Case Report of a Pentacuspid Aortic Valve with Aortic Insufficiency.

Dr Majed Albogmi, Dr Ahmed Alharthi, Dr Mohammad Alharbi, Dr. Mutaz Alareef, Dr Maha Aldohan

Security Forces Hospital Program – Riyadh King Abdullah bin Abdulaziz University Hospital

DOI: https://doi.org/10.56293/IJASR.2023.5604

IJASR 2023
VOLUME 6
ISSUE 6 NOVEMBER – DECEMBER **ISSN: 2581-7876**

Abstract:

Background: Pentacuspid aortic valve is a rare congenital anomaly associated with symptoms ranging from chest discomfort to palpitations, audible murmur, exertional and paroxysmal nocturnal dyspnea. Progressively worsening exertional dizziness, as seen in the following case, was not recorded in published literature.

Case Summary: A 54-year-old male presented with a history of progressive exertional dizziness associated with chest pain and dyspnea. His electrocardiogram (ECG) showed signs of left ventricular hypertrophy and the transoesophageal echocardiography revealed a pentacuspid aortic valve with moderately severe aortic insufficiency, non-coaptation during diastole, and it confirmed the presence of left ventricular hypertrophy. The patient also underwent coronary angiography which revealed normal coronary arteries. The case was managed with an aortic valve replacement surgery using a supra annular mechanical valve.

Discussion: Transoesophageal echocardiography plays an important role in diagnosing valvular abnormalities or other structural heart defects. As seen in this case, the patient's ECG showed signs of left ventricular hypertrophy, which required the use of other imaging modalities to confirm and determine the underlying cause.

Keywords: case report, pentacuspid aortic valve, aortic insufficiency, aortic regurgitation, congenital heart disease

INTRODUCTION

Congenital heart defects or diseases (CHD) are defects with the heart's structure that are present at birth[1]. The worldwide incidence of CHD is estimated to be between 8 to 12 per 1,000 live births[2]. A case of a 54-year-old male with a pentacuspid aortic valve and moderately severe aortic insufficiency detected by transesophageal echocardiography, is presented.

CASE PRESENTATION

A 54-year-old male with no known history of any chronic disease presented with a 4-month history of progressive exertional dizziness that has recently been associated with chest pain and dyspnea. Nevertheless, the patient had no syncope, palpitations, orthopnea, or paroxysmal nocturnal dyspnea. His blood pressure was 131/80 mm Hg and his heart rate was 80 beats per minute. Lung auscultation revealed normal vesicular breathing with equal air entry on both sides and no added sounds. Cardiac examination revealed a normal S1 and S2 with a grade III end diastolic murmur heard at the location of the aortic valve area. Furthermore, the patient had normal routine blood and urine analyses, except for an elevated fasting blood glucose at 6.25 mmol/L. However, his electrocardiogram (ECG) revealed a normal sinus rhythm and an increased R wave amplitude in praecordial leads V3-V6, which is consistent with left ventricular hypertrophy.

Transoesophageal echocardiography (TOE) was performed and showed a pentacuspid aortic valve (PAV), as seen in figure 1, with moderately severe aortic insufficiency and non-coaptation during diastole. It also revealed left ventricular hypertrophy with a mass of 312 g, mild diastolic dysfunction, and an ejection fraction of 55%. Additional TOE findings are listed in table 1. The patient also underwent coronary angiography which revealed normal coronary arteries.

