How Not to Miss a Bicuspid Aortic Valve in the Echocardiography Laboratory

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Diagnosis of bicuspid aortic valve (BAV) in the busy echocardiography laboratory is important, as it is a relatively common cardiac congenital defect (up to 3% of the population),[1] and requires a recommendation for

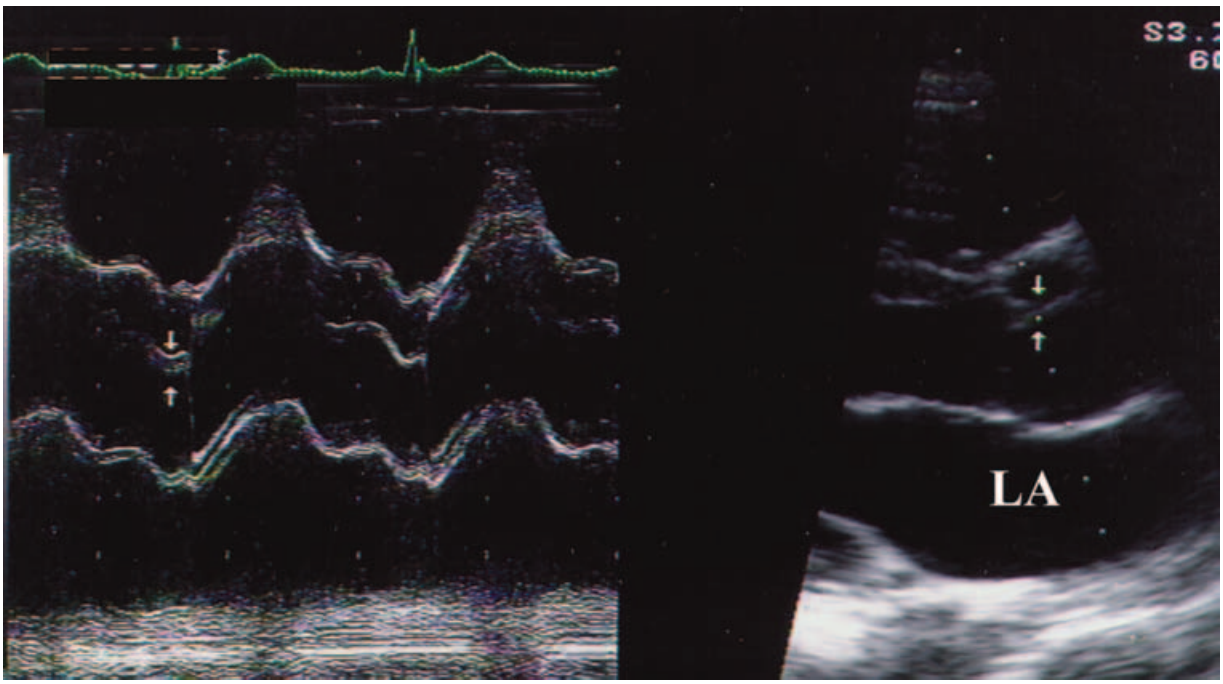

Figure 1. *M-mode and two-dimensional (2D) diastolic frame in the parasternal long-axis view of a bicuspid aortic valve with eccentric closure (arrows). Although not diagnostic, an eccentric line of closure should prompt one to evaluate the aortic valve closely for BAV. LA = left atrium. (Modified with permission from: Kerut EK, McIlwain, Plotnick: Handbook of Echo-Doppler Interpretation 2nd Ed, Blackwell Publishing, Elmsford, New York, 2004, p. 84)*

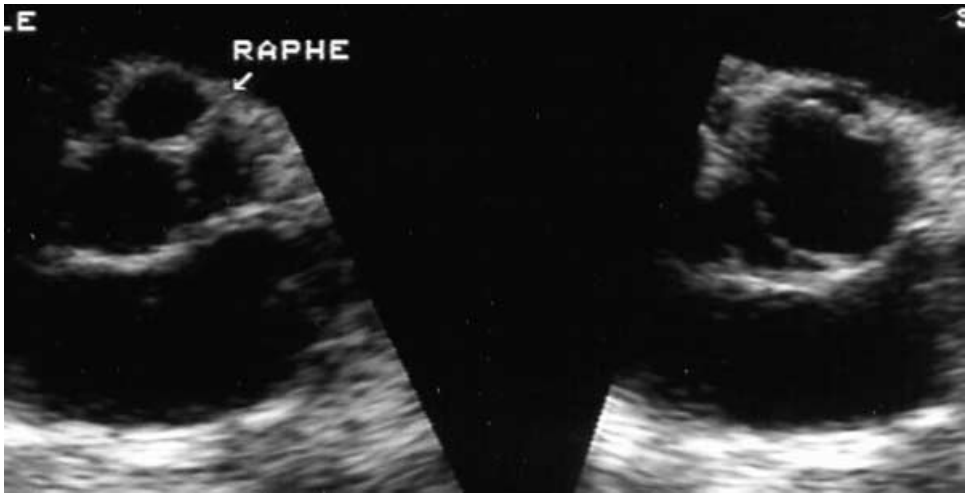

Figure 2. *Diastolic (left panel) and systolic (right panel) two-dimensional (2D) short-axis image of a BAV. In diastole the raphe may appear as a commissure (arrow), appearing to be a normal tri-leaflet valve. However, the systolic frame has a typical "fish-mouth" appearance. (With permission from: Kerut EK, McIlwain, Plotnick: Handbook of Echo-Doppler Interpretation 2nd Ed, Blackwell Publishing, Elmsford, New York, 2004, p. 84)*

