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CASE REPORT
Cardiac imaging

A rare case of anaerobic streptococci endocarditis in a young female with bicuspid aortic valve: case report

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Background Bicuspid aortic valve is the most common congenital cardiovascular malformation and occurs in 1–2% of the population. The haemodynamic changes appear early, leading to tissue damage and predisposing to germs attachment. The development of perivalvular extension is a constant in bicuspid aortic valve endocarditis. Infective endocarditis with anaerobic bacteria is a rare condition with a high rate of mortality.

Case summary We report a case of a young female with bicuspid aortic valve infective endocarditis. Involved bacteria were anaerobic streptococci, and the clinical course of the diseases was very aggressive. The echocardiographic evaluation revealed aortic and mitral regurgitation, perivalvular abscess, ventricular septum defect, and pericardial effusion. The surgery approach consisted of the aortic valve replacement with a mechanical prosthesis after radical resection of aortic root abscess and reconstruction of the annulus. The ventricular septum defect was also closed with a pericardial patch. Anticoagulation started the first day after surgery. The patient was received antibiotic therapy for 10 days before and 4 weeks after surgical intervention. Evolution was very good at 1 and 6 months follow-up.

Discussion This is a severe case of endocarditis, complicated with extensive valvular destruction, aortic root abscess, and fistula. Perivalvular complications are frequent in patients with bicuspid aortic valve endocarditis. The 'take away' message is that echocardiography is an essential tool for diagnosis, management, and follow-up of patients with infective endocarditis.

Keywords Echocardiography • Infective endocarditis • Bicuspid aortic valve • Case report

Learning points

- Infective endocarditis with anaerobic streptococci on a bicuspid aortic valve has an aggressive clinical course.
- A proper clinical examination and echocardiographic evaluation play an essential role in the diagnosis, management, and follow-up.
- When severe complications are present, early surgery is the best option.

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Introduction

Bicuspid aortic valve (BAV) is the most common congenital cardiovascular malformation, occurring in 1–2% of the population.[1] Patients with BAV aortic regurgitation (AR) are at high risk for infective endocarditis (IE).[2] Common BAV IE complications include valve destruction and aortic ring abscess.[5] Anaerobic IE is uncommon but has a high mortality rate.[7]

Timeline

Prior to the presentation (4 days before)	Patient presented to the emergency department with dyspnoea at rest, fatigue upon minimal exertion, fever, leucocytosis, and anaemia. She was investigated by the emergency, infectious disease, and haematology departments.
Day 0	Patient admitted to the cardiology department with signs and symptoms of heart failure, fever, sweats, and heart murmurs. She had no past medical history or regular medications. Transoesophageal echocardiogram (TOE) revealed aortic regurgitation, mitral regurgitation (MR), large vegetations on the bicuspid aortic valve (BAV), perivalvular abscess, ventricular septal defect (VSD), and pericardial and pleural effusion. Blood was collected. Treatment was initiated for heart failure (HF), and empirical antimicrobial treatment for infective endocarditis was started.
Day 1	Transoesophageal echocardiogram confirmed the transthoracic echocardiography findings. Laboratory investigations showed leucocytosis, anaemia, increased erythrocyte sedimentation rate, and increased levels of C-reactive protein. Abdominal ultrasound revealed hepatomegaly and splenomegaly.
Day 4	Blood cultures were positive for anaerobic streptococci susceptible to the initial antibiotic regimen. Patient's fever resolved, but the signs and symptoms of HF persisted.
Day 9	The surgical intervention consisted of resecting the abscess, debridement of the infected tissues, reconstructing the annulus, and replacing the native BAV with a mechanical Sorin Carbomedics 21 mm prosthesis. The VSD was closed with a pericardial patch. Oral anticoagulation was started the first day after surgery. Antimicrobial therapy continued for 4 weeks.
Day 39	Patient was discharged with no remaining symptoms and continued oral anticoagulation.
1 and 6 months	Patient was asymptomatic with continued oral anticoagulation. Transthoracic echocardiography revealed normal functioning of the aortic prosthesis, mild MR, and standard dimensions and an ejection fraction of the left ventricle.

