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Quadricuspid aortic valve: a case report and review of the literature

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Abstract:

Quadricuspid aortic valve (QAV) is a rare congenital anomaly that can present as aortic insufficiency later in life. We report a case of aortic regurgitation associated with a QAV, treated by aortic valve replacement. The patient presented with breathlessness, lethargy and peripheral oedema. Echocardiography and cardiac magnetic resonance revealed abnormal aortic valve morphology and coronary angiography was normal. The presence of a quadricuspid aortic valve was confirmed intraoperatively. This was excised and replaced with a bioprosthetic valve and the patient recovered well postoperatively. Importantly, the literature indicates that specific QAV morphology and associated structural abnormalities can lead to complications. Hence, early detection and diagnosis of QAV allows effective treatment. Aortic valve surgery is the definitive treatment strategy in patients with aortic valve regurgitation secondary to QAV. However, the long-term effects and complications of treatment of this condition remains largely unknown.

Key words: aortic valve, aortic valve insufficiency, thoracic surgery, cardiac surgical procedures

Word count: 799

Introduction

Quadricuspid aortic valve (QAV) is a rare congenital anomaly[1] that can present as aortic insufficiency requiring treatment later in life. We present a case of aortic regurgitation (AR) associated with a QAV, managed by aortic valve replacement and review of the literature surrounding this unusual presentation.

Case presentation

A 65-year-old caucasian lady was referred with progressive effort-related breathlessness for treatment of longstanding AR. She presented with mild bipedal oedema, a regular pulse collapsing in character, with harsh ejection and early diastolic murmurs on auscultation, and no signs of congestive cardiac failure.

Coronary angiography showed normal coronary arteries. A transthoracic echocardiogram (TTE) and cardiac magnetic resonance (CMR) study showed progression to severe eccentric AR, dilatation of the left ventricle (LV) and preserved LV function (EF of 61%). These revealed her aortic valve to be quadricuspid in morphology (figures 1, 2).

Aortic valve replacement surgery was therefore recommended on symptomatic and prognostic grounds.

Surgery was performed without complication. Intraoperative transoesophageal echocardiography (TOE) confirmed the presence of a quadricuspid aortic valve (figure 3) with severe AR yet good LV function. Macroscopically, the quadricuspid valve appeared heavily calcified (figure 4). The leaflets were excised, and annular debridement performed. A bioprosthetic valve was safely implanted, followed by de-airing, temporary pacing wire insertion, successful weaning off cardiopulmonary bypass and decannulation. The post-operative period was uneventful.

Histopathological analysis of the excised tissue revealed four heavily calcified valve leaflets, two of which fused into a single leaflet with raphe, measuring 27x14x2mm overall. The other two leaflets measured 18x14x5mm and 21x13x4mm. This was confirmed as calcific sclerosis of quadricuspid aortic valve, with no active inflammation.

Discussion

The incidence of QAV is reported between 0.001-0.006%[1], with an equal male-to-female ratio, although males more commonly require surgical treatment[2]. Resulting from defective embryological development of the aortic trunk during gestation, QAV is associated with other congenital cardiac conditions, including coronary artery anomalies such as presence of a single coronary ostium, an accessory artery or displaced coronary ostia due to the accessory cusp[3,4]. Other associations include hypertrophic cardiomyopathy, subaortic stenosis, patent ductus arteriosus, ventricular septal defects, ruptured sinus of Valsalva, complete heart block and endocarditis[2].

Historically, variations of QAV are classified by the configuration of their cusps[5], but there is no established correlation between the anatomy and functional status of the aortic cusps, and no resulting prognostic implications.

Of those that present clinically, 75% manifest as aortic insufficiency producing symptoms of exertional breathlessness, presyncope and fatigue, often in the fifth and sixth decades[6], and half require surgical treatment[6]. AR results from inadequate valve closure due to progressive valve leaflet thickening and asymmetric mechanical stress causing abnormal leaflet coaptation[7]. Less commonly, QAV presents with normal valve function or aortic stenosis. The natural history of QAV remains largely unknown but concomitant aortic incompetence can lead to further aortopathy, congestive cardiac failure and death[2].

QAV can be detected by imaging[6]. Echocardiography and cardiac MRI allow characterisation of the valve, its inflow and outflow tracts, its dimensions, associated great vessel anatomy (importantly, the

aortic root and arch), quantification of flow and ventricular volumes and function. QAV is suggested by an 'X' shape seen on short-axis TTE view of the aortic valve[8], more visibly on TOE.

