Quadricuspid aortic valve: a rare cause of aortic regurgitation

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Quadricuspid aortic valve (QAV), a rare congenital anomaly, is an uncommon cause of aortic regurgitation. Echocardiography is the first-line imaging modality in valvular heart disease but cardiovascular magnetic resonance (CMR) is becoming an important tool. We present two cases of aortic regurgitation in patients with quadricuspid aortic valve seen on CMR.

Hurwitz and Roberts\(^1\) described seven morphological types of quadricuspid valve based on the anatomic appearance of the valve.\(^2\) The first case demonstrates a type A QAV where there are four aortic sinuses and four equally sized cusps. There is incomplete leaflet closure in diastole resulting in AR (Figure 1A).

The leaflets fail to close down normally into the aortic annular plane resulting in a central regurgitant orifice arising above the annular plane at the mid-sinus level. In systole the slightly undersized leaflets fail to fold back normally into each sinus resulting in a “square” systolic orifice (Figure 1B).

The second case illustrates an even more unusual variant of QAV. In this case there are four sinuses and four leaflets but each pair of leaflets is fused to produce a functionally bicuspid valve with the typical “fish-mouth” appearance of a BAV (Figures 2A & 2B).

Figure 1A. Short axis view of QAV in diastole, with a small central coaptation defect (arrow)
Figure 1B. Short axis view of QAV in systole demonstrating a square systolic orifice (arrow)

Figure 2A. Short axis view of QAV in diastole, with a small central coaptation defect (arrow)
Figure 2B. Short axis view of QAV during systole demonstrating “fish-mouth” appearance (arrow)

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