

CASE REPORT

A case of isolated bicuspid pulmonary valve

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Summary

Our case highlights the finding of an abnormal pulmonary valve on 2D echocardiography, confirmed to be of bicuspid morphology with 3D imaging. The use of biplane imaging both in transthoracic and transoesophageal echocardiography and routine use of three-dimensional views particularly in transoesophageal echocardiography are of incremental value in better delineating pulmonary valve anatomy.

Key Words

- ▶ 2D echocardiography
- ▶ 2D transoesophageal echocardiography
- ▶ 3D transoesophageal echocardiography

Learning points:

- Bicuspid pulmonary valve as an isolated clinical entity is a rare finding in clinical practice with an incidence of about 0.1%.
- The true prevalence of the condition may be underestimated because of difficulty in visualising the pulmonary valve en-face on standard two-dimensional echocardiography.
- Trans-oesophageal echocardiography may provide better visualization of the pulmonary valve when transthoracic images are affected by interference from the left lung.
- Routine use of 3D echocardiography with biplane and zoomed views should be advocated for a full morphological assessment of the pulmonary valve, whether imaging via the transthoracic or transoesophageal approach.

Background

Bicuspid pulmonary valve is considered a rare congenital anomaly. Although standard 2D trans-thoracic echocardiography can readily detect pulmonary valve dysfunction, morphological assessment may be limited by the close proximity of the pulmonary valve to the lungs and the inability to view the pulmonary valve en face. Our case report suggests that the finding of an abnormal pulmonary valve on standard transthoracic echocardiography should warrant further interrogation with 3D imaging, ideally with a trans-oesophageal approach. This allows a full morphological assessment of the pulmonary valve increasing the chances of detecting a bicuspid anatomy, which may otherwise be missed.

Case presentation

A 73-year-old female presented with a history of nausea, vomiting, abdominal discomfort and fever. She had a past history of breast cancer treated 9 years ago. Her blood pressure was 125/55 with heart rate of 100 beats/min, and no clinical evidence of fluid overload. On auscultation, there was an early diastolic murmur heard best in the left parasternal area, but with no peripheral stigmata of infective endocarditis.

Investigations

Investigations revealed a raised white cell count of 27,500/cu.mm with a neutrophilia. Haemoglobin was

reduced at 81g/L with a normocytic, normochromic pattern consistent with anaemia of a chronic disorder. Platelets were normal and CRP was elevated at 350mg/L. Renal and liver function tests were normal apart from a low albumin of 18g/L; serum calcium was 2.78mmol/L and phosphate was 0.97mmol/L. Chest X-ray showed hilar prominence and CT scan of the thorax also showed a dilated pulmonary trunk and pleural effusion. CT scan of the abdomen showed a heterogeneous pelvic mass with associated calcific foci. CT pulmonary angiography excluded pulmonary embolism, and serial blood cultures were negative. Notably, she had already been empirically treated with antibiotics. Biopsy of the pelvic mass revealed poorly differentiated spindle cell stromal cancer possibly endometrial in origin.

Transthoracic echocardiography (TTE) was performed to look for any evidence of valve infection in view of the diastolic murmur and raised inflammatory markers. Left ventricular size and systolic function were normal. The right ventricle was mildly dilated with normal systolic function. Mitral valve was mildly thickened with mild mitral regurgitation. Tricuspid valve was thin with mild tricuspid regurgitation. The aortic valve was thickened with normal excursion of all coronary cusps and trivial regurgitation. The most notable finding was moderate pulmonary regurgitation (Figs 1 and 2) with thickening and doming of the pulmonary valve (Fig. 3). The peak pulmonary valve gradient was 19mmHg and the pulmonary artery was mildly dilated. The mean pulmonary artery pressure was approximately 34mmHg. A transoesophageal echocardiogram was performed to further evaluate the pulmonary valve and exclude any vegetations. The use of 3D imaging showed a bicuspid pulmonary valve with anterior and posterior commissures (Figs 4 and 5) and moderate regurgitation. There was no evidence of vegetations associated with any of the cardiac valves.

Treatment and outcomes

The patient was managed palliatively for the underlying malignancy.

