

# Multimodality imaging of a rare pulmonary artery sessile mass: a case report

Laura Piscitelli <sup>1\*</sup>, Ilaria Dentamaro<sup>2</sup>, Gaetano Pezzicoli <sup>3</sup>,  
and Carlo D'Agostino<sup>4</sup>

<sup>1</sup>Cardiology Unit, Department of Emergency and Organ Transplant, University Hospital Policlinico of Bari, Piazza Giulio Cesare, 11 70124 (BARI), Italy; <sup>2</sup>Cardiology Unit, Miulli Hospital, Strada provinciale 127 Acquaviva-Santeramo, 70021 Acquaviva delle Fonti (BARI), Italy; <sup>3</sup>Department of Biomedical Sciences and Human Oncology, University Hospital Policlinico of Bari, Piazza Giulio Cesare, 11 70124 (BARI), Italy; and <sup>4</sup>Department of Cardiology, University Hospital Policlinico of Bari, Piazza Giulio Cesare, 11 70124 (BARI), Italy

Received 15 June 2020; first decision 29 July 2020; accepted 8 December 2020

## Background

Primary pulmonary artery masses are unusual entities that mimic pulmonary embolism (PE) in clinical presentation and on imaging studies. It is necessary to perform advanced diagnostic exams, such as transesophageal echocardiography (TEE) and cardiac magnetic resonance imaging (MRI), to determine the proper diagnosis. In unclear cases, laboratory findings, morphological follow-up, and response to anticoagulant therapy can help to clarify the diagnosis.

## Case summary

A 47-year-old previously healthy man with worsening effort dyspnoea underwent chest computed tomography (CT) for suspicion of PE, which showed a pedunculated eccentric mass at the origin of the pulmonary artery causing severe stenosis. The patient was started on anticoagulation therapy, but, after TEE and cardiac MRI, a neoplastic fibroelastic mass was suspected. Unexpectedly, <sup>18</sup>fluorodeoxyglucose (FDG) positron emission tomography (PET)-CT revealed a unique area of glucose uptake in the superior lobe of the left lung and not in the pulmonary artery. The biopsy was consistent with pleomorphic high-grade lung sarcoma. After 3 months of chemotherapy, a CT scan showed progression of the lung disease with no change in the arterial mass, which was therefore confirmed as pulmonary fibroelastoma.

## Discussion

Due to the rarity of pulmonary artery tumours, they can be initially misdiagnosed as PE or a metastasis of a lung sarcoma. Three-dimensional TEE and cardiac MRI are particularly useful in differentiating tumours from PE.

## Keywords

Pulmonary artery • Thromboembolism • Mass • Case report • Multimodality imaging

## Learning points

- The diagnosis of neoplastic masses should be considered in patients with suspicion of pulmonary embolism, especially those with poor response to anticoagulant therapy.<sup>1,2</sup>
- Thrombus and mass are difficult to differentiate. Three-dimensional transesophageal echocardiography and cardiac magnetic resonance imaging are helpful for the complete morphological and functional evaluation of cardiac masses.<sup>3</sup>

\* Corresponding author. Tel: +393495438622, Email: [piscitellilaura92@gmail.com](mailto:piscitellilaura92@gmail.com)

Handling Editor: Inga Voges

Peer-reviewers: Dmitry Duplyakov and Ciro Santoro

Compliance Editor: Hibba Kurdi

Supplementary Material Editor: Elhosseyn Guella

© The Author(s) 2021. Published by Oxford University Press on behalf of the European Society of Cardiology.

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact [journals.permissions@oup.com](mailto:journals.permissions@oup.com)

## Introduction

Primary cardiac masses are rare entities that can be easily mistaken for thromboembolic events, with serious prognostic implications. We report a case in which a pulmonary artery mass and its clinical presentation played a pivotal role in diagnosing a high-grade pulmonary sarcoma that was clinically silent.

