

Cardiac Computed Tomography and Quadricuspid Aortic Valve: A Case Report

Carrie B. Chapman, MD; Takushi Kohmoto, MD; Annie F. Kelly, MD;
Frank Thornton, MD; Jon G. Keevil, MD

ABSTRACT

A quadricuspid aortic valve is rare and often incidentally found by echocardiography, surgically, or on post mortem examination. Aortic regurgitation is common and if severe enough can lead to symptoms of dyspnea. We report a case of a quadricuspid aortic valve, which was found by cardiac multidetector computed tomography during a pre-operative assessment for severe aortic regurgitation.

CASE DESCRIPTION

A 35-year-old previously healthy woman was sent for echocardiography after her allergist auscultated a murmur during an evaluation for persistent cough. Her history was significant for allergies, controlled hypertension, mild glucose intolerance, and occasional tobacco use. She had no family history of coronary artery disease (CAD), sudden death, or known valvular abnormalities. Symptomatically, the patient reported worsening shortness of breath over the previous several months associated with exertional chest heaviness.

A transthoracic echocardiogram (TTE) demonstrated severe aortic regurgitation by color Doppler. The short axis view, however, was inadequate to evaluate the numbers of aortic valve cusps, likely due to the patient's body mass index of 38. The left ventricle was found to be severely enlarged, with an end-diastolic dimension of 79 mm and end-systolic dimension of 56 mm, and the ejection fraction was mildly reduced to 50%. Additionally, the ascending aorta was noted to be mildly enlarged at 40 mm.

Given this information, it was decided to pursue cardiac computed tomography (CT) both to define the size of the patient's aorta and to rule out coronary atherosclerosis. Coronary CT angiography (CTA) is known to be highly accurate compared to conventional coronary angiography and has been validated in the preoperative setting for chronic aortic regurgitation.¹ Thus, invasive cardiac catheterization, which was previously uniformly done prior to aortic valve surgery, could be avoided.

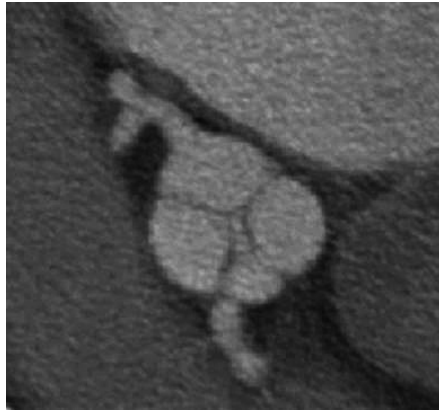
The cardiac CT was performed on a 64-slice GE VCT scanner (General Electric, Waukesha, Wisconsin). The patient underwent standard protocol with beta blockade and nitroglycerin sublingually prior to scanning. The images were obtained by retrospective gating and reconstructed using a dedicated Advantage workstation. The patient was administered 21.9 mSv of radiation and 87 ml of contrast. Overall, the quality of the CT images was good.

Figure 1A shows the aortic valve in diastole with both the right and left coronary arteries at their insertion points. The valve can be described as a Type B variant with 3 equal cusps and 1 smaller accessory cusp,² now felt to be the second most common described in the literature.³ Type A variant is more common and has 4 equal cusps; however, Type B is more likely to lead to aortic regurgitation. The 3 larger cusps include a left coronary cusp, right coronary cusp, and non-coronary cusp. The CT image demonstrates the lack of coaptation of the valve leaflets with a regurgitation orifice that measured 20 mm². The remainder of the CTA demonstrated large coronary arteries free from calcium or plaque. The ascending aortic root was only mildly enlarged at 43 mm. The left ventricular ejection fraction was calculated to be 55%, and the end-systolic volume was 145 ml.

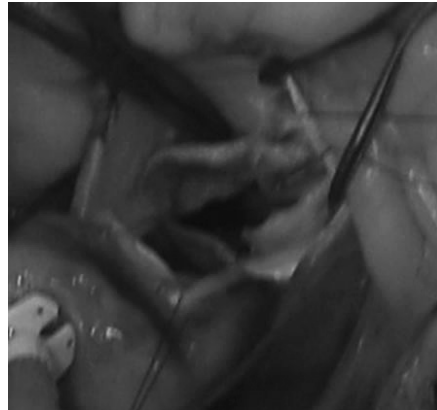
The patient subsequently underwent surgical replacement of her severely insufficient aortic valve. Intraoperative pictures by a transesophageal echocardiogram (TEE) and photography confirmed the diag-

Author Affiliations: Division of Cardiovascular Medicine, University of Wisconsin Hospital and Clinics, Madison, Wis (Chapman, Kelly, Keevil); Division of Cardiothoracic Surgery, University of Wisconsin Hospital and Clinics, Madison, Wis (Kohmoto); Division of Radiology, University of Wisconsin Hospital and Clinics, Madison, Wis (Thornton).