Discussion

The isolated finding of a bicuspid pulmonary valve is rare and most often diagnosed at surgery or post-mortem. In a European homograft bank registry from unselected

donors, the incidence was found to be 0.1% among 3861 donors (1), whereas the incidence of bicuspid aortic valve is 1–2%. The true incidence of bicuspid pulmonary valve could be underestimated owing to difficulty in visualising the pulmonary valve on conventional two-dimensional echocardiography. Improper fusion or dedifferentiation of the endocardial cushion is responsible for abnormal semilunar valves and so a bicuspid pulmonary valve is more often associated with other congenital abnormalities such as tetralogy of Fallot or ventricular septal defect (2). As most patients with bicuspid pulmonary valves are asymptomatic, the diagnosis is often made incidentally after suspicious auscultatory findings and subsequent imaging. Trans-thoracic echocardiography remains the first-line investigation for outlining pulmonary valve morphology. The pulmonary valve is usually visualised in the longitudinal view on conventional 2D TTE conferring an inherent difficulty in delineating the number of cusps. Doming of the valve seen in our case was a crucial finding and should raise the suspicion of an abnormal semilunar valve when observed in the pulmonary or aortic position. Trans-oesophageal echocardiography (TOE) may provide better spatial resolution for imaging the pulmonary valve. Probe manipulation to provide modified 2D views, complemented with the 3D X-plane and zoom views can delineate the pulmonary valve in a short axis slice to better assess valve morphology. Use of the latter 3D modalities with TTE may also provide similar diagnostic information, but imaging difficulty may arise from the close proximity of the pulmonary valve to the left lung.

Other cardiac imaging modalities such as CT and magnetic resonance imaging have contributed greatly in outlining complex cardiac anatomy and consequently increasing cases of isolated bicuspid pulmonary valves have been reported with these imaging tools. Quadricuspid pulmonary valves have also been described with the increasing use of cardiac CT imaging. It is difficult to know the true prevalence of quadricuspid pulmonary valves as the data come from autopsy studies and incidental case reports, but this may occur as an isolated finding or in association with other congenital cardiac anomalies, such as bicuspid or quadricuspid aortic valve (3). Quadricuspid pulmonary valves have been described more commonly in men and tend to remain clinically silent but may lead to isolated severe pulmonary regurgitation or concomitant stenosis and regurgitation.

Multi-detector ECG-gated cardiac CT provides good 3D spatial orientation, and therefore, enables detailed evaluation of small cardiac structures, which may be less amenable to echocardiography. Although the technique

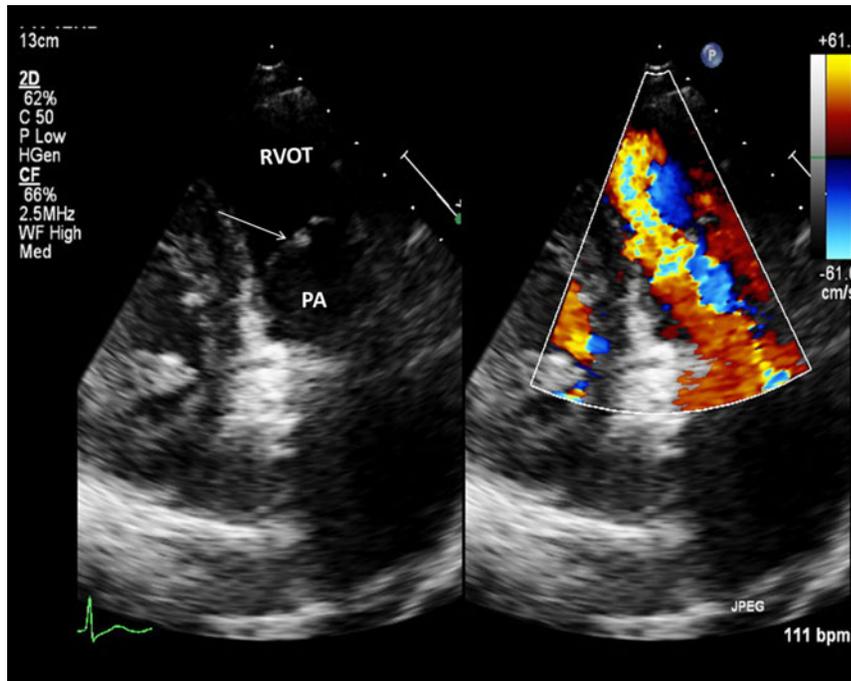


Figure 1
Simultaneous two-dimensional (left) and colour flow Doppler (right) images of the pulmonary valve (arrow) in early diastole showing a jet of pulmonary regurgitation.

