Two Reports of Quadricuspid Aortic Valve With Aortic Insufficiency

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We report two cases of a quadricuspid aortic valve with severe aortic incompetence. Both patients presented with dyspnea on exertion. Their physical examinations demonstrated wide pulse pressures with diastolic murmurs. Bedside transthoracic Doppler echocardiography revealed preserved left ventricular systolic function and possible quadricuspid aortic valve with severe aortic incompetence in both patients. We proceeded with transesophageal echocardiography that confirmed a quadricuspid aortic valve with severe aortic incompetence in both patients. Left ventricular systolic function was preserved in both cases. Both patients had a preoperative cardiac catheterization, which showed normal coronary arteries. They were referred to cardiothoracic surgery and underwent successful aortic valve replacement with bioprosthetic valves.

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KEY WORDS

Quadricuspid aortic valve • Aortic insufficiency • Aortic valve replacement

Case Reports

Case 1

A 60-year-old woman was referred to the cardiology clinic with dyspnea on exertion for several months; she had no noteworthy past medical or family history, and was a lifelong nonsmoker.

On examination, she had a pulse rate of 82 beats/min with collapsing character and a blood pressure of 110/44 mm Hg. She had no raised jugular venous pressure and no pedal edema. On auscultation, an early diastolic murmur at the aortic area and a soft second heart sound were detected.

Transesophageal echocardiography (TEE) showed a normal-sized left ventricle with normal systolic function and quadricuspid aortic valve (QAV) with malcoaptation (Figure 1). Severe central aortic incompetence was observed (Figure 2). Effacement of the sinuses with mild mid-aortic dilatation of 4 cm was also detected. The mitral valve was structurally and functionally normal with an intact intra atrial septum. The patient then proceeded to have a successful aortic valve replacement with a bioprosthetic valve.

Case 2

A 64-year-old woman with a history of lobectomy for a bronchial adenoma was referred to the rapid-access chest pain clinic with dyspnea on exertion. She was a nonsmoker and had no family history of cardiac illnesses.

On examination, she had a pulse rate of 90 beats/min with collapsing character and a blood pressure of 125/55 mm Hg. She had no raised jugular venous pressure and no pedal edema. Auscultation detected an early diastolic murmur at the aortic area.

TEE showed a dilated left ventricle with preserved systolic function and QAV (Figure 3) with severe aortic incompetence directed centrally (Figure 4). A sinus of Valsalva aneurysm was noted. A structurally and functionally normal mitral valve and normal aortic root dimensions above the level of the sinus of Valsalva were also observed. The patient underwent a successful aortic valve replacement with a bioprosthetic valve along with repair of the dilated sinus with the help of interrupted sutures incorporated into the aortic annulus.

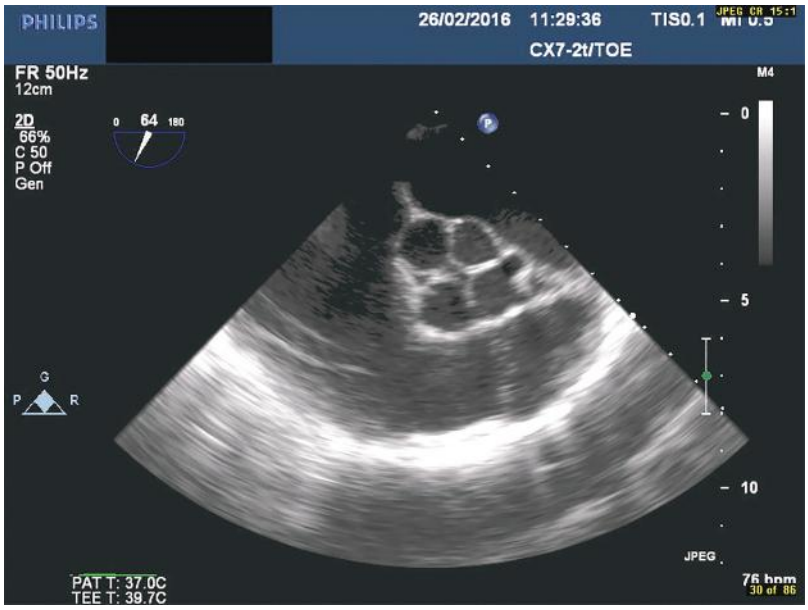

Figure 1. Image of an upper position transesophageal echocardiography view showing a quadricuspid aortic valve in a 60-year-old woman with dyspnea (Case 1).