endocarditis prophylaxis.[2] BAV usually occurs in isolation but is associated with other anomalies in 20% of cases. The most common associated anomaly is coarctation of the aorta, but occasionally patent ductus arteriosus (PDA), ascending aortic aneurysm, and DeBakey Type I aortic dissection is found.[3–5] One-half of patients younger than 75 years old, with symptomatic aortic stenosis, have BAV.[6] Valvular stenosis appears to progress most rapidly when the leaflets are unequal in size and have an anteroposterior leaflet configuration. Severe aortic regurgitation (AR) is found in 1.5%–3% of patients with BAV.[7] As there exists familial clustering of BAV (male: female ratio of 3:1), echocardiographic screening of first-degree family members is prudent.[8,9]

A BAV may have a right and left cusp (the origin of the right coronary artery is in the right cusp, and left coronary artery in the left cusp), or an anterior and posterior cusp (both right and left coronary arteries originate in the anterior cusp). M-mode of BAV may demonstrate an eccentric line of closure (Fig. 1) but an eccentric line of closure may also be seen in patients with a tricuspid leaflet aortic valve, and a "normal" line of closure may be seen in patients with BAV. From the parasternal short axis, aortic valve anatomy may appear normal in diastole, as a raphe may simulate three commissural lines (Fig. 2). However, in systole the valve will have a "fish-mouth" appearance. In the parasternal long axis, the valve may appear domed in systole (Fig. 3) and prolapse in diastole. With BAV, the ascending aorta may have a normal appearance; however it should be

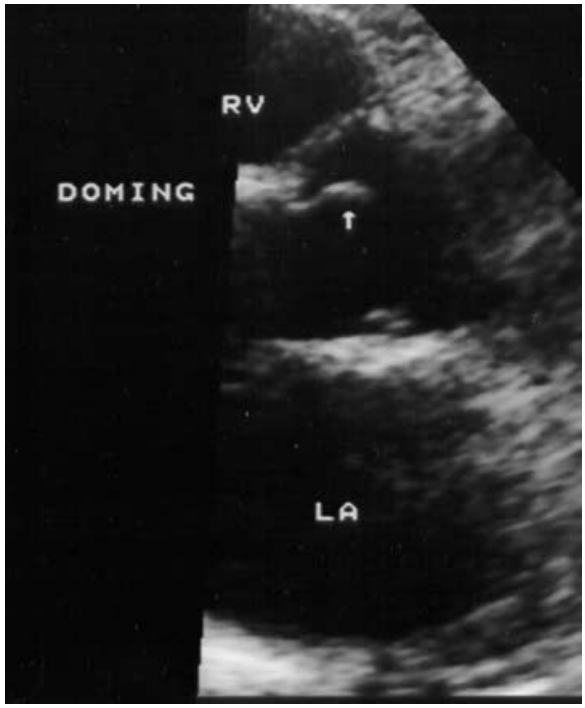

Figure 3. *Systolic parasternal long-axis view of a patient with a BAV. Doming (arrow) is noted. Normally the leaflet should be parallel to the aortic wall when fully open during systole. LA = left atrium; RV = right ventricle.*

inspected for aortic root size and structure, as ascending aortic aneurysm and dissection may be associated.

Some "pointers" to look for a BAV include:

(i) From a parasternal long-axis window, look for systolic valve doming (normally the leaflets are parallel to the aorta), and diastolic aortic valve prolapse.
(ii) As aortic regurgitation (AR) is not "normal," any AR noted by color Doppler warrants a closer look at the aortic valve, to make sure it is not bicuspid.
(iii) As a matter of routine, one should always image the descending aorta from the suprasternal window, particularly with a BAV. Si, aortic coarctation, or PDA should prompt the sonographer to closely evaluate the aortic valve.
(iv) As a confirmatory step in our laboratory, we have a "checkbox" on our echo worksheet, for the sonographer to identify the aortic valve as either tricuspid, bicuspid, or "cannot tell" (difficult image). This is done while imaging in systole and diastole from a parasternal short-axis view.
(v) With a tricuspid leaflet aortic valve, one should note the valve commissures extending to the base of the valve. If this cannot be clearly visualized, consider BAV.

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