Valvular Mischief: A Case of a Quadricuspid Aortic Valve Mistaken for a Bicuspid Aortic Valve

Shelley Fontal[1], Asish Katta[1], Teresa Samson[2], Steven Kindsvater[3]

[1]University of the Incarnate Word School of Osteopathic Medicine

[2]Texas Institute for Graduate Medical Education and Research

[3]Baylor Scott & White Heart Hospital

Background

A quadricuspid aortic valve (QAV) is a rare phenomenon characterized by four aortic valvular leaflets, or cusps, instead of three common leaflets.

The most common QAV variation is three equal-sized coronary and noncoronary cusps with one smaller supernumerary cusp. This subtype also correlates with higher rates of aortic regurgitation due to the presence of one smaller cusp.

The incidence of QAVs lies between 0.0059% and 0.0065% in patients undergoing TTE examination, while BAVs occur at a rate between 0.5% and 2%.

Imaging with a transthoracic echocardiogram (TTE) poses diagnostic challenges as it has lower sensitivity in comparison to a transesophageal echocardiogram (TEE). Consequently, QAVs are often missed or misdiagnosed initially as BAVs.

Presentation

A 54-year-old male initially presented for cardiac evaluation for exertional chest pain.

Coronary CT angiography with CT fractional flow reserve revealed a dilated left ventricle with a reduced LV ejection fraction of 27% and a 4.4 cm ascending aortic dilation.

TTE was suspicious for bicuspid aortic valve:

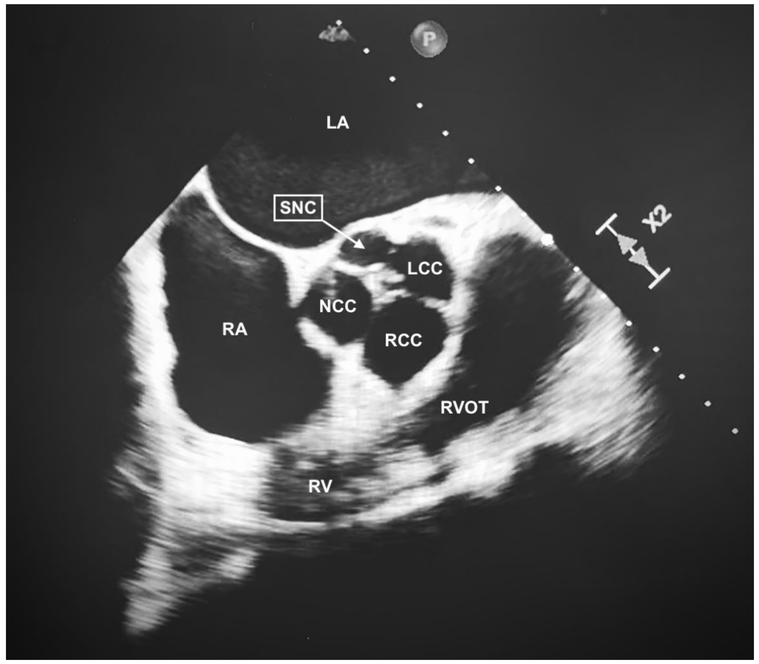
- Moderate aortic insufficiency with PHT of 280 msec
- 2 sclerotic aortic cusps

TEE revealed quadricuspid aortic valve:

- Severe aortic insufficiency with PHT of 200 msec
- 4 equal-sized aortic cusps

The patient underwent a supported Ross procedure to reconstruct the aortic valve and correct the quadricuspid aortic valve.

Figures



LA = left atrium, RA = right atrium, RV = right ventricle, RVOT = right ventricular outflow tract, NCC = non-coronary cusp, RCC = right coronary cusp, LCC = left coronary cusp, SNC = supernumerary cusp

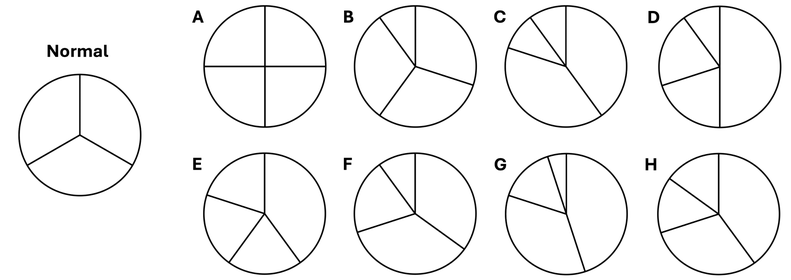
Discussion

While a TTE is the preferred imaging modality for detecting aortic valvular defects, there are many drawbacks with its utility in identifying QAVs:

- External acoustic windows, lower resolution
- Inadequacy of parasternal long- or short-axis views if the SNC is smaller or overlaps
- Interference from other pathology (calcification, aortic root dilatation, etc.)

Utilization of a TEE overcomes many of these obstacles:

- Internal acoustic window overcomes body habitus
- Deep transgastric and trangastric long-axis views are most accurate for viewing the LVOT even in the presence of other pathology



Classification according to Hurwitz & Roberts, 1973

A	4 equal cusps
B	3 equal large cusps + 1 small cusp
C	2 equal large cusps + 2 equal small cusps
D	1 large cusp + 2 equal medium cusps + 1 small cusp
E	1 large cusp + 3 equal small cusps
F	2 equal large cusps + 2 unequal small cusps
G	4 unequal cusps
H	1 large cusps + 1 medium cusp + 2 equal small cusps

Conclusions

QAVs are rare anomalies that are underdiagnosed or often misdiagnosed as BAVs. Detection of these valvular defects with TTE may be insufficient due to variable acoustic windows, poor resolution, limited viewing angles, and potential interference from other pathologies.

Evaluation with TEE overcomes many of these obstacles and is warranted, especially in the presence of severe aortic insufficiency, to appropriately visualize QAVs.

While aortic valve repair, whether with the Ross procedure or other techniques, and aortic valve replacement are available options, a patient-centered and tailored approach should be considered to choose the best intervention for patients with QAVs.

References

Aboud et al. 2021; Alomari et al. 2024; Bietry et al. 2022; D'Errico et al. 2022; Hurwitz & Roberts, 1973; Idrees et al. 2015; Xiao et al. 2010; Zhu et al. 2013; Saith S, Saith S, Murthy A, 2022; Yuan, 2016