involves exposure to radiation, acquisition times are rapid. Cardiac MR is the reference standard for the assessment of chamber volumes, myocardial mass and tissue characterisation, but has a lesser role in the assessment of cardiac valves. The technique does not involve radiation, but metallic implants such as pacemakers tend to be contraindicated. The use of multiple imaging modalities depends on the clinical context, and each modality

should be considered complementary in the diagnosis of complex cardiac conditions. Nevertheless, the ready availability of 3D echocardiography, may limit the use of the latter imaging techniques in the future. Without the benefit of 3D echocardiography, bicuspid pulmonary valves are likely to have been underdiagnosed in the past. The use of 3D echocardiography may give an artificially thicker appearance to thin structures making cusp

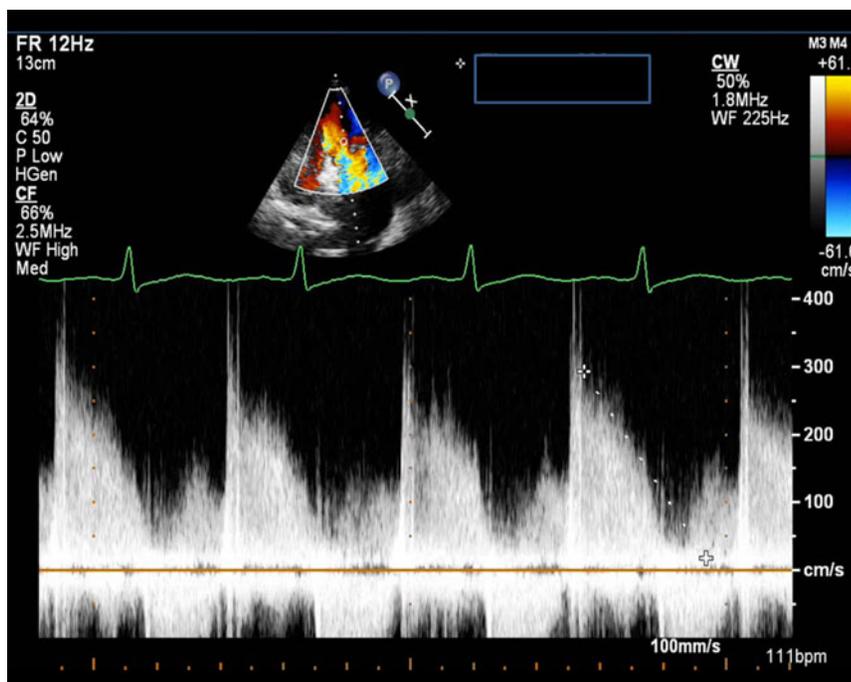


Figure 2
Continuous wave Doppler profile of pulmonary regurgitation.

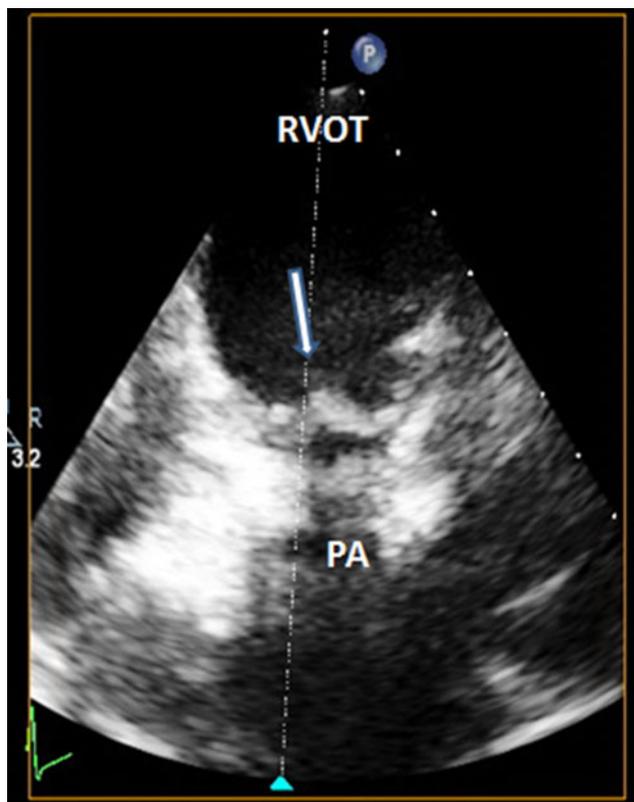


Figure 3
Two-dimensional parasternal view of the pulmonary valve showing marked thickening and systolic doming of the leaflet (arrow). PA, pulmonary artery; RVOT, right ventricular outflow tract.