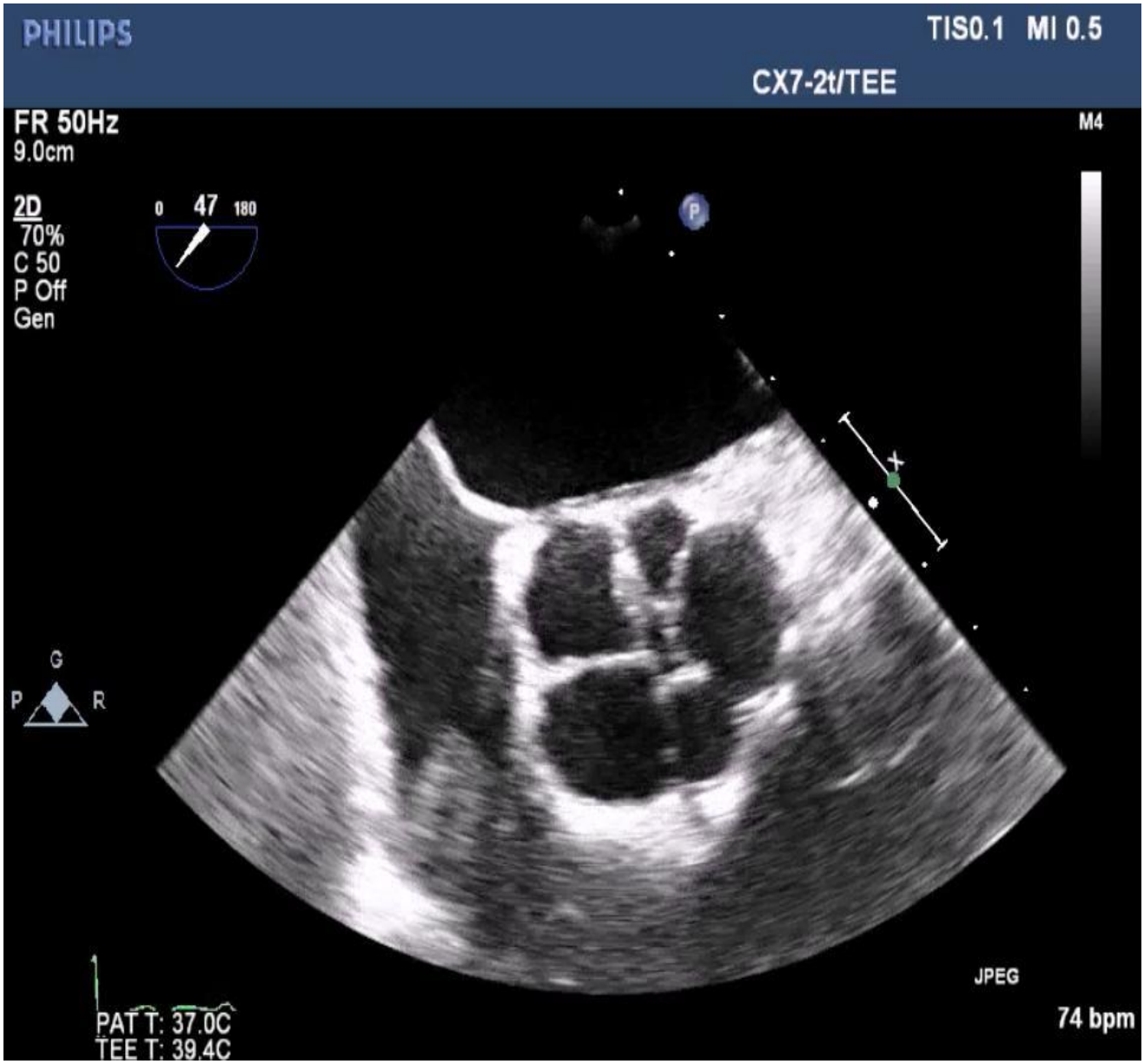
The patient had aortic valve replacement surgery using a 24 mm ATS AP360 supra annular mechanical valve. He recovered without complications, except for a fever on the fifth day post-operatively which later subsided. The patient was discharged on the sixth day post-operatively on aspirin, warfarin, bisoprolol (beta-blocker), furosemide (loop diuretic), amplodipine (calcium channel blocker), perindopril (angiotensin converting enzyme [ACE] inhibitor), and paracetamol.

