamically significant coarctation of the aorta.

Discussion

Bicuspid aortic valve (BAV) is a common congenital heart defect occurring in 1-2% of the population, with a male predominance.[1] Patients with BAV are at increased risk for a variety of complications that includes aortic valvular disease, endocarditis, ascending aortic aneurysm and aortic dissection.[1-4] There is also an association of BAV with coarctation of the aorta.[5,6] While this BAV-coarctation relationship is widely known to exist in patients with Turner syndrome, this is also seen in non-syndromic individuals.

The detection of a bicuspid aortic valve is often incidental, but once found requires routine surveillance for its associated complications. Echocardiography remains the initial study of choice for most patients. However, MRI, capable of functional evaluation using cine bright blood techniques and of aortography using MRA, is well suited for further evaluation of patients with BAV for associated valvular and aortic conditions.

Functional impairment of the aortic valve—namely aortic stenosis and aortic regurgitation—is the most common complication (in up to 68-85% of patients [1,3]) seen in patients with BAV. Aortic stenosis may manifest as a systolic ejection murmur, best heard in the second right intercostal space. Aortic insufficiency, on the other hand, is characterized by a decrescendo diastolic murmur, best heard at the left sternal border. On echocardiography, of course, the severity of the valvular stenosis or insufficiency and co-existence of a BAV is typically evident.

Patients with BAV also have an increased incidence of aortic disease.[7-11] While it is well known that aortic valvular dysfunction is associated with increased risk for aortic aneurysm, patients with BAV appear to have additional risks for aortic disease. Nistri et al.[12] reported significant aortic root enlargement in healthy patients with normally functioning BAV when compared to healthy subjects with normally functioning tricuspid aortic valves. Similarly, Hahn et al.[2] report significantly larger aortic root sizes in patients with BAV at all grades of valvular dysfunction when compared to subjects with tricuspid aortic valves matched for age, gender and grade of valvular disease. These studies suggest that the predisposition for aortic disease in patients with BAV is due to factors other than, or in addition to, traditional hemodynamic considerations.

De Sa et al.[8] and others [7,9,11] have reported severe degenerative changes, such as cystic medial necrosis, elastic fragmentation and alterations of smooth muscle orientation, in the media of the ascending aorta of patients with BAV. Moreover, De Sa et al.[8] found more severe degenerative changes not only in the media of the ascending aorta but also in the pulmonary arteries of patients with BAV, compared to that of patients with tricuspid aortic valves. These studies suggest that the association of BAV with aortic disease results from a common developmental error and not only hemodynamic forces.

In patients with aortic dilatation and BAV, the risk for aortic rupture and aortic dissection are considerably higher.[13] The risk for aortic dissection, in particular, has been reported to be increased 5 to 9 fold in patients with BAV.[10,14] For this reason, some surgeons have advocated more aggressive and earlier elective surgical repair in patients with BAV, aortic valvular dysfunction and aortic dilation.[13]

The clinical presentation of patients with BAV varies and depends on the presence and severity of its associated complications. Often the finding is incidental, such as being found during a routine echocardiogram for evaluation of suspected aortic stenosis or aortic insufficiency. On occasion, it is identified during the evaluation of a dilated aorta as in this case. Alegret et al.[15] reported BAV in 20 percent of their patients with aortic root dilatation. Echocardiography will continue to be the initial study of choice in most instances. However, with the increased use of MRI in the evaluation of

aortic pathology, the evaluation of the aortic valve should be considered during an MRI study especially in instances of aortic dilatation or in cases where there is suspicion or known aortic valvular dysfunction.

On MRI, aortic caliber is best evaluated using ECG-gated black blood pulse sequences (Figures 1A and 1B). Note that these images are typically performed during diastole and measurements may not necessarily correlate exactly with echocardiographic measurements. The aortic valve, on the other hand, is best seen using cine bright blood acquisitions, notably steady state free precession (SSFP; also known as TrueFISP, FIESTA and balanced FFE). To evaluate valvular function, cine images should be performed perpendicular to the left ventricular outflow track (Figure 1E). To evaluate the number of valve leaflets, imaging using cine SSFP (or cine phase contrast, Figure 1F) images should be performed parallel to the aortic valve plane. The determination of BAV is made by the configuration of blood flow across the actual aortic valve. In patients with BAV, the aortic valve orifice is shaped like a lens (Figure 1F); in patients with a tricuspid aortic valve, like a triangle.

MRI is well suited for the determination of aortic caliber and valvular pathology, but can also reliably detect aortic dissection.[16] This can be performed using a combination of black and bright blood pulse sequences. However, contrast-enhanced three-dimensional MRA (Figure 1G) can particularly improve the diagnostic confidence for the diagnosis or exclusion of an intimal tear and or aortic coarctation.[17]

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