Particularly important for surgical management, is to delineate valvular anatomy and identify displacement of the coronary ostia to avoid obstructing coronary blood flow during valve surgery[9]. Coronary angiography or multidetector coronary CT (MDCT) is therefore recommended in the preoperative assessment of QAV[10].

Non-operative treatment includes medical management of symptoms and secondary prevention of cardiac failure. Long-term survival is good in patients who do not develop any haemodynamic compromise[2].

Aortic valve surgery provides the best long-term survival in QAV with valve insufficiency[2]. The most common indication for surgery is severe AR[2,6]. Although intraoperative findings ultimately dictate surgical strategy, aortic valve replacement using a bioprosthetic valve is preferable to valve repair[1]. Furthermore, valve replacement permits histopathological analysis to exclude concurrent microscopic valve disease.

Aortic valve repair is feasible. Tricuspidization restores normal geometry to the leaflets and functional aortic annulus, restoring a normal surface of coaptation[1]. However, such techniques are often reserved for the paediatric population and patients that benefit from avoiding a prosthetic valve, depending on specific QAV morphology and functional status.

Alternatively, subcoronary aortic valve replacement with a patient's own pulmonary valve (the Ross procedure), has been reported as an effective treatment for QAV to mitigate the risk of aortic root dilation[2,11]. However, the true risk of such complications is unknown and more research is needed to establish the long-term sequelae, before supporting this recommendation[1,12].

Conclusions

Early detection and diagnosis of QAV has significant implications for treatment. Delineating specific QAV morphology and identifying any associated structural abnormalities is vital to avoid complications and determine the most effective treatment strategy. Aortic valve surgery is the definitive treatment in patients who develop severe aortic valve regurgitation secondary to QAV.

References

1. Idrees J, Roselli E, Arafat A, Johnston D, Svensson L, Sabik J et al. Outcomes after repair or replacement of dysfunctional quadricuspid aortic valve. J Thorac Cardiovasc Surg. 2015;150(1):79-82.
2. Tsang M, Abudiab M, Ammash N, Naqvi T, Edwards W, Nkomo V et al. Quadricuspid Aortic Valve. Circulation. 2016;133(3):312-319.
3. Bogers A, Gittenberger-de Groot A, Poelmann R, Péault B, Huysmans H. Development of the origin of the coronary arteries, a matter of ingrowth or outgrowth?. Anatomy and Embryology. 1989;180(5):437-441.
4. Yassine N, Shahram J, Body S. Pathogenic Mechanisms of Bicuspid Aortic Valve Aortopathy. Frontiers in Physiology. 2017;8.
5. Hurwitz L, Roberts W. Quadricuspid semilunar valve. The American Journal of Cardiology. 1973;31(5):623-626.
6. Tutarel, O. The quadricuspid aortic valve: a comprehensive review. The Journal of Heart Valve Disease. 2004;13(4), 534-537.
7. Yuan S. Quadricuspid Aortic Valve: A Comprehensive Review. Brazilian Journal of Cardiovascular Surgery. 2016;31(6):454–460.
8. Jagannath A, Johri A, 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.
9. Savino K, Quintavalle E, Ambrosio G. Quadricuspid aortic valve: A case report and review of the literature. Journal of Cardiovascular Echography. 2015;25(3):72.

10. Douglas H, Moore M, Purvis J. Comprehensive assessment of a quadricuspid aortic valve and coronary arteries by multidetector cardiac CT. Heart. 2012;98(24):1838-1838.
11. Misfeld M, Christiansen F, Sievers HH. Ross procedure in a quadricuspid aortic valve. Ann Thorac Surg. 2005;80:1110-1.
12. Timperley J, Milner R, Marshall A, Gilbert T. Quadricuspid aortic valves. Clinical Cardiology. 2002;25(12):548-552.

Tables None.

Figures

Figure 1. TTE.

