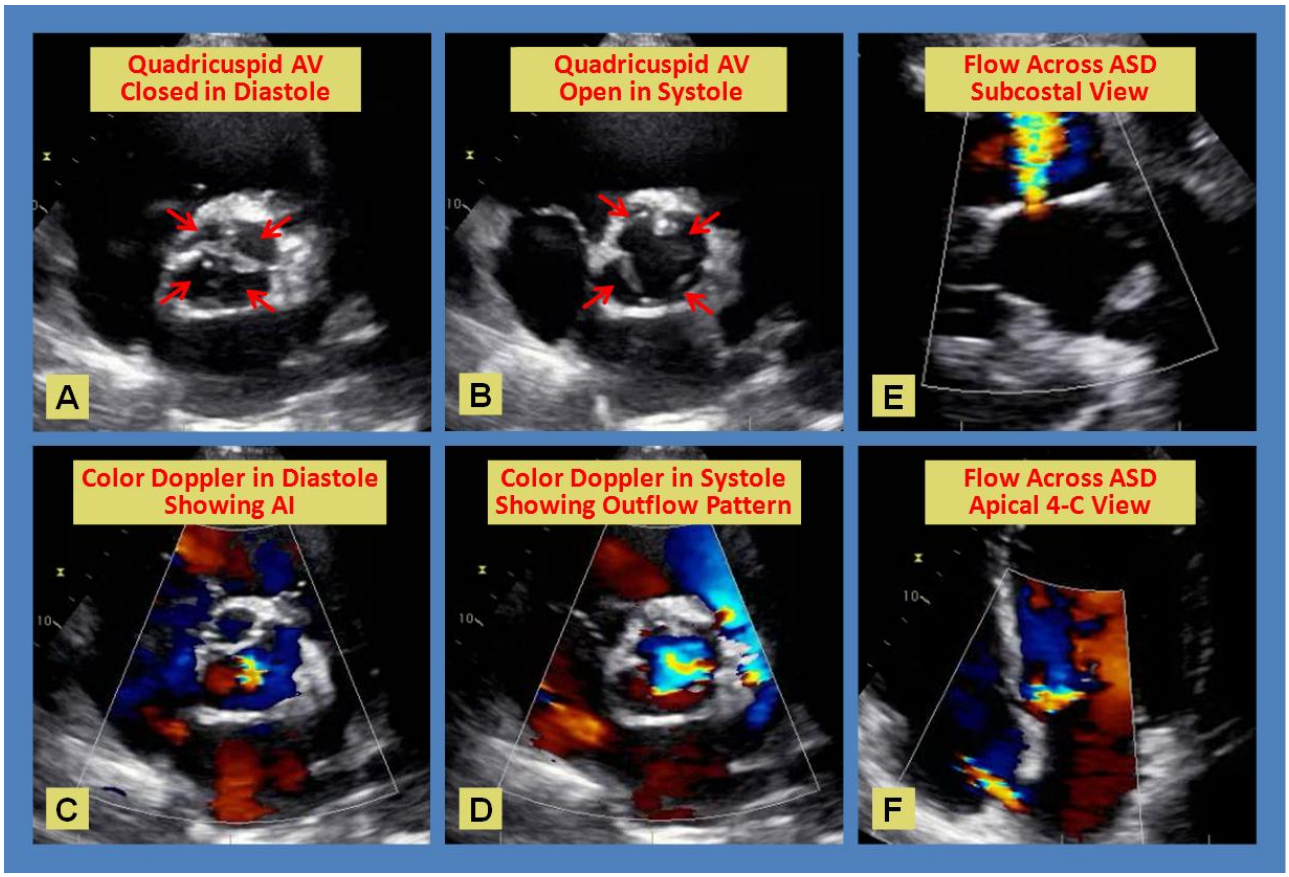
Quadricuspid Aortic Valve..*Associated ASD!*

Sagar Kumar, M.D.[a], Mahmoud N. Hamad[b], Sarina Sachdev, M.D.[a], Christopher Malozzi, D.O.[a], Bassam Omar, M.D., Ph.D.[a,c]



Description

The figure above shows a 2-dimensional parasternal short axis view of a quadricuspid aortic valve in closed (A) and open (B) position with arrows pointing to the individual cusps. Note the sclerosis affecting the coapt points. Color flow Doppler across the valve in diastole (C) shows a central jet of mild aortic regurgitation, and the outflow pattern across the valve in systole (D). The subcostal view (E) and the apical 4-chamber view (F) both demonstrate spontaneous left to right shunting across the interatrial septum consistent with an ostium secundum atrial septal defect.

