5-2020

Quadricuspid aortic valve with aortic insufficiency: a rare echocardiographic finding

Zachary Demertzis
Henry Ford Health System, ZDemert1@hfhs.org

Paul Nona
Henry Ford Health System, PNONA1@hfhs.org

Bryan Zweig
Henry Ford Health System, BZWEIG1@hfhs.org

Quadricuspid aortic valve with aortic insufficiency: a rare echocardiographic finding

Zachary Demertzis DO, Paul Nona MD, Bryan Zweig MD

Department Internal Medicine and Division of Cardiology Henry Ford Health System, Detroit, Michigan

Introduction

- Quadriscupid aortic valve (QAV) is a rare congenital heart defect typically found incidentally without any associated cardiac defects
- Functional status of QAV is pure aortic insufficiency (AI)
- Clinical manifestations are dependent on the functional status of the valve, presenting in the fifth or sixth decade of life due to progressive degeneration of the leaflets
- In our case, we present a young female who developed post-partum dyspnea with elevated brain natriuretic peptide levels concerning for heart failure where on transesophageal echocardiography (TEE) revealed a QAV with AI

Case Presentation

Patient: 37-year-old female with no past medical history 5 days post cesarean section

Chief Complaint: dyspnea (NYHA III)

Physical examination: significant for decrescendo diastolic murmur at the aortic area and bibasilar rales

Pertinent labs: brain natriuretic peptide level elevation of 345 pg/mL with normal troponin I levels (<40 ng/L)

Imaging:

- Chest radiograph significant for pulmonary congestion
- Transthoracic echocardiogram was suggestive for AI
- Transesophageal echocardiogram revealed a QAV with all four leaflets equal in size with normal thickness and mobility. Moderate malcoaptation of all valves was present and severe AI was visualized. Planimetry of aortic regurgitant orifice was measure at 0.29 cm^2, the AI jet was greater than 65% of left ventricular outflow tract, pressure half time calculated at 252 ms, and vena contracta measured 0.6 cm. Her systolic (ejection fraction 60%) and diastolic function were both preserved.

Management:

- She was diuresed with furosemide 40 mg IV twice daily, achieving euvolemia and symptom relief, discharged on furosemide 20mg PO daily

Post-Hospitalization:

- Seen by structural heart and cardiac surgery team, recommended to undergo CT coronary angiography for surgical aortic valve replacement, but lost to follow up

Images

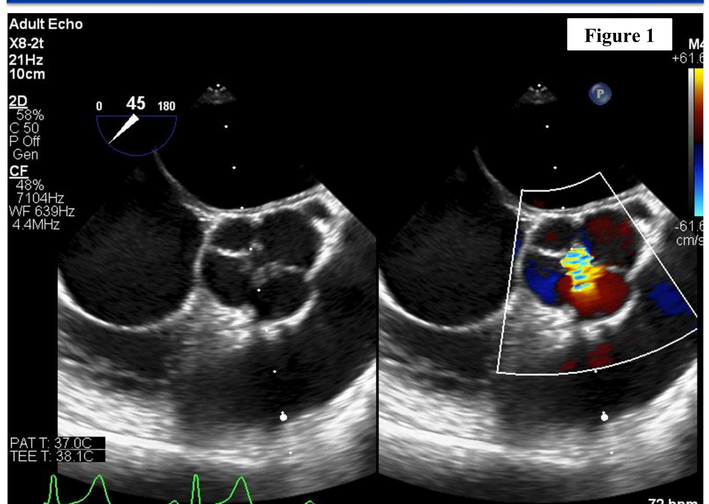

Figure 1

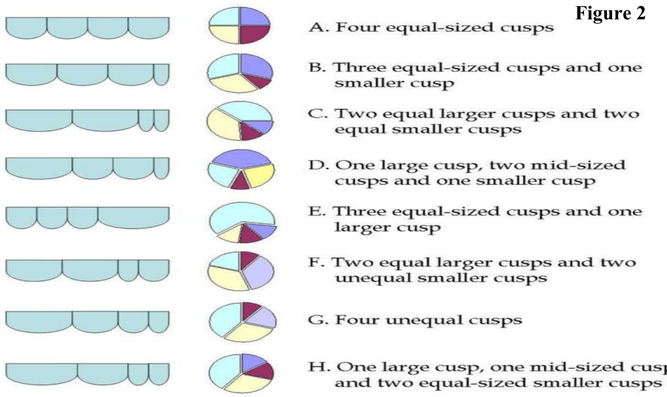

Figure 2

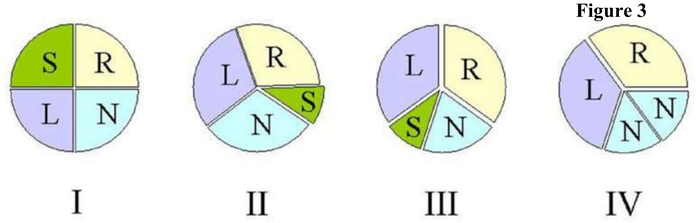

Figure 3

Figure 1. Transesophageal echocardiogram. Mid-esophageal level, aortic valve short axis view. Doppler significant for severe AI.

Figure 2. Hurwitz & Roberts classification of QAV with Vali et al. supplementation

Figure 3. Nakamura et al. simplified classification of QAV

Discussion

- QAV is a rare congenital cardiac anomaly with an incidence rate of 0.00028-0.00033%
- Echocardiography allowed for visualization of the aortic valve and for quantification of the degree of AI
- QAV can present in various anatomic morphologies and can be further classified
- Most prevalent complication of QAV is AI (74.7%)
- Untreated AI can lead to valvular heart failure
- Patients with QAV are at increased risk for infective endocarditis due to progressive degeneration of the leaflets from the asymmetric mechanical stress around the four cusps
- In our case, the peripartum physiologic changes of increased intravascular volume lead to further valvular stress resulting in acute valvular heart failure
- Definitive treatment of QAV with AI is valve replacement, which was recommended to our patient

Conclusion

- QAV is a rare, isolated congenital disease
- Most commonly manifests with AI in the fifth and sixth decade
- QAV is typically found incidentally and best visualized by TEE
- Definitive management of QAV is surgical valve replacement

References

1. Hurwitz LE, Roberts WC. Quadricsupid semilunar valve. Am J Cardiol 1973;31(5):623-626
2. Jagannath AD, Johri AM, Liberthson R, Larobina M, Passeri J, Tighe D, et al. Quadricuspid aortic valve: a report of 12 cases and a review of the literature. Echocardiography. 2011;28(9):1035–1040.
3. Nakamura Y, Taniguchi I, Saiki M, Morimoto K, Yamaga T. Quadricuspid aortic valve associated with aortic stenosis and regurgitation. Jpn J Thorac Cardiovasc Surg. 2001;49(12):714–716
4. Suraci N, Horvath SA, Urina D, Rosen G, Santana O. Quadricuspid aortic valve: case serires and review of literature. Echocardiography 2019;36(2):406-410
5. Vali Y, Rajendra R, Nishtar S. A previously undescribed type of quadricuspid aortic valve: type H. J Heart Valve Dis. 2010;19(6):792–793.
6. Yuan SM. Quadricuspid aortic valve: a comprehensive review. Braz J Cardiovasc Surg 2016;31(6):454-460