Case presentation

We report a case of a 29-year-old female who was hospitalized with dyspnoea at rest and fatigue with minimal exertion over the preceding 3 weeks. The patient also complained of fever and sweating. She had no past medical history or regular medications. Four days before her hospitalization, she presented to the emergency department with these symptoms. At that time, neither cardiac nor abdominal ultrasounds were performed and the patient was referred to the infectious diseases department. There, laboratory investigations revealed leucocytosis, anaemia, and increased erythrocyte sedimentation rate (ESR). An abdominal ultrasound showed hepatomegaly and splenomegaly. Analysis of a 50 mL serous liquid from the right pleura excluded tuberculosis. The patient continued to be evaluated by the haematology department, where clinical examination identified heart murmurs. Because of the fever and murmur, IE was suspected, even though she had no prior history of cardiovascular disease.

A physical examination by the cardiology department revealed orthopnoea, rales in the lung bases, and cardiomegaly. Her blood pressure was 110/60 mmHg, and her heart rate was 112 beats/min. Cardiac auscultation identified a Grade III/VI systolic murmur and a diastolic murmur at the third left sternal border. The spleen was palpable. A 12-lead electrocardiography showed sinus rhythm and a chest X-ray confirmed the cardiomegaly and diffuse interstitial prominence (*Figure 1A*): Laboratory investigations showed anaemia (haemoglobin: 8.3 g/dL), leucocytosis [white blood cell count : 8700 per mm^3, 85% polymorphonuclear cells, increased levels of C-reactive protein (16.58 mg/dL), and increased ESR (30 mm/1 h)]. Blood cultures were collected before administering antibiotics. The anaerobic streptococci identified on the fifth day of admission were susceptible to the initial antimicrobial regimen.

Transthoracic echocardiography (TTE) revealed a BAV with two cusps (right and left) with a dome opening (*Figure 1B*, Supplementary material online, *Video S1*) and mobile masses with dimensions of 12 mm × 2.3 mm and 6 mm × 1.6 mm (*Figure 1C*) severe AR, (*Figure 1D*, Supplementary material online, *Videos S2 and S3*) moderate mitral regurgitation (MR) (*Figure 1E*, Supplementary material online, *Video S3*) aborted abscess (pseudoaneurysm) located in the mitral-aortic intervalvular fibrous area (Supplementary material online, *Video S2*) fistulae in the membranous ventricular septum between the left and right ventricles (*Figure 1F* and Supplementary material online, *Video S4*) pericardial and right pleural effusion (Figure 1C, D, E, F and Supplementary material online, *Videos S3 and S4*). Left atrial volume index (LAVi) was increased: 40 mL/m^2. Left ventricle diameters were also increased: end-diastolic diameter (LVEDD): 62 mm, end-systolic diameter (LVESD): 48 mm. Left ventricle ejection fraction (LVEF) was