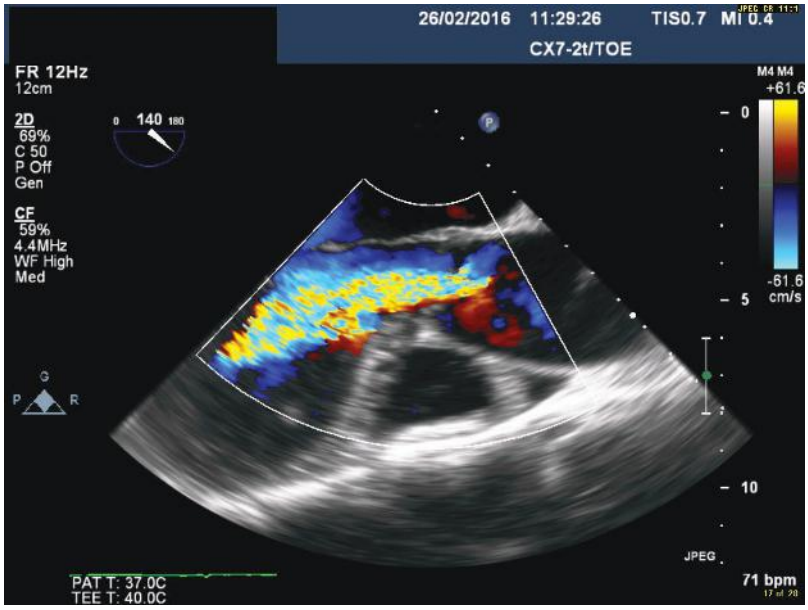

Figure 2. Transesophageal echocardiography image of a color-flow across the aortic valve showing regurgitation in a 60-year-old woman with dyspnea (Case 1).

QAV is a rare cardiac valvular anomaly characterized by the presence of four cusps, instead of the usual three cusps, in the aortic valve.

Discussion

QAV is a rare cardiac valvular anomaly characterized by the presence of four cusps, instead of the usual three cusps, in the aortic valve. QAV is an uncommon clinical finding[1,2]; its prevalence ranges between 0.013% and 0.043%.[3] Some reports suggest that this anomaly may occur in up to 1% of individuals who present for aortic valve surgery.[4] QAV is typically detected by echocardiography, or at the time of aortic valve surgery by aortography, or during autopsy.[5] Due to the advancements in echocardiographic imaging, the diagnosis