## Timeline

Day 0	The patient presented with sudden dyspnoea. Computed tomography (CT): pulmonary artery obstruction, probably due to embolism. An oral anticoagulant therapy was started. Electrocardiogram (ECG): right axial deviation. High T waves.
Week 2	Transesophageal echocardiography: 17 × 13 mm sessile mass at the right ventricle outflow tract. Magnetic resonance imaging (MRI): 13 × 53 mm fibrous vegetation, extended to pulmonary bifurcation.
Week 3	<sup>18</sup> F-FDG PET-CT: area of glucose uptake at the superior lobe of the left lung, no uptake in the heart.
Month 2	New admission for: control CT and US-guided biopsy of the pulmonary mass that revealed a pleomorphic high-grade sarcoma.
Month 3	Admission in oncology with new cardiac MRI. Starting of 1st line chemotherapy: doxorubicin-ifosfamide.
Month 7 (after 3 cycles)	Follow-up CT: increase in the size of the lung sarcoma, with no change of the pulmonary artery mass.
Month 7	Starting of the 2nd line chemotherapy: gemcitabine-paclitaxel.
Month 12	Patient's death

## Case presentation

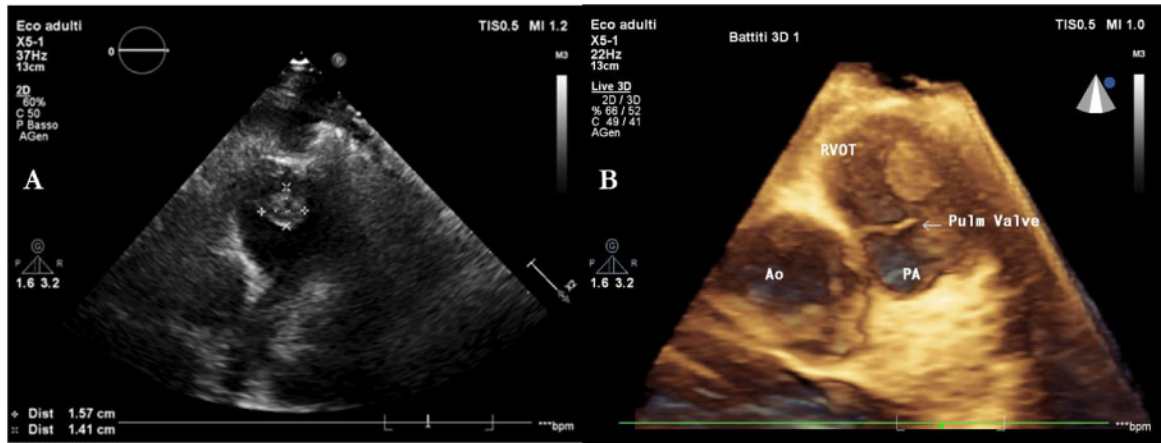
We report the case of a 47-year-old white male smoker with a family history of lung cancer. No major comorbidities were reported. Initially, he presented with sudden dyspnoea on exertion, without any other symptoms such as fever, weight loss, or night sweats (B symptoms in lymphoma clinical presentation). The haemodynamic parameters were stable (BP 140/90 mmHg, HR 70 b.p.m.) and the physical examination was normal. Electrocardiogram (ECG) showed right axial deviation and high T waves in precordial leads. He underwent chest computed tomography (CT) for suspicion of pulmonary embolism (PE). Computed tomography imaging showed a filling defect ~22 mm in diameter at the origin of the pulmonary artery causing severe stenosis. Although the patient was started on anticoagulant therapy, thoracic echocardiogram (TTE) and transesophageal

echocardiography (TEE) confirmed a fixed 17 × 13 mm sessile mass at the right ventricular outflow tract (RVOT), just before the pulmonary valve, and severe stenosis of the pulmonary artery main trunk, with a maximum gradient of 100 mmHg and a mean gradient of 68 mmHg (n.v. maximum gradient <36 mmHg).