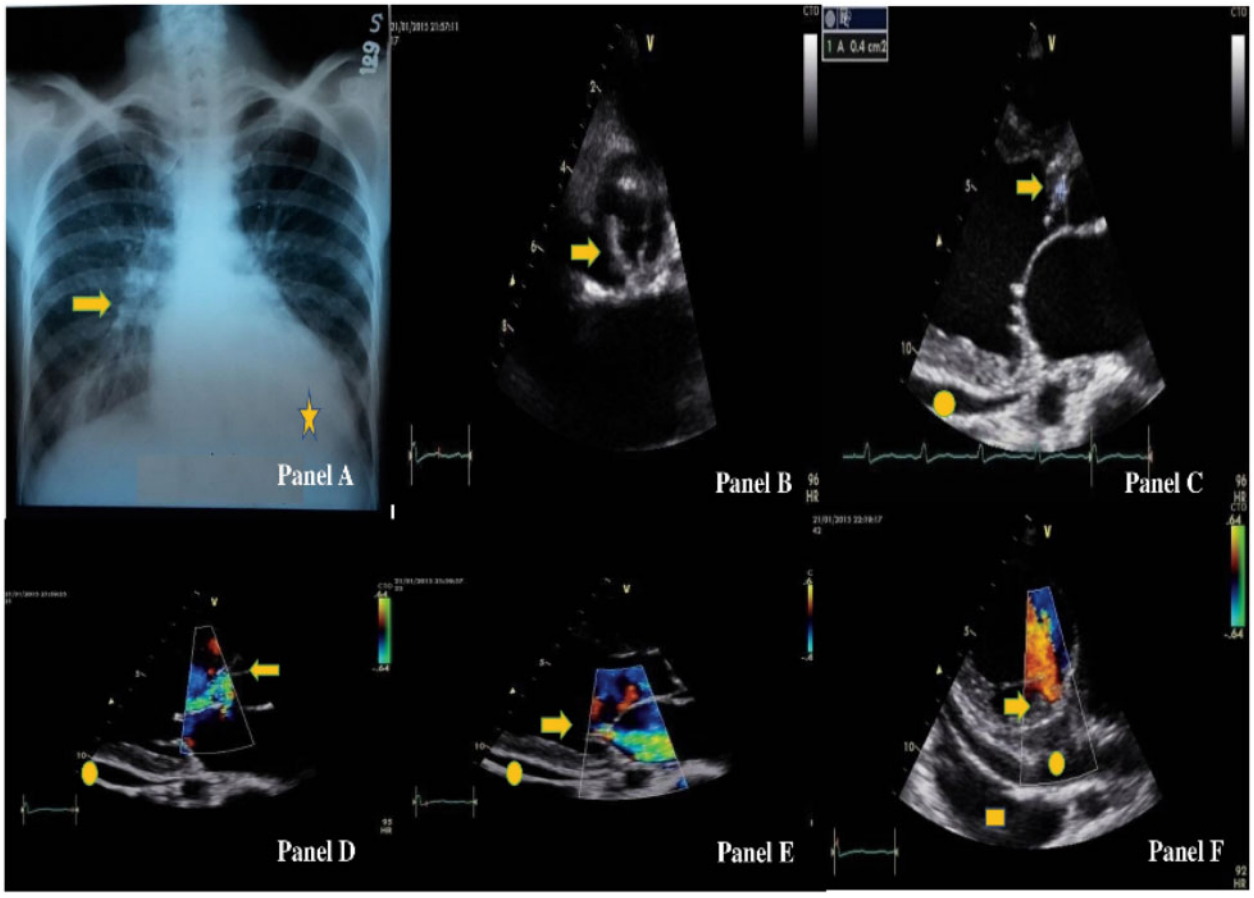

Figure 1 (*A*) Chest radiographs—cardiomegaly and diffuse interstitial prominence. (*B*) Transthoracic echocardiography—parasternal short-axis view at the level of great arteries with zoom—bicuspid aortic valve with two thick aortic leaflets and dome opening. (*C*) Transthoracic echocardiography—parasternal long-axis view—vegetation at the level of aortic valve, pericardial effusion. (*D*) Transthoracic echocardiography—parasternal long-axis view—aortic regurgitation, pericardial effusion. (*E*) Transthoracic echocardiography—parasternal long-axis view—mitral regurgitation, pericardial effusion. (*F*) Transthoracic echocardiography—modified parasternal short-axis view at the level of papillary muscles: ventricular septum defect, pericardial effusion, pleural effusion.

55%. The dimensions of the right cavities were normal. Tricuspid regurgitation (TR) was mild and pulmonary artery systolic pressure (PASP): 35 mmHg. A transoesophageal echocardiogram (TOE) confirmed the TTE findings of vegetations on the BAV (*Figure 2A–D*, Supplementary material online, *Video S5* and *S6*), aborted abscess (*Figure 2B, C* and Supplementary material online, *Videos S5* and *S6*), AR (*Figure 2C*, Supplementary material online, *Video S6*), MR (*Figure 2D*), and restrictive perimembranous ventricular septal defect (VSD) (*Figure 2E*) with left-to-right shunting and a systolic gradient of 112 mmHg (*Figure 2F*). Holter monitoring for 24 h did not detect arrhythmias.

According to the modified Duke criteria, the patient had a definite diagnosis of IE.[8] Empirical antimicrobial treatment started with amoxicillin (2 g every 6 h) and gentamycin (3 mg/kg/day intravenously). She also received treatment for heart failure (HF). Her fever rapidly diminished, but the symptoms of HF persisted. Based on the guidelines for managing IE, urgent surgical intervention was indicated due to HF, large vegetations, and uncontrolled local infection.[8] The decision was made to transfer her to the cardiovascular surgery department. Intraoperative TOE confirmed the TTE and TOE findings and provided detailed information. The VSD was 2 mm with morphological features of a perforation. Mitral regurgitation was moderate, without vegetations or structural abnormalities of the mitral valve. After 10 days of antibiotics, surgical correction consisted of abscess resection, debridement of the surrounding infected tissues, reconstruction of the annulus, then replacing the native BAV with a mechanical Sorin Carbomedics 21 mm prosthesis. The VSD was closed with a pericardial patch. Oral anticoagulation started the first day after surgery, and antibiotic therapy continued for 4 weeks.