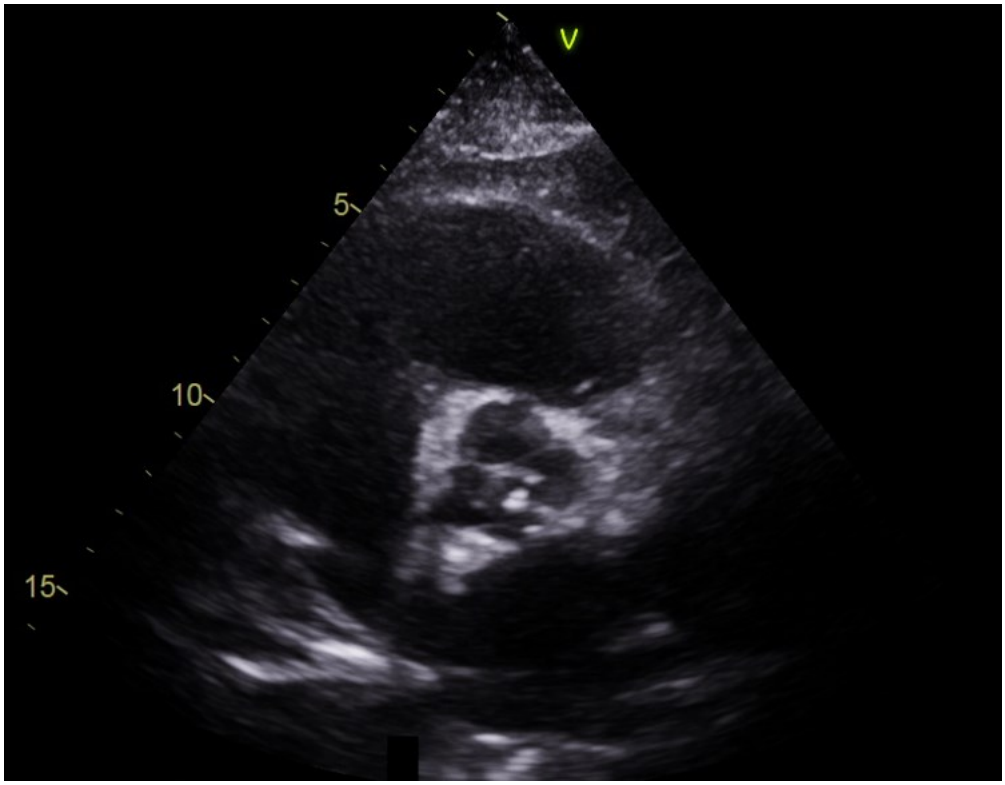


Figure 2. CMR.

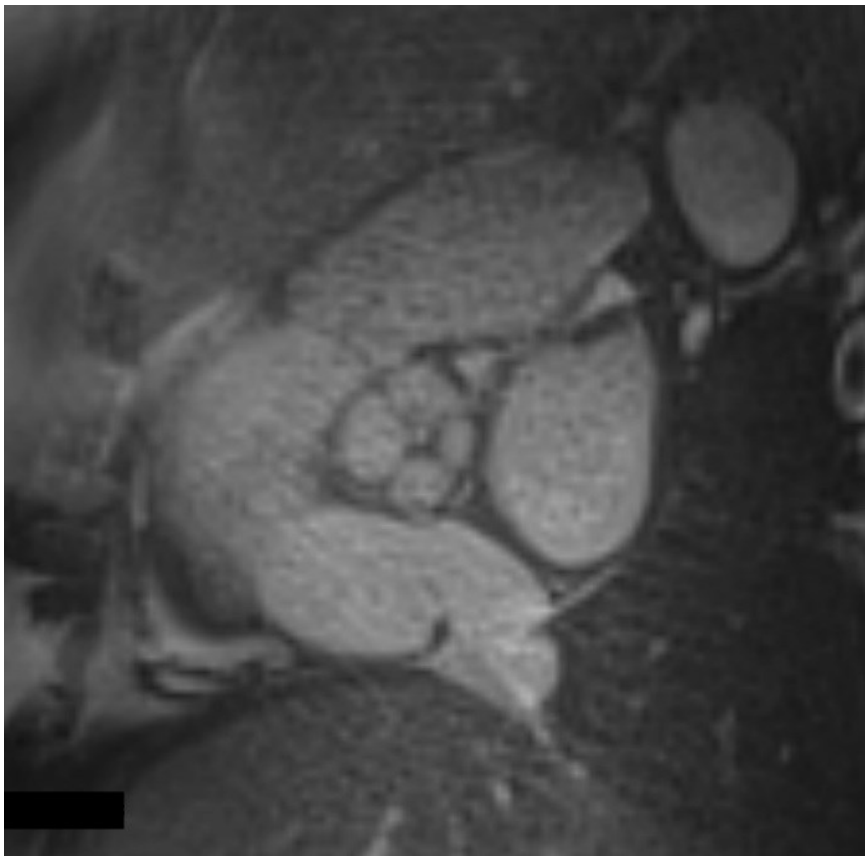


Figure 3. TOE.