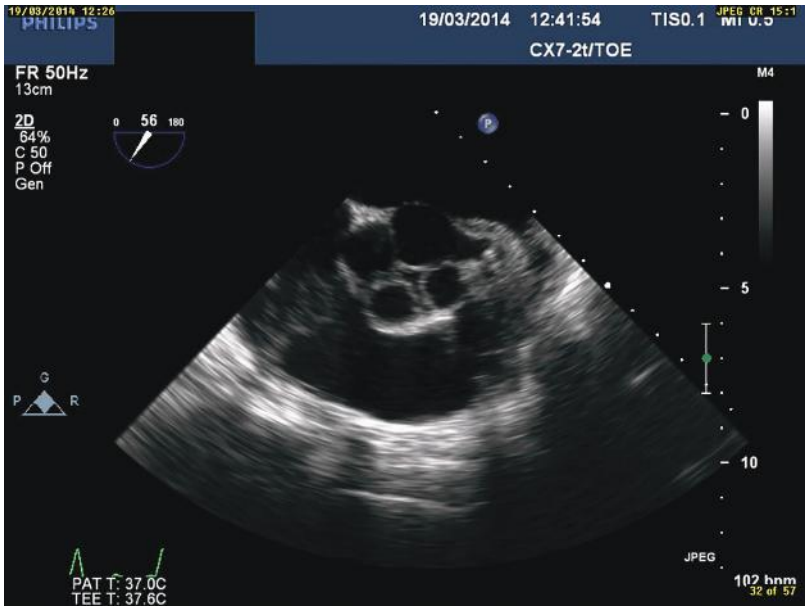

Figure 3. Image of an upper position transesophageal echocardiography view showing a quadricuspid aortic valve in a 64-year-old woman with a history of lobectomy for a bronchial adenoma and dyspnea on exertion (Case 2).

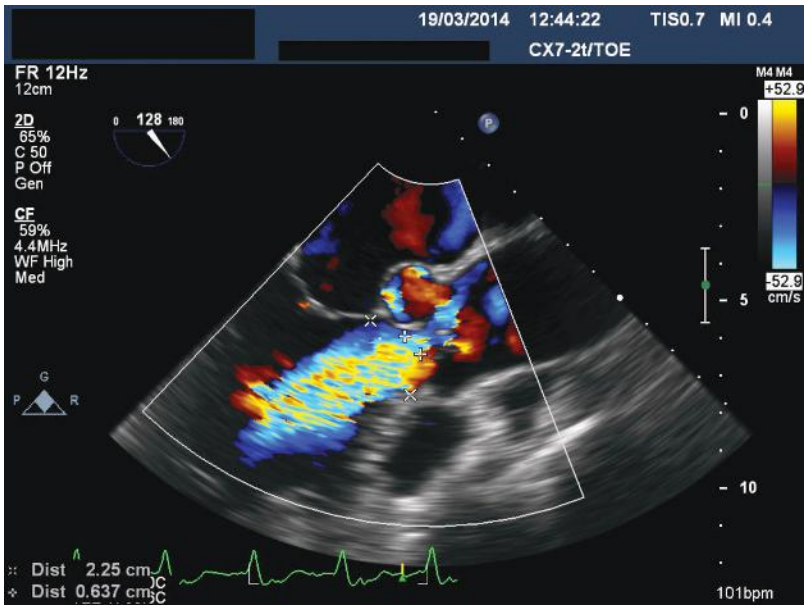

Figure 4. Transesophageal echocardiography image of a color-flow across the aortic valve showing regurgitation in a 64-year-old woman with a history of lobectomy for a bronchial adenoma and dyspnea on exertion (Case 2).

The first known case of QAV was reported by Balington in 1862.[6] The first case diagnosed with aortography in the left anterior oblique view was reported by Peretz and colleagues.[7] It is important to identify the anomaly for two reasons: (1) for its association with aortic insufficiency, and (2) for the possibility of anomalously placed coronary ostia due to the presence of an additional leaflet. A comprehensive review of 186 previously published cases found pure aortic regurgitation of any degree in almost 75% of cases.[8] Another study that included 50 patients with QAV revealed that moderate or severe aortic incompetence was present in 26% of these patients, and 29% had aortic dilatation at the time of diagnosis.[9]

Hurwitz and Roberts categorized quadricuspid valves based on the size of the leaflets into seven anatomic variations (Table 1).[10] Over 85% of the reports of QAV are of type A, B, or C (valves with four equal cusps, three equal cusps with one smaller cusp, or two equal larger and two equal smaller cusps, respectively). The echocardiographic appearance of the valve can sometimes be different from its appearance during surgery. Aortic incompetence usually occurs secondary to fibrous thickening with incomplete coaptation, resulting in unequal distribution of stress. Aortic regurgitation is rarely seen in young individuals with QAV; aortic stenosis can occur but is rare.[11] Some QAVs are associated with other abnormalities, such as displacement of the coronary sinus and ostium, ventricular septal

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Some QAVs are associated with other abnormalities, such as displacement of the coronary sinus and ostium, ventricular septal defect, patent ductus arteriosus, subaortic stenosis, cardiomyopathy, Valsalva aneurysm, and mitral valve regurgitation.