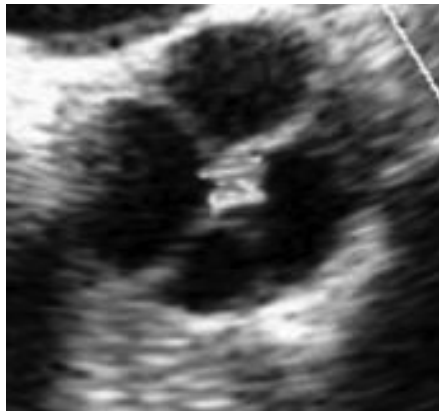
Corresponding Author: Carrie B. Chapman, MD, 600 Highland Ave, Madison, WI 53792; phone 608.263.0891; fax 608.263.0405; e-mail cbchapma@medicine.wisc.edu.



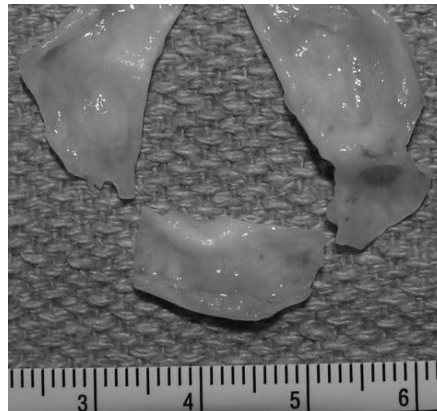
A. Cardiac CT of aortic valve and coronary arteries.



C. Intraoperative view of aortic valve with coronary arteries identified by forceps.



B. TEE short axis of aortic valve with aortic insufficiency.



D. Post-operative gross specimen of aortic valve.

Figure 1. Quadricuspid Aortic Valve.

nosis obtained by cardiac CT (Figures 1B, 1C, 1D). The cusps are labeled to coincide with the CT image.

DISCUSSION

A quadricuspid aortic valve remains a rare finding. In 2004, Tutarel performed an in-depth literature review and identified 186 cases published.³ Aortic regurgitation was found in approximately 75% of the cases, with 9% having both aortic stenosis and regurgitation, and 16% having a normal functioning aortic valve. Symptoms and progressive valvular dysfunction were significant enough to require surgery for 45.2% of the subjects in this series. Several associated cardiac abnormalities were also identified, including hypertrophic cardiomyopathy, atrial septal defects, patent ductus arteriosus, and, most commonly, anomalous coronary arteries.^{3,4}

The estimated incidence of quadricuspid aortic valve has varied from 0.008% by autopsy to 0.043% by TTE,

both of which likely underestimate the true incidence.^{4,5} The first reported case was in 1862, described at autopsy by Balington.⁶ In 1969, a quadricuspid aortic valve was imaged by aortography and described by Peretz.⁷ Transthoracic 2-dimensional echocardiography became available in the 1970s with several cases of quadricuspid aortic valves being described in 1984.^{8,9} Today, TEE is considered the superior imaging test for a clear anatomical image of a quadricuspid aortic valve. It is particularly useful in defining abnormally placed coronary ostia, which can affect valve replacement surgery.^{10,11} Three-dimensional TTE has been used but currently produces images inferior to TEE.¹² Recent reports of cardiac magnetic resonance imaging to diagnose a quadricuspid aortic valve have also been published with good image quality^{13,14} and case reports using multidetector CT imaging are now being published, also with excellent images.^{14,15}

In our patient, cardiac CT provided clear simultaneous images of a quadricuspid aortic valve, mild aortic root dilation, location of coronary ostia, and absence of coronary atherosclerotic disease. Pathologic specimens verified the quadricuspid valve that was identified. In this case, the transthoracic echocardiographic images were inadequate to completely define our patient's anatomy. Cardiac CT allowed for a non-invasive modality to obtain this information and also allowed for improved pre-surgical planning by our cardiothoracic surgeons.

There are currently multiple appropriate uses of cardiac CT that have been established by the American College of Cardiology. Several examples include coronary evaluation in patients with intermediate risk of CAD presenting with acute chest pain who have both negative enzymes and ECG, patients with an uninterpretable stress test, and patients with suspected coronary anomalies, congenital heart disease, or valvular abnormalities.¹⁶ The negative predictive value of CTA has been reported near or at 100%. A lesion of >50% or more by CTA would, however, subsequently need to be evaluated by catheterization. In 1 study of subjects needing aortic valve surgery for aortic regurgitation, catheterization could have been avoided in 70% of patients.¹

Cardiac CT could conceivably replace both TEE and cardiac catheterization preoperatively for aortic valve surgery, which would reduce the number of diagnostic studies performed and patient risk. Radiation and contrast exposure with cardiac catheterization is similar compared to CTA; however, several risks associated with the invasive nature of catheterization—including stroke, myocardial infarction, and bleeding—are not a factor. In conclusion, for the appropriate patient, cardiac CT is a reasonable modality for the preoperative evaluation of aortic valve insufficiency and can detect a previously undiagnosed quadricuspid aortic valve.

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