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**CASE REPORT**
*Congenital heart disease*

# A rare late finding in corrected tetralogy of Fallot: a case report

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**Introduction** Isolated pulmonary valve endocarditis is a rare phenomenon. Pulmonary prosthesis endocarditis is even more unusual, with only about 50 descriptions in worldwide literature, and its diagnosis and treatment is a challenge. Due to the increasing number of surgically corrected tetralogy of Fallot (TOF) patients, that often include pulmonary valve implantation, this clinical scenario is likely to become more frequent.

**Case presentation** We describe a 37-year-old man with a previously implanted biologic pulmonary prosthesis after a TOF correction that presented to the emergency department with new-onset fever, orthopnoea, and lower limb oedema. Blood cultures were positive for *Streptococcus mitis*. Transthoracic echocardiography showed a large mobile mass in the right ventricular outflow tract, apparently originating from the pulmonary prosthesis. Transoesophageal echocardiography (TOE) showed the presence of multiple mobile structures arising from the arterial surface of the prosthesis, extending into the right pulmonary artery and causing right ventricular obstruction. Antibiogram guided treatment was administered and surgery was performed, removing a 9 cm vegetation and replacing the valve. Patient recovered well and was discharged 35 days after.

**Discussion** In right-sided endocarditis, surgery indications and its timing are much less clear than in left-sided infections, but current literature describes it as associated with a significant morbidity, mortality, and high likelihood of requiring surgery. Large vegetations and clinical signs of haemodynamic impact should prompt consideration of early surgical intervention. The combination of transthoracic and TOE allowed a correct diagnosis and a timely treatment.

**Keywords** Tetralogy of Fallot • Case report • Pulmonary valve • Endocarditis • Right ventricle obstruction

## Learning points

- In surgically corrected tetralogy of Fallot patients with fever of unknown origin, prosthetic pulmonary valve endocarditis needs to be adequately investigated.
- As echocardiography of the pulmonary valve is technically difficult, a high clinical suspicion is needed and signs of right heart failure should always be searched for.
- General guidelines do not address this issue; we recommend early surgical intervention when large vegetations, sustained fever, and clinical signs of haemodynamic impact are present.

## Introduction

Prosthetic valve endocarditis is a rare complication of valve replacement surgery with an incidence of 0.3–4.2% per patient-year,[1,2] and accounts for approximately 20% of all cases of infective endocarditis (IE).[3] Most cases of prosthetic valve endocarditis regard adults with prosthetic aortic or mitral valve infections; reports of prosthetic pulmonary valve endocarditis (PVE) are rarely described outside major cohort reviews that usually focus on left-sided heart valves.[4] PVE has about 50 descriptions in worldwide literature,[4] but due to the increasing number of surgically corrected tetralogy of Fallot (TOF) patients (that often include pulmonary valve implantation) this

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scenario is likely to become more frequent, and the number of PVE has been rising, especially in percutaneously treated patients.[2] It is associated with high morbidity and mortality,[5] and its diagnosis is challenging. Due to the anterior position and variable fibrosis of the pulmonary valve (PV), transoesophageal echocardiography (TOE) use is technically difficult.[6] A high clinical suspicion is needed and thus reporting PVE presentation and imaging features is essential.

## Timeline

| | |
|---|---|
| Day 1 | Hospital admission: fever >72 h of unknown origin + signs of right heart failure |
| Day 3 | Positive blood cultures for relatively resistant to penicillin *Streptococcus mitis* |
| | Transthoracic echocardiogram (TTE) suggestive of pulmonary valve endocarditis (PVE) |
| | Started on iv amoxicillin 200 mg/kg/day plus iv gentamicin 3 mg/kg/day |
| Day 4 | Transoesophageal echocardiography confirmed diagnosis of PVE with right ventricular outflow tract obstruction |
| Day 5 | Patient was transferred to the surgical centre |
| Day 7 | Sustained fever. Repeated blood cultures—identical result |
| | Antibiotic treatment was empirically changed to iv Vancomicin 30 mg/kg/day plus iv gentamicin 3 mg/kg/day |
| Day 10 | Sustained fever. Repeat TTE—identical to previous exams |
| Day 12 | Surgical valve replacement with a Freestyle valve n° 25 implantation |
| Week 6 | Hospital discharge |