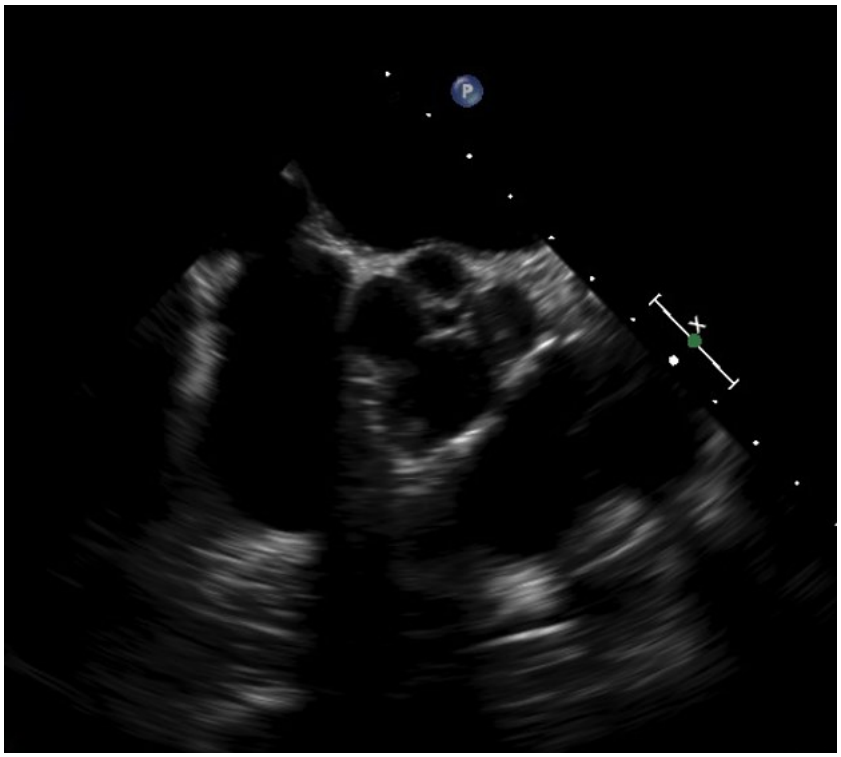
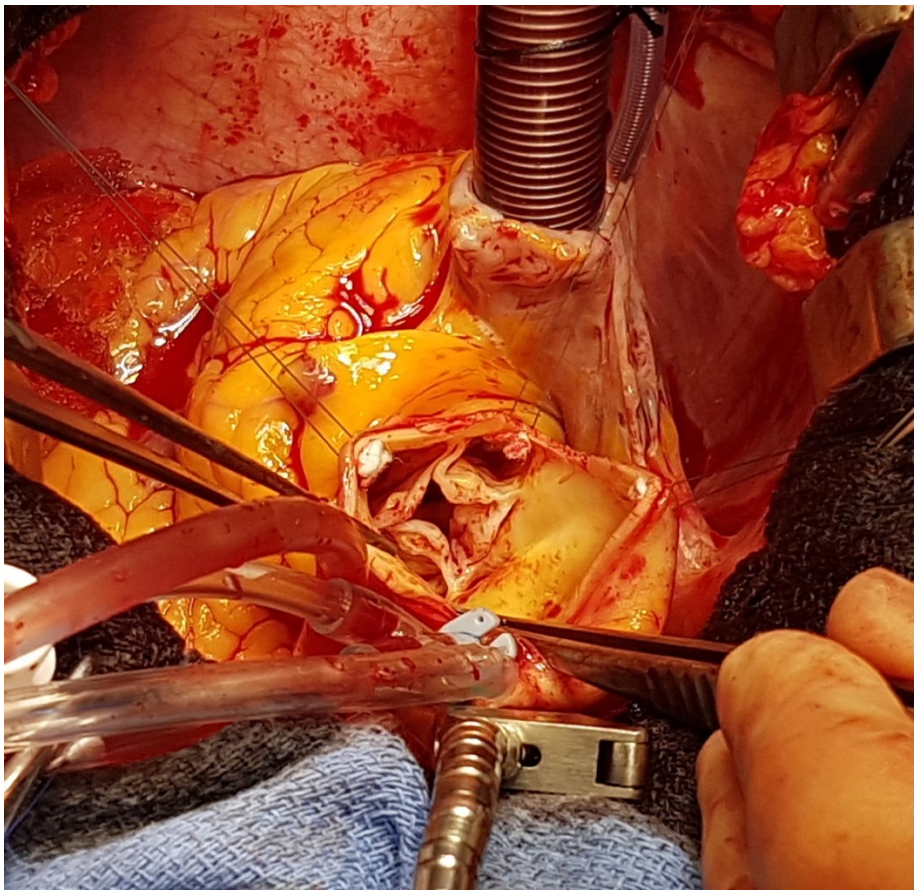


Figure 4. Macroscopic appearance.



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