Table 1 – Transoesophageal Echocardiography Findings and Aortic Valve Insufficiency Quantification

Jet width to left ventricular outflow tract ratio	50 %
Pressure half time*	400 ms
Vena contracta	9.8 mm
Effective regurgitant orifice	0.186 cm^2

*not accurate due to coexistent diastolic dysfunction

Figure 1 – Transoesophageal Echocardiography Image of the Pentacuspid Aortic Valve



DISCUSSION

The aortic valve is a tricuspid semilunar valve derived from subendocardial valve tissue and bulbar ridges. Congenital anomalies of the aortic valve involve the number of cusps; hence, it can be unicuspid, bicuspid, quadricuspid and pentacuspid. The bicuspid aortic valve anomaly is common with a prevalence of approximately 1.3% worldwide[3], while pentacuspid aortic valve is rarely seen and reported in clinical practice; with the first case to our knowledge in 1982[4].

Transoesophageal echocardiography is the preferred diagnostic approach for PAV or other valvular abnormalities, but other diagnostic modalities such as a three dimensional transoesophageal echocardiography, a multidetector computed tomography (CT), or virtual intraaortic endoscopy by multidetector-row CT may be confirmatory.

Case reports of PAV describe patients between the age of 19 and 48. Patients symptoms ranged from presenting with palpitations, chest discomfort, audible murmur to exertional and paroxysmal nocturnal dyspnea[5-9]. However, the main complaint of progressively worsening exertional dizziness, as seen in this case, was not found in published literature. Most reported cases discovered a variation between the sizes of the cusps, although the presence or absence of haemodynamic changes or aortic insufficiency was unclear. Differences in cusp size may be attributed to dysplastic changes associated with PAV[9]. Aortic insufficiency was due to failure of the five cusps to coapt.

This case highlights the importance of transoesophageal echocardiography in diagnosing valvular abnormalities. Since the patient's ECG showed signs of left ventricular hypertrophy, it resulted in the use of other imaging modalities to determine the underlying cause.

PATIENT CONSENT

Written consent for submission and publication of this case report including images and associated text has been obtained from the patient.

FUNDING

There was no funding for this study.

ACKNOWLEDGEMENTS

Personal acknowledgements should precede those of institutions.

REFERENCES

1. National Heart, Lung, and Blood Institute. Congenital Heart Defects. Bethesda, MD: NHLHBI. Available from: https://www.nhlbi.nih.gov/health-topics/congenital-heart-defects/ (29 May 2018).
2. Qu Y, Liu X, Zhuang J, Chen G, Mai J, Guo X, Ou Y, Chen J, Gong W, Gao X, Wu Y, Nie Z. Incidence of Congenital Heart Disease: The 9-Year Experience of the Guangdong Registry of Congenital Heart Disease, China. PLoS One. 2016;11(7): 0159257.
3. Verma S, Siu S. Aortic Dilatation in Patients with Bicuspid Aortic Valve. N Engl J Med. 2014;370(20):1920-1929.
4. Bogers J, Aytug Z, Hendriks F, Huysmans H. Quinticuspid aortic valve causing aortic valve incompetence and stenosis. Thorax. 1982;37(7):542-543.
5. Kuroki H, Hirooka K, Ohnuki M. Pentacuspid aortic valve causing severe aortic regurgitation. The J Thorac Cardiovasc Surg. 2012;143(2):e11-e12.
6. Cemri M. Pentacuspid aortic valve diagnosed by transoesophageal echocardiography. Heart. 2000;84(4):9e-9.
7. Wang S, Meng X, Zhang H, Wang J, Han J. Pentacuspid Aortic Valve With Severe Aortic Regurgitation. Ann Thorac Surg. 2010;89(6):2034-2036.
8. Kamata S, Sakagoshi N, Ohata T, Sasako Y. Successful surgical treatment of pentacuspid aortic valve with severe aortic regurgitation. Jpn J Cardiovasc Surg. 2008; 37: 53–55

9. Ozyilmaz S, Akgul O, Guzeltas A, Ozyilmaz I. Diagnosis of Pentacuspid Aortic Valve with Severe Regurgitation Using Three-Dimensional Transesophageal Echocardiography. Echocardiography. 2014;32(2):393-394.