At the 1-month follow-up visit, the patient was asymptomatic with normal electrocardiography. TTE revealed normal prosthesis function (*Figure 3A–C*). Six months after surgery, she had no symptoms, and TTE showed a standard range of prosthesis function (*Figure 3E*), mild MR (*Figure 3D*), LVEDD: 57 mm, LVESD: 38 mm, LVEF: 56%, and LAVi: 39 mL/m^2.

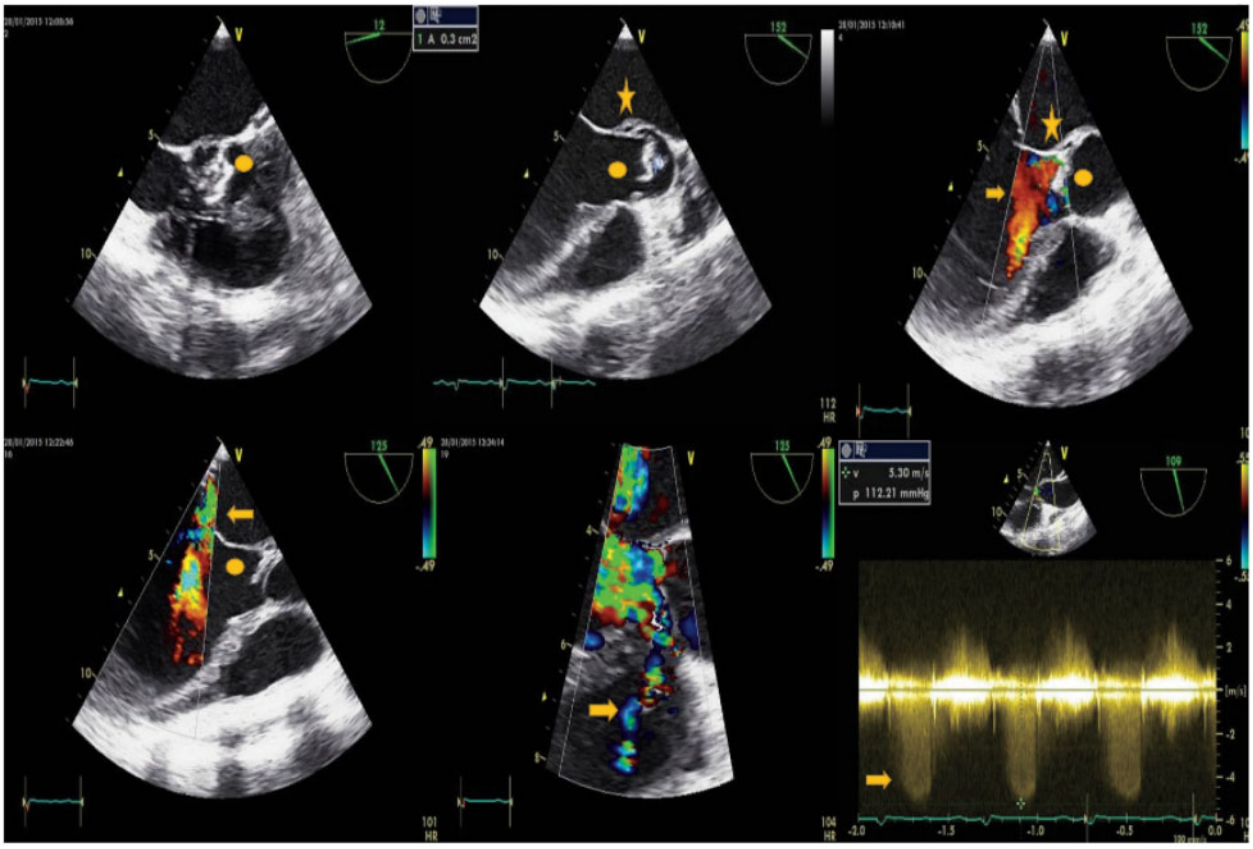

Figure 2 (*A*) Transoesophageal echocardiogram—short-axis view at the level of aortic valve: bicuspid aortic valve with vegetations, aborted abscess at the level of mitral-aortic intervalvular fibrosa area. (*B*) Transoesophageal echocardiogram—long axis at the level of aortic valve: vegetations at the level of aortic valve, aborted abscess at the level of mitral-aortic intervalvular fibrosa area. (*C*) Transoesophageal echocardiogram—long-axis view at the level of aortic valve: aortic regurgitation, vegetations at the level of aortic valve, aborted abscess at the level of mitral-aortic intervalvular fibrosa area. (*D*) Transoesophageal echocardiogram—long-axis view at the level of aortic valve: mitral regurgitation, vegetations at the level of aortic valve. (*E*) Transoesophageal echocardiogram—long-axis view: ventricular septum defect. (*F*) Transoesophageal echocardiogram—CW Doppler: restrictive ventricular septum defect with left to right flow, pressure gradient 112 mmHg.

Discussions

This case report is notable for its rare aetiology and complications in a young female with IE on a BAV. Haemodynamic changes appear early on malformed aortic valves[3], leading to tissue damage and predisposing the valve to the microbial attachment.[4] Infective endocarditis develops on 16.22% of BAVs[5] and may lead to aortic root abscess.[6]

