Case report

**Quadricuspid aortic valve not discovered by transthoracic echocardiography**

Magnus Dencker*¹ and Martin Stagmo²

Address: ¹Dept of Clinical Sciences, Unit of Clinical Physiology and Nuclear Medicine, University Hospital MAS, Lund University, Malmö, Sweden and ²Dept of Cardiology, University Hospital MAS, Lund University, Malmö, Sweden

Email: Magnus Dencker* - magnus.dencker@skane.se; Martin Stagmo - martin.stagmo@skane.se

* Corresponding author

**Abstract**

**Background:** Quadricuspid aortic valve is a rare congenital heart defect. Several different anatomical variations of a quadricuspid aortic valve has been described and aortic regurgitation is the predominant valvular dysfunction associated with quadricuspid aortic valve.

**Case presentation:** A 68-year-old woman presented with almost a years history of increasing dyspnoea on exertion. The patient have had two previous transthoracic echocardiographic exams in the last six years and they had only documented moderate aortic regurgitation. Transoesophageal echocardiography displayed a quadricuspid aortic valve with three cusps of equal size and one larger cusp. The malformation was associated with severe aortic regurgitation.

**Conclusion:** Liberal use of transoesophageal echocardiography is often warranted if optimal display of valvular morphology is desired.

**Background**

Quadricuspid aortic valve is a rare congenital heart defect. It has in the past been an incidental finding at open heart surgery or at autopsy. There has in recent years been a few case report, with the increasing use of echocardiography [1-8].

**Case presentation**

A 68-year-old woman presented with almost a years history of increasing dyspnoea on exertion. On physical examination, her blood pressure was 200/90 mm Hg and a diastolic murmur of grade 5/6 was heard at the left sternal border. ECG displayed, sinusrhythm with high QRS-voltage and ST-T configuration consistent with left ventricular hypertrophy. Chest X ray was normal. The patient have had two previous transthoracic echocardiographic exams in the last six years and they had only documented moderate aortic regurgitation. Transoesophageal echocardiography displayed a quadricuspid aortic valve. Three cusps were of equal size and one cusp was larger (Figure 1 in diastole and Figure 2 in systole). Planimetric evaluation of the 4 separate cusps performed on an end diastolic still frame resulted in following cusp areas (cm²): 1.4, 1.4, 1.5, and 2.2. The three equal cusps had minimal differences in cusp area whereas the larger cusp had a approximately 50% larger cusp area. In this patient the quadricuspid aortic valve malformation was associated with severe aortic regurgitation (Figure 3).

**Discussion**

Knowledge of the existence of quadricuspid aortic valve is not new, the first reported case dates back to 1862 [8].
Transoesophageal images, in diastole, of a quadricuspid aortic valve with three cusps of equal size and one larger cusp.  

Figure 1
Transoesophageal images of a quadricuspid aortic valve in systole.  

Figure 2
incidence from autopsy studies matches that detected by echocardiography, around 0.01%, with a slight male pre-
dominance [9]. Hurwitz and Roberts introduced a classi-
fication nomenclature for quadricuspid aortic valve that
included 7 different types named A to G [10]. The type
described in this case report, with three equal cusps and
one larger, is type E and extremely rare. Tutarel published
the most comprehensive review of the literature concern-
ing quadricuspid aortic valve, in 2004 [9]. This review
included 186 cases of quadricuspid aortic valve and only
4 was designated type E [9]. Aortic regurgitation is the pre-
dominant valvular dysfunction and is seen in up to 75%
of documented cases, on the other hand, quadricuspid
aortic valve is rarely associated with aortic stenosis [9].
The most prevalent other cardiac malformation associated
with quadricuspid aortic valve was anomalies of the coro-
nary arteries, which have been reported in 10% of the
cases [9]. Other malformation associated with quadricus-
pid aortic valve include stenosis of pulmonic valve, non-
obstructive cardiomyopathy, subaortic stenosis, and
ventricular septal defect [8,9]. Also, cases of bacterial
endocarditis affecting a quadricuspid aortic valve have
been reported [2,3].

The prevalence of quadricuspid aortic valve is too low to
study the diagnostic accuracy of transthoracic versus tran-
soesophageal echocardiography in the detection of this
malformation. A recent study by Alegret and co-workers
concerning bicuspid aortic valve morphology gives an
hint to what might be the case [11]. Alegret et al inves-
tigated 59 selected patients and 15 out of 32 bicuspid aortic
valves were missed on transthoracic, but detected on tran-
soesophageal echocardiography [11].

**Conclusion**

Noteworthy is that two consecutive transthoracic echocar-
diographic exams had fail to document the quadricuspid
aortic valve and this is one more example that liberal use
of transoesophageal echocardiography is often warranted if optimal display of valvular morphology is desired. Also, the increasing use of transoesophageal echocardiography will probably lead to an increase in the detection of quadricuspid aortic valve. It is therefore of importance to know this anomaly and its associated defects.

Competing interests
The author(s) declare that they have no competing interests.

Authors’ contributions
MD did the echocardiography of this case.

MD, MS jointly performed review of the literature and wrote the paper.

References
1. Cooke JC, Dupuche DR, Gay TJ: Quadricuspid Aortic Valve. Echocardiography 2000, 17:699.
2. Takeda N, Ohakaki E, Kashigawa H, Tobaru T, Sumiyoshi T: Infective endocarditis associated with quadricuspid aortic valve. Jpn Heart J 2003, 44:441-5.
3. Watanabe Y, Taketani Y, Takei Y, Tanaka K, Watanabe Y: Complete heart block resulting from quadricuspid aortic valve penicillin-resistant pneumococcal endocarditis: a case report. Circ J 2003, 67:275-6.
4. Hwang DM, Feindel CM, Butany JW: Quadricuspid semilunar valves: Report of 2 cases. Can J Cardiol 2003, 19:938-42.
5. Ninuma H, Yoshioka K, Ogino Y, Kawazoe K: Three-dimensional demonstration of tetracuspid aortic valve by 16-row multidetector-row computed tomography: comparison with transesophageal echocardiography. Eur J Cardiothorac Surg 2005, 28:326.
6. Recupero A, Pugliatti P, Rizzo F, Arrigo F, Coglitore S: Quadricuspid aortic valve: a rare cause of aortic insufficiency diagnosed by doppler echocardiography. Report of two cases and review of the literature. Ital Heart J 2005, 6:927-30.
7. Schulze MR, Srassser RH, Unser, Bi, Triv., and Quadricuspid Aortic Valves. N Engl J Med 2006, 355(12):1262.
8. Timperley J, Milner R, Marshall JA, Gilbert TJ: Quadricuspid aortic valves. A review. Clin Cardiol 2002, 25:548-52.
9. Tutarel O: The quadricuspid aortic valve: A comprehensive review. J Heart Valve Dis 2004, 13:534-37.
10. Hurwitz LC, Roberts WC: Quadricuspid semilunar valve. Am J Cardiol 1973, 31:623-6.
11. Alegret JM, Palazon O, Duran I, Vernis JM: Aortic valve morphology definition with transthoracic combined with transesophageal echocardiography in a population with high prevalence of bicuspid aortic valve. Int J Cardiovasc Imaging 2005, 21:213-7.