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Osman Ratib, MD, PhD; Joseph K. Perloff, MD; John S. Child, MD

Congenitally abnormal aortic valves are defined morphologically according to the number and type of cusps and commissures. The bicuspid aortic valve, first recognized by Leonardo da Vinci in the early 16th century, is the most common congenital anomaly to which that structure is subject. The estimated incidence is 0.9% to 2% of the general population, or a prevalence in the United States of ≈ 4 million. Two varieties of bicuspid aortic valves are based on commissural fusion, namely, fusion between the left and right cusps or fusion between the right and noncoronary cusps. Three varieties of bicuspid aortic valves are based on cuspal size, namely, 2 cusps of equal size, 2 cusps of unequal size, or a conjoined cusp that is twice the size of its mate. The greatest degree of cuspal inequality is represented by the conjoined cusp. The images shown here represent a unique variety of cuspal inequality caused by aneurysmal dilatation of the nonconjoined cusp (Figure).

Dilatation of the ascending aorta, which is a common coexisting feature of a bicuspid aortic valve, is caused by an inherent medial abnormality that attenuates the ascending aortic wall, and is coupled with the bicuspid morphology of the congenitally malformed valve rather than its functional state. The aneurysmal cusp shown in the Figure might harbor a tissue abnormality analogous to the inherent medial fault that attenuates the dilated ascending aorta.

The 2 coronal images on the left are from dynamic cine MRIs. Left, Upper image in diastole shows aneurysmal dilatation (Aneur) of 1 cusp of a congenitally bicuspid aortic valve. There is no regurgitation. The ascending aorta (AscAo) is moderately dilated. Lower image in systole shows unobstructed flow across the left ventricular outflow tract (LVOT), LV indicates left ventricular. Right, These 2 oblique images were acquired across the bicuspid aortic valve. Upper image in diastole shows the aneurysmal nonconjoined aortic cusp (Aneur) and the smaller conjoined second cusp with its false raphe (FR). Lower image in systole shows the unobstructed bicuspid valve (BicuspV), which was functionally normal despite the cuspal aneurysm.