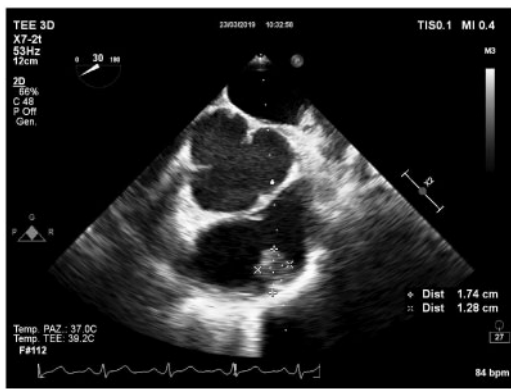
The right ventricle was mildly dilated, hypertrophic, and showed a mild decrease in contractile function (Figures 1 and 2, Videos 1–3). To investigate the nature of the lesion, the patient underwent cardiac magnetic resonance imaging (MRI), which characterized the sessile mass as a 13 × 53 mm fibrous vegetation next to the anterior leaflet of the pulmonary valve that extended to the pulmonary bifurcation (Figure 3). For the high suspicion of a neoplastic origin, the patient underwent <sup>18</sup>fluorodeoxyglucose (<sup>18</sup>FDG) positron emission tomography (PET)-CT. Unexpectedly, PET showed an area of intense glucose uptake at the superior lobe of the left lung, in the subpleural region. The patient was re-admitted with a second CT that showed persistence of the pulmonary artery mass despite anticoagulant therapy with warfarin (2 months) and no peripheral filling defects of the pulmonary arteries. The CT also revealed a 7 cm-wide solid neofor- mation at the superior lobe of the left lung. An ultrasonography (US)-guided biopsy of the lung parenchymal area was therefore performed. The pathology report was consistent with a pleomorphic high-grade sarcoma. A team including an US specialist, a cardiovascular surgeon, an oncologist, and a pathologist was assembled to decide the best therapeutic management. No surgical approach was possible, so the patient was started on first-line chemotherapy including doxorubicin (25 mg/m<sup>2</sup> per day, Days 1–3) plus ifosfamide (10 g/m<sup>2</sup> over 4 days), with mesna (0.5 g/m<sup>2</sup> by intravenous bolus before ifosfamide, 1.5 g/m<sup>2</sup> concurrent with ifosfamide, and 1 g/m<sup>2</sup> orally 2 and 6 h after completion of ifosfamide infusion) and pegfilgrastim (6 mg subcutaneously, Day 5) as support, in 3-week cycles, up to a maximum of six cycles.<sup>4</sup> After 3 months, the clinical condition of the patient worsened, with chest pain and rest dyspnoea occurrence. The restaging CT and MRI showed no change in the size of the pulmonary artery mass (Figures 4 and 5), but an increase in the size of the pulmonary mass, which now reached a diameter of 18 cm (Figure 5). Moreover, a new lesion was observed in the inferior lobe of the right lung. According to response evaluation criteria in solid tumours (RECIST), the patient was considered to have progressive disease, and therefore, a second line of chemotherapy was started, consisting of gemcitabine (900 mg/m<sup>2</sup> on Days 1 and 8) and docetaxel (70 mg/m<sup>2</sup> on Day 8) in 3-week cycles.<sup>5</sup> After 5 months, the patient died.

## Discussion

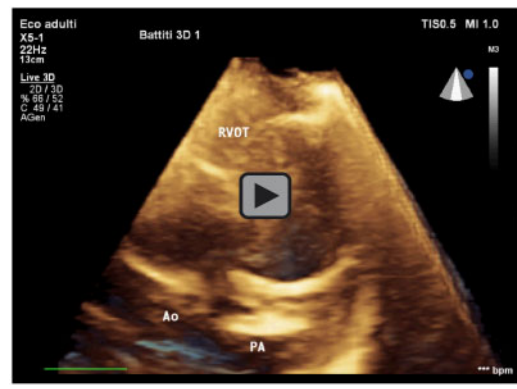
This case is interesting, first, because of the diagnostic path: the differential diagnosis of PE and cardiac masses remains a challenge. And second, because multimodality advanced imaging techniques such as 3D TEE and cardiac MRI play an important role in characterizing the precise location of the pulmonary artery tumour (Supplementary material online, Table).<sup>3</sup> A multidisciplinary team including cardiologists, oncologists, radiologists, and pathologists is essential to determine the most probable diagnosis and choose the optimal therapy. The unchanged persistence of the pulmonary artery mass together with the absence of glucose uptake, in contrast to the lung sarcoma, allow us to diagnose its benign nature with unlikely relationship



**Figure 1** (A) Two-dimensional transthoracic echocardiogram, pulmonary off-axis view. It shows right ventricular outflow tract, pulmonary valve, and pulmonary artery in longitudinal axis. Right ventricular outflow tract is mildly dilated with a fixed 15 × 14 rounded isoechogenic mass inside. (B) Three-dimensional transthoracic echocardiogram, PSAX focused on large vessels (aorta and pulmonary artery). Three-dimensional technique gives a volumetric characterization of the sessile mass located in the right ventricular outflow tract eccentrically, just before the pulmonary valve.