morphology easier to demonstrate than with 2D imaging. The true prevalence of the condition may be uncovered by the routine use of 3D echocardiography for pulmonary valve assessment (4).

The clinical course of isolated bicuspid pulmonary valve is usually benign and does not need any therapeutic intervention unless associated with haemodynamically significant pulmonary stenosis or regurgitation (5). Infective endocarditis is one of the most important diagnoses to be considered in patients presenting with fever and a new cardiac murmur. Isolated infective endocarditis of the pulmonary valve is very rare. In a study of 155 cases of infective endocarditis encountered over a period of 14 years, only 9 cases of pulmonary valve endocarditis have been identified. Autopsy has been performed in all 9 cases, of whom 3 were found to have bicuspid pulmonary valves (6). Two of these cases had associated congenital heart defects and one had an isolated bicuspid pulmonary valve. In our patient, echocardiography was performed to exclude infective endocarditis and a bicuspid pulmonary valve was identified, but with no clear evidence of

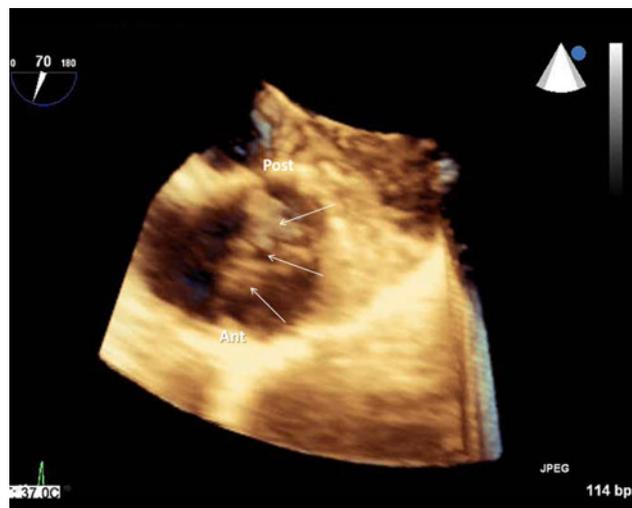


Figure 4
Three-dimensional view of the bicuspid pulmonary valve seen en-face and showing marked thickening and malcoaptation of the leaflets (arrows). Ant, anterior commissure; Post, posterior commissure.

a vegetation or microbiological confirmation of endocarditis. Trans-oesophageal echocardiography tends to be more sensitive than TTE as spatial resolution is improved with TOE. As with this case, TOE should be used in those with a high index of suspicion and a negative trans-thoracic echocardiogram.

The incidence of infective endocarditis varies globally depending on the prevalence of underlying cardiac disorders. In developed countries, predispositions to infective endocarditis such as degenerative valve disease, prosthetic valves and cardiac devices tend to be more common, whereas rheumatic carditis tends to predominate in other geographical regions. In North America, the incidence of infective endocarditis approximates to 3–10/100,000 population and despite advances in diagnosis and treatment, the mortality rate has remained unchanged at about 25–30% (7). Early diagnosis of infective endocarditis is vital for improving the clinical outcome of the individual patient. Patients with poor oral hygiene, intravenous drug abuse, long-term indwelling intravenous lines, inflammatory conditions particularly malignancy, immune compromised state, on haemodialysis and diabetes are at a higher risk of infective endocarditis. High-risk pre-existing cardiac conditions include complex congenital heart disease, prosthetic valves, intracardiac devices and previous endocarditis.