Anaerobic bacteria are an unusual but ominous aetiology of IE, with a mortality rate of 21–43%. Anaerobic cocci generally originate from the lower respiratory tract.[7] They are fastidious, difficult to isolate, and often overlooked.[7] Our patient had no prior history of heart disease, which delayed the diagnosis. She was admitted at a late stage of the disease when the echocardiographic examination revealed large vegetations and extensive tissue damage. Sensitivity and specificity for vegetation and abscess detection are high for both TTE and TOE.[8]

One of the challenges of this case was to determine if the interventricular communication was a congenital defect or a fistula. An abscess develops on the weakest side of the aortic annulus, near the membranous septum, and may rupture into adjacent chambers creating an intracardiac fistula.[9] We considered the VSD to be an extension of the disease because the right side of the heart was unremarkable on echocardiography, with mild TR and an estimated PASP of 35 mmHg. The intraoperative morphology also suggested that the VSD was a complication of the disease.

Previous studies have shown that IE causes more than half of the cases of severe AR in patients with BAV.[1] In our case, the AR may have been due to the BAV or a complication of IE because anaerobic bacterial endocarditis produces severe valve destruction. The infection may extend to the anterior mitral leaflet from a primary AR jet.[10] The mechanism of the MR, in this case,s was secondary to left ventricle dilatation because the echocardiogram did not detect vegetations or damage to the mitral valve. Intraoperative TOE and surgical