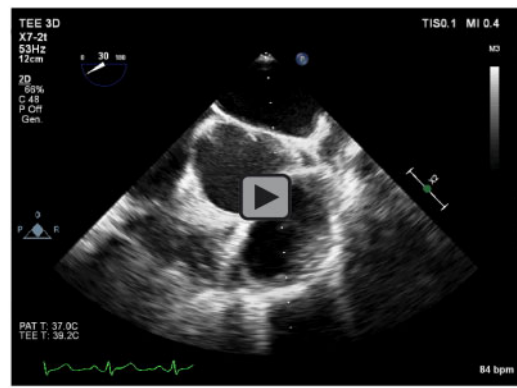
**Figure 2** Two-dimensional transesophageal echocardiography 30 degrees short axis at great vessels level. It shows the aortic valve in cross-section and, just below, a 17 × 13 mm sessile mass at the right ventricular outflow tract (RVOT).



**Video 2** Three-dimensional transthoracic echocardiogram shows the mass at the right ventricular outflow tract causing an important flow acceleration indicative of severe obstruction.

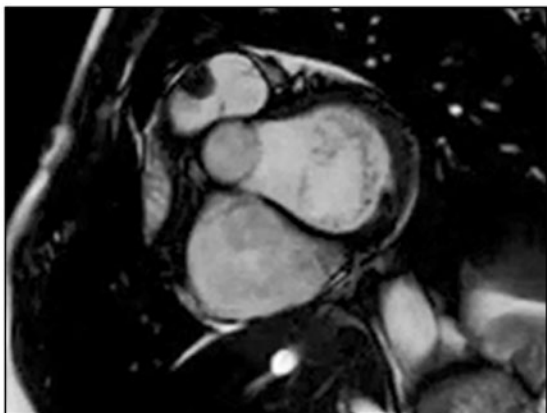


**Video 1** Two-dimensional transthoracic echocardiogram, pulmonary off-axis view shows a dilated right ventricle and the rounded sessile mass inside.



**Video 3** Two-dimensional transesophageal echocardiography 30 degrees short axis at great vessels level. It shows, below the aortic valve, a sessile mass at the right ventricular outflow tract (RVOT), just before the pulmonary valve.

between this and the primary lung masses identified ([Supplementary material online, Table](#)). Given its location, shape, and structure, the most likely differential diagnosis is a sessile fibroelastoma, a benign cardiac neoplasia that arises from the endocardium and that can be dangerous due to the risk of peripheral embolization.<sup>6</sup> Other plausible hypotheses include a fibroelastic reaction to the lung neoplasia (paraneoplastic syndrome) or, less likely, a fibrotic organized thromboembolic lesion caused by the lung neoplasia. This latter possibility was excluded because of the low mass mobility and the



**Figure 3** Cardiac magnetic resonance imaging, March 2019: Segmented steady-state free precession cine imaging sequences in short-axis view. We can see a small and round mass, with a homogenous hypointense signal intensity, attached to the right ventricular outflow tract by a short stalk.

absence of modification despite intensive anticoagulant therapy ([Supplementary material online, Table](#)). Determining the true nature of the mass is complex: a biopsy with a histological diagnosis would be required to definitively diagnose the mass. However, in this case, that would have been too dangerous for the patient and would likely not have changed the therapeutic strategy, which was largely determined by the lung tumour.

