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# Case Report

# Cardiovascular computed tomography for the detection of quadricuspid aortic valve: A case report<sup>☆</sup>

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#### ARTICLE INFO

Article history: Received 23 April 2023 Revised 20 June 2023 Accepted 23 June 2023

Keywords: Aortic regurgitation Cardiac CT Valvular heart disease Aortic valve Cardiac surgery

## ABSTRACT

Aortic regurgitation (AR) represents a significant cause of morbidity and mortality. Due to its low cost and widespread availability, echocardiography remains the frontline for aortic valve (AV) assessment. However, poor sonographic windows may limit the assessment of valve morphology with this technique. Cardiovascular computed tomography (CCT) is increasingly utilized prior to structural AV interventions. Due to its excellent spatial resolution, CCT provides exceptional characterization of aortic leaflets. Accordingly, we present a case of a quadricuspid valve diagnosed by CCT. Here, CCT led to a new diagnosis of quadricuspid valve, highlighting the potential for CCT for the characterization of aortic leaflet morphology. CCT may be particularly useful in patients with contraindications to transesophageal echocardiography or those undergoing structural or robotic interventions.

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## Background

Aortic regurgitation (AR) represents a significant cause of morbidity and mortality. Due to its low cost and widespread availability, echocardiography remains the frontline for aortic valve (AV) assessment. However, poor sonographic windows may limit the assessment of valve morphology with this technique. Cardiovascular computed tomography (CCT) is increasingly utilized prior to structural AV interventions. Due to its excellent spatial resolution, CCT provides exceptional characterization of aortic leaflets. Accordingly, we present a case of a patient with equivocal AV morphology on TTE that is ultimately diagnosed with quadricuspid AV by preoperative CCT.

## **Case description**

A 49F with a history of asthma was evaluated in primary care for progressive leg swelling. The patient was referred to a cardiologist. An echocardiogram was performed, demonstrating

Abbreviations: AR, aortic regurgitation; AV, aortic valve; AVR, aortic valve replacement; CCT, cardiovascular computed tomography.

 $<sup>^{\</sup>star}\,$  Competing Interests: The authors affirm that they have no relevant conflicts of interest to disclose.

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https://doi.org/10.1016/j.radcr.2023.06.056

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Fig. 1 – Key echocardiographic findings in the case. Parasternal long axis demonstrating aortic leaflet malcoaptation (A, Solid Arrow) with severe aortic regurgitation (B, Dashed Arrow). Leaflets were incompletely visualized without clear evidence of quadricuspid morphology (C). Suprasternal notch view demonstrating holodiastolic flow reversal (D, Arrow Head) consistent with severe aortic regurgitation.

aortic leaflet malcoaptation with severe, centrally directed aortic regurgitation; the aortic root was normal in size, and aortic valve leaflet morphology was not well visualized due to technically difficult sonographic windows (Figs. 1 A-C). There was holodiastolic flow reversal in the descending aorta consistent with severe AR (Fig. 1D). The patient was referred to our institution for evaluation for robotic endoscopic aortic valve replacement (AVR) as a less invasive option. A CCT was performed for preoperative planning, revealing a quadricuspid aortic valve with significant central malcoaptation as the mechanism of AR. (Fig. 2, Supplemental Video 1). The patient subsequently underwent successful robotic endoscopic aortic valve repair, where the accessory cup was excised and the noncoronary cusp was reconstructed to approximate the size of a normal cusp, with the end result being a trileaflet valve. Intraoperative transesophageal echocardiogram (TEE) confirmed the finding of quadricuspid valve seen on CCT

(Fig. 3, Supplemental Video 2). The patient ultimately had a good functional recovery with only mild residual AR.

### Discussion

Quadricuspid aortic valve is a rare defect that most commonly leads to AR [1]. Hurwitz and Robertson's classification system has identified seven subtypes. Type B is the most common subtype, involving three equal-sized larger cusps and one smaller cusp (Fig. 4) [1]. Thus, the theory for AR is from unequal sheer forces and leaflet degeneration. Patients should be managed according to guidelines: AVR is recommended for those with severe AR symptoms, LV dilation or dysfunction, or concurrent cardiac surgery.



Fig. 2 – CCT findings demonstrating quadricuspid leaflet morphology on both 2-D and 3-D imaging with left (L), right (R), noncoronary (N), and diminutive fourth (\*) cusp present.



Fig. 3 – Intraoperative TEE findings confirm the diagnosis of quadricuspid aortic valve with left (L), right (R), noncoronary (N), and diminutive fourth (\*) cusp present (Hurwitz Type B).



Fig. 4 – Morphological classification for quadricuspid aortic valve, as proposed by Hurwitz & Roberts (From Braz J Cardiovasc Surg. 2016 Nov-Dec; 31(6): 454–460.).

A unique aspect of our case was that in this patient, CCT was critical for making the correct diagnosis of quadricuspid AV. While echocardiography is the frontline for AR evaluation, its accuracy for detecting aortic leaflet number may be limited, for instance with poor image quality or excessive leaflet calcification. Due to excellent spatial resolution, CCT provides exceptional visualization of aortic valve leaflets. Previous studies have demonstrated superior accuracy of CCT compared to transthoracic echocardiography for differentiation between bicuspid and trileaflet valves [2].

The identification of quadricuspid AV by CCT was crucial in our case as this finding ultimately led the surgeons to pursue aortic valve repair rather than replacement. Although AVR is often the standard of care for aortic regurgitation, it is less ideal in younger patients given the increased potential for structural valve degeneration, need for reoperation, or thromboembolism [3,4]. Aortic valve repair, on the other hand, has been studied to have significant survival benefits compared to aortic valve replacement for this cohort of patients [5]. CCT is important for aortic valve repair, as precise preoperative planning is necessary - requiring accurate measurements such as the aortic annulus, sinus of Valsalva, sinotubular junction, and ascending aorta [6]. Thus, CCT is not only important in facilitating the diagnosis of quadricuspid aortic valves, but it is also essential for preoperative planning for patients to undergo successful aortic valve repair. For this and other reasons, is our standard practice to perform CCT prior to robotic surgery for preoperative planning.

In summary, CCT led to a new diagnosis of quadricuspid valve, highlighting the potential for CCT for the characterization of aortic leaflet morphology. CCT may be particularly useful in patients with contraindications to transesophageal echocardiography or those undergoing structural or robotic interventions.

## Patient consent

We affirm that informed written consent was obtained from the patient for the publication of this case report. The patient affirmed that de-identified clinical information and images would be used for the purposes of this publication. The authors of the manuscript have retained this informed consent which may be provided to the journal if specifically requested.

#### Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2023.06.056.

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