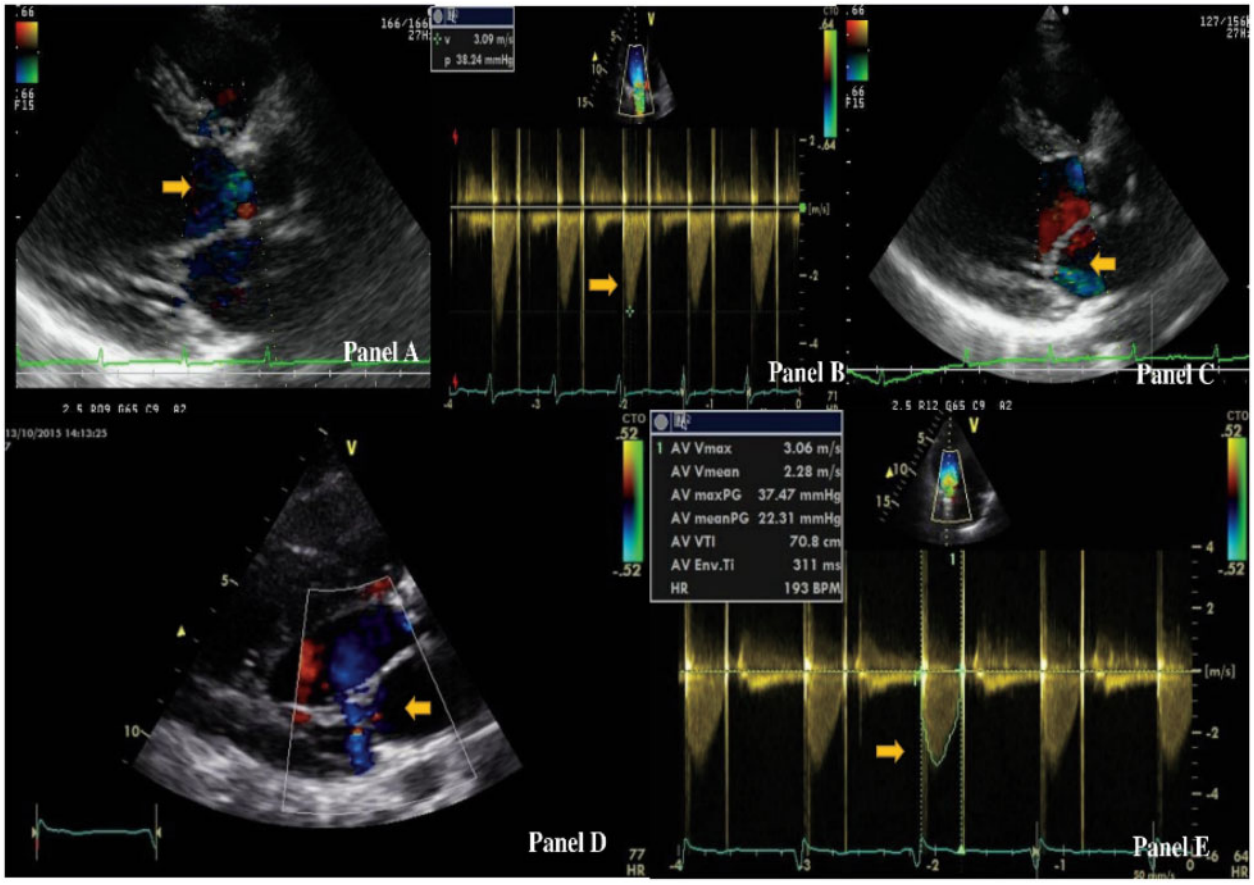

Figure 3 (*A*) Transthoracic echocardiography—parasternal long-axis view: mechanical prosthesis in aortic position—at 1-month follow-up visit. (*B*) Transthoracic echocardiography—parasternal long-axis view: mechanical prosthesis in aortic position with colour Doppler showing normal flow—at 1-month follow-up visit. (*C*) Transthoracic echocardiography—CW Doppler at the level of mechanical prosthesis in aortic position- maximal pressure gradient 38.24 mmHg—at 1-month follow-up visit. (*D*) Transthoracic echocardiography—parasternal long-axis view: mild mitral regurgitation—at 1-month follow-up. (*E*) Transthoracic echocardiography—CW Doppler at the level of mechanical prosthesis in aortic position-maximal pressure gradient 37.47 mmHg—at 6-month follow-up visit.

examination confirmed this hypothesis. The MR was mild when evaluated by TTE 6 months after surgery.

According to guidelines and the aggressive nature of anaerobes on a malformed aortic valve, early surgery was the best option to avoid progressive HF, perivalvular extension, and systemic embolism.[11] Bicuspid aortic valve is an abnormality with a 30-fold higher risk of IE than the general population.[12] The present guidelines recommend prophylaxis only high-risk patients[12] and do not categorize BAV as high risk of IE.[13] The clinical course is aggressive and complications are more common in patients with BAV.[14]

Conclusions

This was a rare case of IE due to anaerobic streptococci on a BAV. Both the aggressive nature of the microbe and the aortic valve malformation contributed to a severe disease course. The presentation emphasizes the importance of an adequate diagnosis and therapeutic approach for a BAV IE that involves anaerobic streptococci.

Lead author biography

Despina-Manuela Toader graduated from the University of Medicine and Pharmacy Craiova-Faculty of Medicine- Romania. She trained in cardiology at the University Hospital Bucharest and the Institute of Cardiovascular Disease CCIliescu Bucharest. Dr Toader received her Doctorate in Medicine from the University of Medicine and Pharmacy Craiova – The Faculty of Medicine- Romania. The title of her thesis was: Diastolic dysfunction following acute myocardial infarction. She is certificated by EACVI in transthoracic and transesophageal echocardiography. She is currently a senior

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Supplementary material

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

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s 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.

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