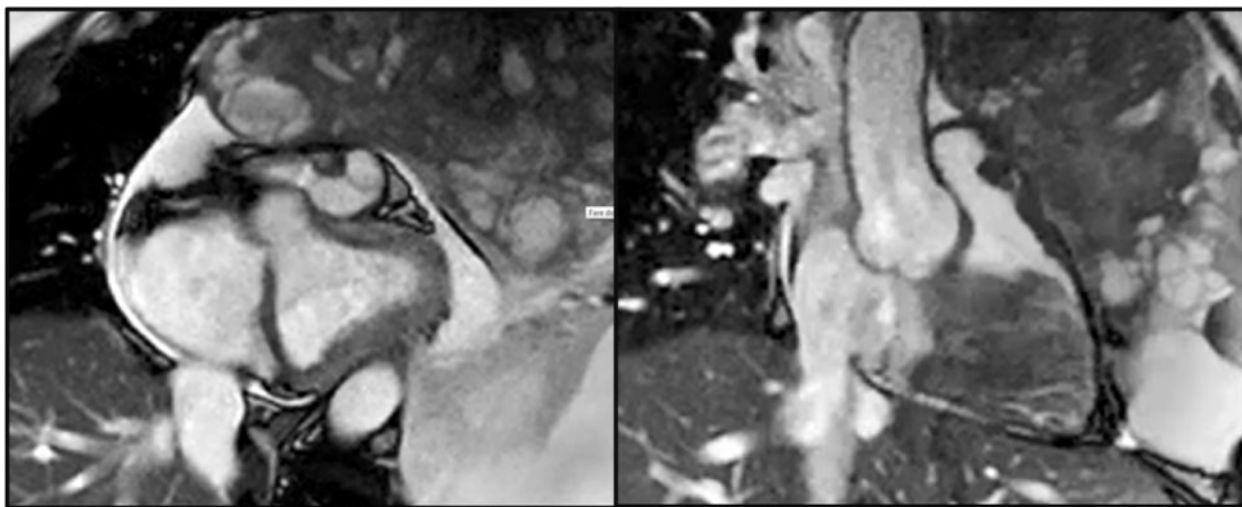
## Conclusion

Because of the low incidence of pulmonary artery tumours, together with their clinical presentation with dyspnoea, they can be easily mistaken for a thromboembolic process. Thanks to multimodality cardiac imaging (TEE and MRI), it was possible to make a difficult diagnosis.<sup>4</sup> Additionally, having an interdisciplinary team was fundamental for us to obtain a more complete overview of this rare condition and determine the best management.

