

Introduction

Quadricuspid aortic valve (QAV) is a rare congenital valvular anomaly characterized by four aortic cusps instead of the normal three. It results from abnormal cusp formation during valve morphogenesis. Although often discovered incidentally, QAV is clinically important because it commonly leads to aortic regurgitation (AR) and may be associated with ascending aortic dilation.

Classification

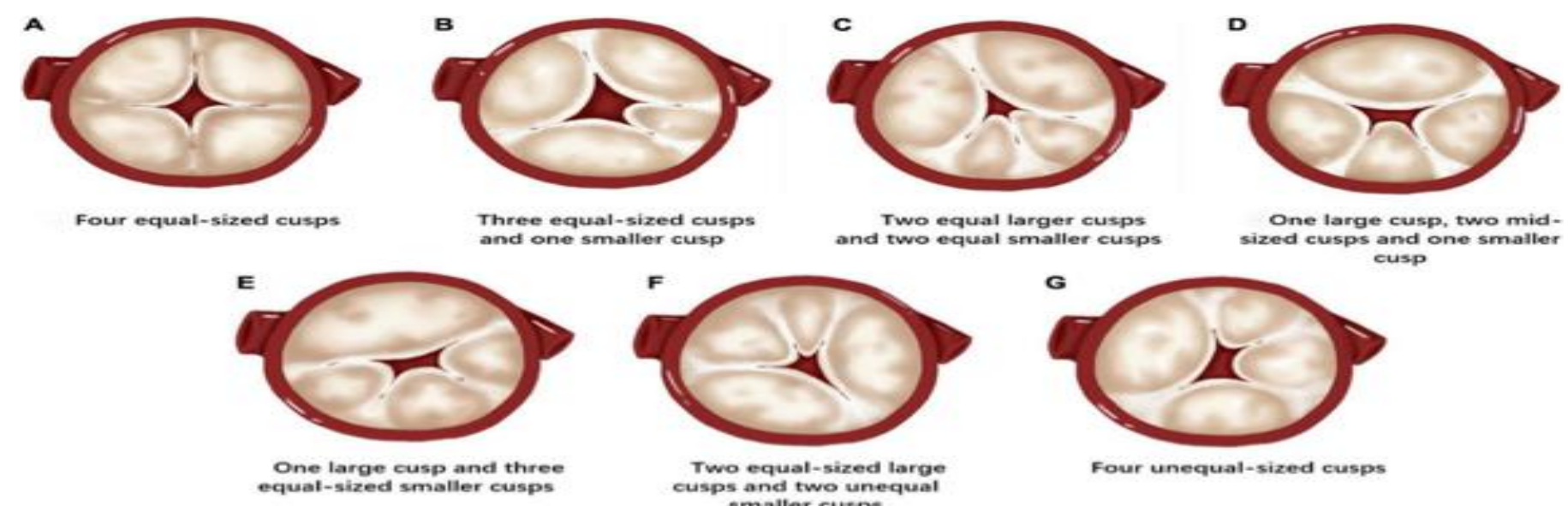
Embryology:

- QAV results from abnormal cusp formation during valvulogenesis.

Classifications:

There are two utilized QAV anatomical classification systems. The first one is Hurwitz and Roberts classification system, seven subtypes were included, lettered A to G, based on the four cusps’ relative sizes, with an additional type H supplemented by Vali et al. Based on their findings, approximately 85% of QAVs are of type A, B, or C.

Type	Description	Prevalence
A	Four equal cusps	32%
B	Three equal larger cusps + one small cusp	41%
C	Two large equal + two small equal cusps	15%
D	One large, two intermediate, one small	3%
E	One large + three equal small cusps	2%
F	Two equal large + two unequal small	2%
G	Four unequal cusps	2%
H	One large, one intermediate, two equal small	Very rare



Clinical relevance:

Morphology influences coaptation quality and therefore the severity of AR; asymmetric configurations are more prone to central regurgitant jets.

Case Description

A 61-year-old female presented with progressive chest discomfort, shortness of breath, and rest pain. Non-contrast chest CT, ordered to rule out an abdominal aortic aneurysm, revealed a QAV. Transthoracic echocardiography (TTE) with 2D, Doppler, and M-mode confirmed QAV with four mildly thickened cusps, mild aortic regurgitation (AR) (AVA 1.80–2.00 cm², peak gradient 17.8 mmHg, LVIDd 4.1cm, PHT not well quantified), and preserved LVEF (64%). Short-axis views showed an “X-shaped” systolic pattern and rectangular diastolic appearance. Aortic root diameter was 33.5 mm (sinus-to-sinus) and the ascending aorta was dilated (41.5 x 40.7 mm). The descending thoracic aorta measured 20.4 x 21.6 mm and the main pulmonary artery was 22.7 mm in diameter. Transesophageal echocardiography (TEE) is planned to assess cusp morphology and the degree of aortic insufficiency.