While bicuspid aortic valve is the most common anomaly of the aortic valve, with a global incidence of 1.3%, followed by unicuspid aortic valve, quadricuspid aortic valve is a rare finding, with a reported incidence of less than 0.05%, which is least among the abnormal variants of aortic valve [1]. The first case of quadricuspid aortic valve was reported in 1862 by Belington [2].

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a Division of Cardiology, University of South Alabama, Mobile, AL, USA

b Royal College of Surgeons, Dublin, Ireland

c Corresponding Author: Bassam Omar, MD, PhD. Division of Cardiology, University of South Alabama, 2451 USA Medical Center Dr., Mobile, AL 36617, USA. Email: bomar@health.southalabama.edu

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Hurwitz and Roberts [3] classified quadricuspid aortic valves into seven types based on variations in the size of the cusps. Type B, which is composed of three cusps of same size and one smaller cusp, as shown in the figure above, is the most prevalent type (41%) followed by type A (32%) in which all four cusps are of same size. Prevalence of aortic regurgitation is 77% in type A and 60% in type B.

Although quadricuspid aortic valve is often an isolated anomaly, coexistent cardiac malformations have been reported, especially coronary anomalies [4]. Knowledge of the coronary anatomy is very important to avoid damage to anomalous coronary ostia during valve surgery [5, 6]. Sudden cardiac death has been reported in young patient with quadricuspid aortic valve with complete isolation of the left coronary artery by an adherent aortic cusp leading to myocardial infarction [7].

Other reported cardiac abnormalities associated with quadricuspid aortic valve include VSD [8], PDA [9], Pulmonary stenosis [10], complete Heart Block [11], dilated and noncompaction cardiomyopathies [12, 13], and hypertrophic cardiomyopathy [14]. Several case reports have also been published establishing an association of quadricuspid aortic valve with a patent foramen ovale and an atrial septal defect, as in the figure above [15 – 19]. Cases of infective endocarditis [20], ischemic stroke [21], aortic dissection [22] and left ventricular hemangioma [23] have also been reported in patients with quadricuspid aortic valve.

The mean age of presentation in patients with a quadricuspid valve is 50.7 years, with a slight male predominance with a male to female ratio of 1.6 [24]. Most common associated functional abnormality is aortic regurgitation (75% of the cases), followed by combined aortic regurgitation and aortic stenosis (8% of the cases), while isolated aortic stenosis is rare (0.7% of the cases). Approximately 16% of the cases of quadricuspid valve are normally functioning with no associated functional abnormalities.

Although echocardiography is the gold standard for diagnosing quadricuspid aortic valve, occasional cases may be missed [25]. Advanced imaging has been shown to help better visualize quadricuspid valves including 3-dimensional (3-D) transthoracic echocardiography [26], transesophageal echocardiography (TEE) [27], intraoperative TEE [28] and 3-D TEE [29]. Visualization using cardiac magnetic resonance imaging [30] and cardiac computed tomography [31] can be diagnostic or supplementary to echocardiography. Multimodality imaging is often required to better visualize a quadricuspid valve and characterize associated anomalies [32].

Quadricuspid aortic valve is a rare congenital heart defect with a high potential for serious complications. Patients with this condition should be carefully evaluated for associated anomalies and require close follow up [1]. Treatment of the quadricuspid aortic valve is dependent on associated anomalies and severity of the valve dysfunction (often aortic regurgitation). When there is need for surgical intervention, treatment may entail aortic valve replacement [33], aortic valve repair [34], and more recently, transcatheter aortic valve replacement (TAVR) [35]. Close post-operative long term follow-up is required given the ongoing risk of aortic root and ascending aorta dilatation [36].

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