## Lead author biography

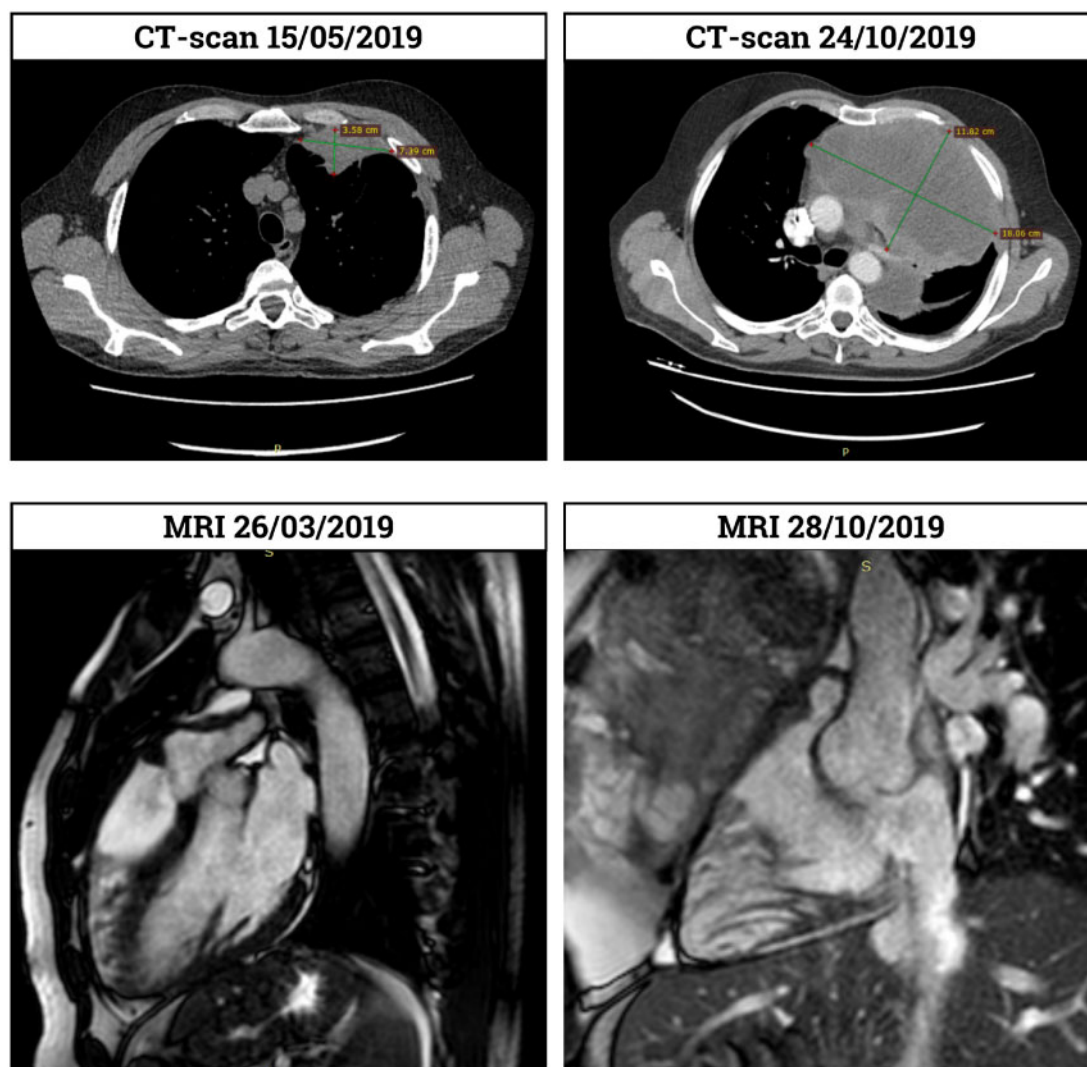


Dr Laura Piscitelli is actually a Cardiology resident in the Cardiology Department of University Hospital Policlinico of Bari, in Italy. She graduated in the University of Bari in 2017. She has a knee interest in Cardiac Imaging, in particular advanced echocardiography applied in valvular heart disease.



**Figure 4** Cardiac magnetic resonance imaging, October 2019: Cine imaging sequences in short-axis and oblique coronal views showing the mass with the same signal intensity and no change in its size. However, a large pericardial effusion appears.





**Figure 5** Computed tomography and magnetic resonance imaging showing an increase of the lung disease with no change in the size of the cardiac mass.

## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

## Acknowledgements

We would like to acknowledge the significant contribution of Prof. Paolo Colonna as imaging expert and reviewer of the manuscript.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

**Funding:** none declared.

## References

1. Kahlbau H, Antunes HT, Rodrigues I, Carvalho R, Fragata J. Pulmonary artery sarcoma masquerading as subchronic pulmonary thromboembolism. *J Card Surg* 2016;**31**:529–530.
2. Chen P-W, Liu P-Y. Pulmonary artery sarcoma mimicking pulmonary embolism. *BMJ Case Rep* 2018;**2018**:bcr2018226999.
3. Gulati G, Sharma S, Kothari SS, Juneja R, Saxena A, Talwar KK. Comparison of echo and MRI in the imaging evaluation of intracardiac masses. *Cardiovasc Intervent Radiol* 2004;**27**:459–469.
4. Judson I, Verweij J, Gelderblom H, Hartmann JT, Schöffski P, Blay JY et al.; European Organisation and Treatment of Cancer Soft Tissue and Bone Sarcoma Group. Doxorubicin alone versus intensified doxorubicin plus ifosfamide for first-line treatment of advanced or metastatic soft-tissue sarcoma: a randomised controlled phase 3 trial. *Lancet Oncol* 2014;**15**:415–423.
5. Hara H, Kawamoto T, Fukase N, Kawakami Y, Takemori T, Fujiwara S et al. Gemcitabine and docetaxel combination chemotherapy for advanced bone and soft tissue sarcomas: protocol for an open-label, non-randomised, Phase 2 study. *BMC Cancer* 2019;**19**:725.
6. George JC, Tang A, Markowitz A, Gilkeson R, Hoit BD. Papillary fibroelastoma of the pulmonic valve: evaluation by echocardiography and magnetic resonance imaging. *Echocardiography* 2008;**25**:433–435.