Diagnosis

Modern clinical practice diagnostics continue to be basis of detection and assessment of QAV. The primary modes of assessment include:

- Two-dimensional transthoracic echocardiography (with spectral doppler assessment)
- Real-time three-dimensional transesophageal echocardiography (with spectral doppler assessment)
- Computed tomography angiogram
- Cardiac magnetic resonance imaging
- Surgical examination

TEE is noted to be more sensitive compared to TTE in defining valve morphology and function

Echocardiographic Criteria to Grade Aortic Regurgitation

		Mild	Moderate	Severe
			Mild to moderate	Moderate to severe
PHT	ms	>500	500-200	< 200
Vena contracta	mm	< 3	3 – 6	> 6
Jet / LVOT	%	< 25	25-45	46-64
Regurgitant Volume	ml	< 30	30-44	45-59
Regurgitant Fraction	%	< 30	30-39	40-49
EROA	cm ²	< 0.10	0.10-0.19	0.20-0.29
Aortic Backflow	cm	Early diastolic		Holodiastolic > 15

Images

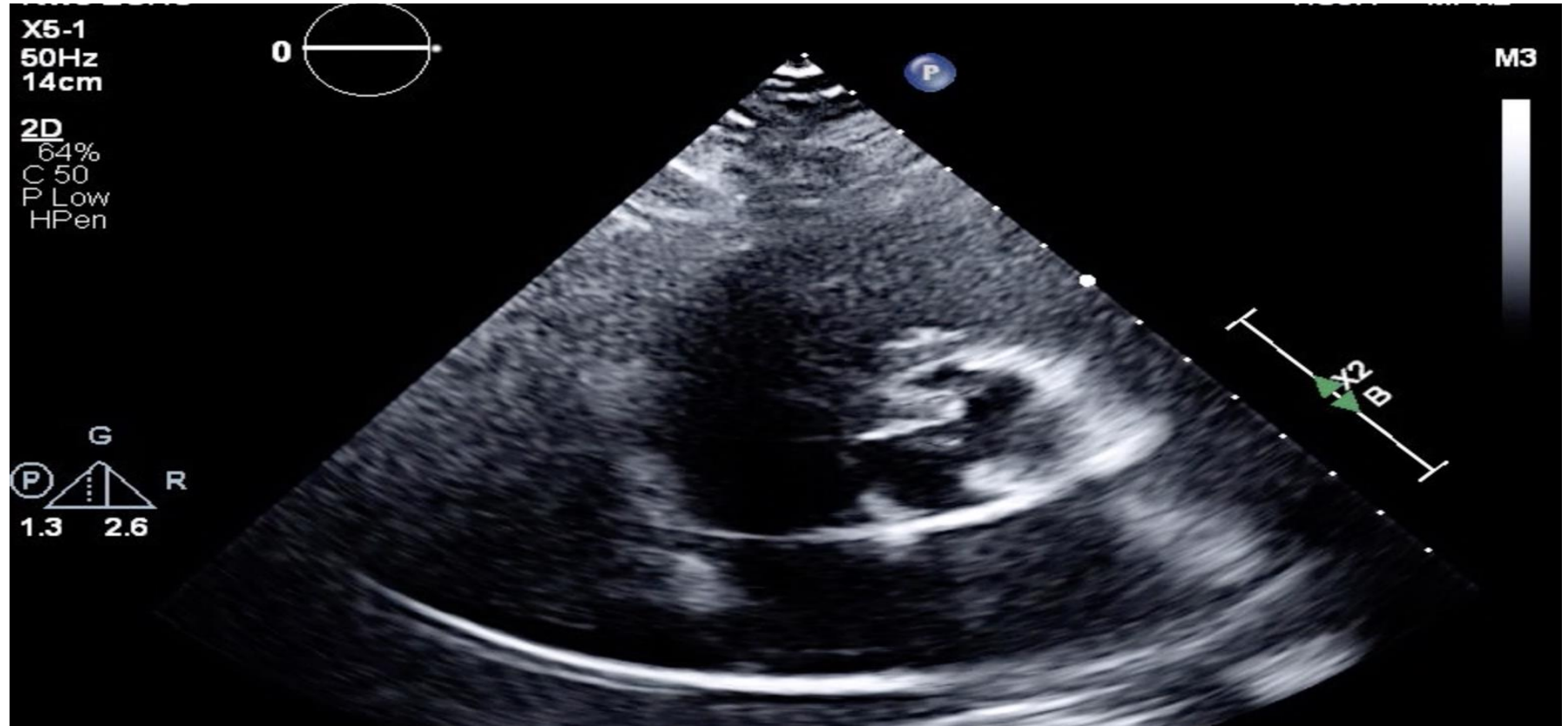


Figure A. 2D TTE of aortic valve in short axis



Figure B. CTA heart and coronary arteries with contrast.
#1: Normal coronary artery origins. Left dominant coronary circulation. CAD RADS 0 disease. No coronary stenosis or plaque noted.
#2: Coronary calcium score: 0 AU.
#3: Aortic Valve: Suspect quadricuspid AV (small accessory 4th cusp noted between the NCC and RCC) Only diastolic images were acquired on this CT scan.

Management

Management depends on symptoms, degree of regurgitation, and associated aortopathy.

Conservative Management: Appropriate for asymptomatic or mild AR Routine echocardiographic surveillance Blood pressure optimization	Surgical Intervention: Indicated for: Symptomatic AR Severe AR with LV dilation or reduced EF Concomitant ascending aortic dilation Options include aortic valve repair, valve replacement, or aorta replacement when necessary.	Transcatheter Approach: High-risk surgical candidates may be considered for: TAVR/TAVI, though experience is limited due to rarity
--	---	--

References

- Mathew JP, Swaminathan M, Ayoub CM. Clinical Manual and Review of Transesophageal Echocardiography. 3rd ed. McGraw Hill; 2019.
- Tsang MYC, et al. Quadricuspid aortic valve: characteristics and clinical outcomes. Circ Cardiovasc Imaging. 2016;9(5):e003483.
- Jagannath AD, et al. Quadricuspid aortic valve: a review of 12 cases. Echocardiography. 2011;28(9):1035–1040.
- JACC Case Rep. 2025;30:105375
- Saith S, Saith S, Murthy A. Quadricuspid Aortic Valve: An Introduction for Clinicians. Cardiol Res. 2022 Feb;13(1): 2-10. doi: 10.14740/cr1308