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TABLE 1

Hurwitz and Roberts QAV Classification

QAV Type	Description
A	Four equal cusps
B	Three equal cusps and one smaller cusp
C	Two equal larger and two equal smaller cusps
D	One large, two intermediate and one small cusp
E	Three equal cusps and one larger cusp
F	Two equal larger cusps and two unequal smaller cusps
G	Four unequal cusps

QAV, quadricuspid aortic valve.

defect, patent ductus arteriosus, subaortic stenosis, cardiomyopathy, Valsalva aneurysm, and mitral valve regurgitation.[12-14]

The typical method of treatment is through surgery such as aortic valve reconstruction surgery and aortic valve replacement, usually with a synthetic valve.[15] The duration of follow-up for asymptomatic patients is not clear, but close follow-up is recommended because valve replacement is frequently required in the fifth or sixth decade of life.[11] ■

References

1. Tang YF, Xu JB, Han L, et al. Congenital quadricuspid aortic valve: analysis of 11 surgical cases. *Chin Med J (Engl)*. 2011;124:2779-2781.
2. Holm H, Jacobson S, Reul GJ, Stainback RF. Quadricuspid aortic valve. *Tex Heart Inst J*. 2004;31:450-451.
3. Xiao Z, Meng W, Zhang E. Quadricuspid aortic valve by using intraoperative transesophageal echocardiography. *Cardiovasc Ultrasound*. 2010;8:36.
4. Shankar B, Mehrotra R, Bansal M, et al. Quadricuspid aortic valve. *J Assoc Physicians India*. 2012;60:54-55.
5. Robicsek F, Sanger PW, Daugherty HK, Montgomery CC. Congenital quadricuspid aortic valve with displacement of the left coronary orifice. *Am J Cardiol*. 1969;23:288-290.
6. Balington J. *London Medical Gazette*. July 1862.
7. Peretz DI, Changfoot GH, Gourlay RH. Four-cusped aortic valve with significant insufficiency. *Am J Cardiol*. 1969;23:291-293.
8. Tutarel O. The quadricuspid aortic valve: a comprehensive review. *J Heart Valve Dis*. 2004;13:534-537.
9. Tsang MY, Abudiab MM, Ammash NM, et al. Quadricuspid aortic valve: characteristics, associated structural cardiovascular abnormalities, and clinical outcomes. *Circulation*. 2016;133:312-319.
10. Hurwitz LE, Roberts WC. Quadricuspid semilunar valve. *Am J Cardiol*. 1973;9:623-626.
11. Timperley J, Milner R, Marshall AJ, Gilbert TJ. Quadricuspid aortic valves. *Clin Cardiol*. 2002;25:548-552.
12. Janssens U, Klues HG, Hanrath P. Congenital quadricuspid aortic valve anomaly associated with hypertrophic non-obstructive cardiomyopathy: a case report and review of the literature. *Heart*. 1997;78:83-87.
13. Ozlü MF, Ozcan F, Cagli K, Gölbasi Z. Quadricuspid aortic valve in a patient with mitral valve prolapse. *Can J Cardiol*. 2008;24:e27.
14. Pitta SR, Kondur A, Afonso L. Quadricuspid aortic valve associated with unruptured sinus of Valsalva aneurysm. *Eur J Echocardiogr*. 2008;9:575-576.
15. Song MG, Yang HS, Lee DH, et al. Mid-term results in patients having tricuspidization of the quadricuspid aortic valve. *J Cardiothorac Surg*. 2014;9:29.

MAIN POINTS

- It is important to consider such a rare, but possible, anomaly in individuals presenting with symptoms and signs suggestive of valvular regurgitation.
- Quadricuspid aortic valve (QAV) predominantly causes aortic regurgitation.
- We should regularly follow up patients with QAV, because most patients may require valve replacement with worsening aortic regurgitation in the fifth or sixth decade of life.