Good oral hygiene is of paramount importance in preventing infective endocarditis in general and particularly among individuals with pre-existing cardiac disorders. Commercially available oral mouthwashes

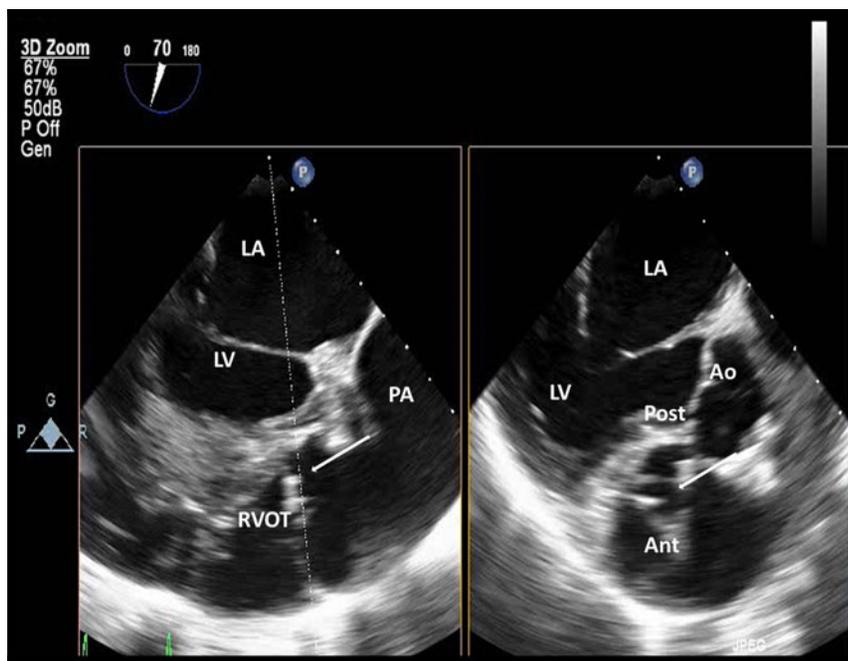


Figure 5
Trans-oesophageal echocardiogram showing X-plane-derived orthogonal views of the pulmonary valve leaflets (arrows). The image on the left shows the pulmonary valve in the long-axis view and on the right shows the bicuspid morphology in a modified short axis view. Ao, aortic root; LA, left atrium; LV, left ventricle; PA, pulmonary artery; Post, posterior commissure and Ant, anterior commissure of the pulmonary valve; RVOT, right ventricular outflow tract.

are able to destroy the common oral pathogens in planktonic solution, and so may be helpful in preventing endocarditis in susceptible individuals (8). Cancer is an inflammatory condition, which is commonly associated with a hypercoagulable state. It has been estimated that around 15% of cancer sufferers will develop a thrombophilic event, which can rarely manifest as a non-bacterial thrombotic endocarditis, referred to as marantic endocarditis (9). The latter should be considered a possibility when a presumed vegetation has been identified in cancer patients. The pathology of marantic endocarditis relates to a hypercoagulable state resulting in aggregates of platelets and fibrin giving rise to the appearance of vegetation, which is in fact sterile in nature.

In a large study over a period of 6 years, 5.6% of patients with infective endocarditis had a prior history or new diagnosis of cancer, and this was mainly as a result of medical interventions. In individuals presenting with infective endocarditis, hidden cancer particularly of the gastrointestinal or urinary tract should be suspected if the relevant organism has been isolated (10).

The Ross procedure is commonly performed in young adults and in the paediatric population for severe aortic valve disease. This involves implanting the individual's own pulmonary valve (autograft) in the aortic position and placing a bioprosthetic valve in the pulmonary position. The longevity of the autograft in the aortic position is greater than a bioprosthetic valve. The presence of a bicuspid pulmonary valve is an uncommon,

relative contra-indication for the Ross procedure because of the lack of durability of this type of valve in the aortic position.

In summary, our case highlights the finding of an abnormal pulmonary valve on 2D echocardiography, confirmed to be of bicuspid morphology with 3D imaging. The routine use of biplane and 3D imaging in both transthoracic and transoesophageal echocardiography has great value in delineating the anatomy of the pulmonary valve.

Declaration of interest

The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this case report.

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Patient consent

Written permission was posthumously obtained from the patient's spouse.

Author contribution statement

Dr Ramasamy Manivarmane collected the clinical details, performed the literature search and drafted the article. Dr Rebecca Taylor contributed to the collection of data. Dr Rajdeep Khattar gave academic cardiological

advice and edited the article. Permission has been sought from the clinician in charge of the patient.

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