## Case presentation

A 37-year-old man came to the emergency department with complaints of orthopnoea, non-productive cough and fever >38.5°C for over 72 h. Physical examination showed a lower limb oedema, positive hepatojugular reflux, and a cardiac systolic murmur. His blood analysis showed leucocitosis of 11.67 × $10^9$ cells/L (4.5–11) with neutrofilia of 9.37 × $10^9$ cells/L (2.0–8.5) and an elevated C-reactive protein of 198 mg/L (<5 mg/L). His chest X-ray had no signs of pulmonary congestion. His previous medical history included a TOF that was corrected in 1999, with a pulmonary biologic valve replacement 2 years later (with a Carpentier-Edwards Perimount® bioprosthesis), secondary to major pulmonary regurgitation.

After hospital admission, blood cultures were positive for relatively resistant to penicillin *Streptococcus mitis*, and directed intravenous antibiotics (amoxicillin plus gentamicin) were given. The transthoracic echocardiogram (TTE) showed a mobile mass arising from the pulmonary prosthesis (*Figure 1*, red arrow). In order to better define this finding, a TOE was performed, showing the presence of multiple mobile structures arising from the arterial surface of the prosthesis and also from the pulmonary trunk (PT), extending into the right pulmonary artery and causing right ventricular obstruction, with a mean and max right ventricular outflow tract (RVOT)-PT gradient of 35 and 52 mmHg, respectively (*Figures 2–4*).

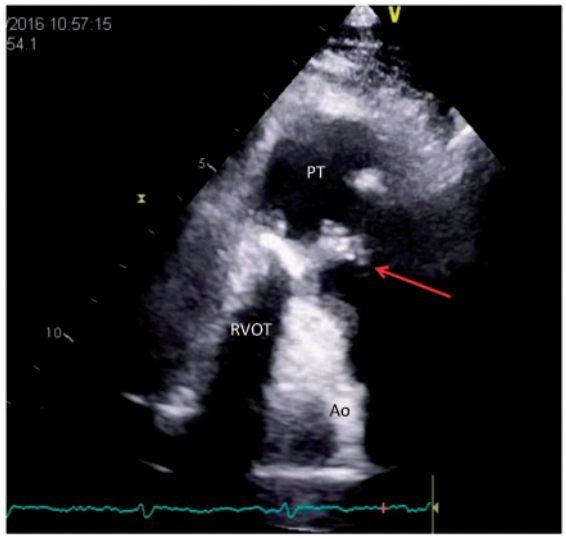


**Figure 1** Transthoracic echocardiogram. Ao, aortic valve; PT, pulmonary trunk; RVOT, right ventricular outflow tract.

The information obtained through both echocardiography techniques allowed for the definitive diagnosis of pulmonary prosthesis endocarditis. Due to sustained fever and positive blood cultures despite directed IV-therapy, cardiac surgery was performed with PT opening during cardiopulmonary bypass to allow the removal of a 9 cm vegetation and a Freestyle valve n° 25 implantation. The patient was discharged after 6 weeks course of antibiotics and, after 1 year follow-up, is currently well. He was advised to perform prophylaxis according to current guidelines.[1]

## Discussion

We report a case of a pulmonary prosthesis endocarditis causing RVOT obstruction and right heart failure. At admission, the patient had fever of unknown origin and signs of right heart failure. After appropriate investigation, definitive diagnosis was obtained according to the modified Duke criteria for the diagnosis of IE, with two major and two minor criteria.[7] After an early surgical correction, the patient recovered well.

To recognize prosthetic PVE implies a high clinical suspicion, since imaging of the right side of the heart is difficult and many times incorrectly disregarded. The biggest case series published to date concluded that TTE and TOE should complement each other in the evaluation of patients with suspected PVE, but still fail to diagnose more than 10% of cases.[6]

In this case, the identified microorganism was a gram positive bacterium: *S. mitis*. The most commonly identified organisms in previously

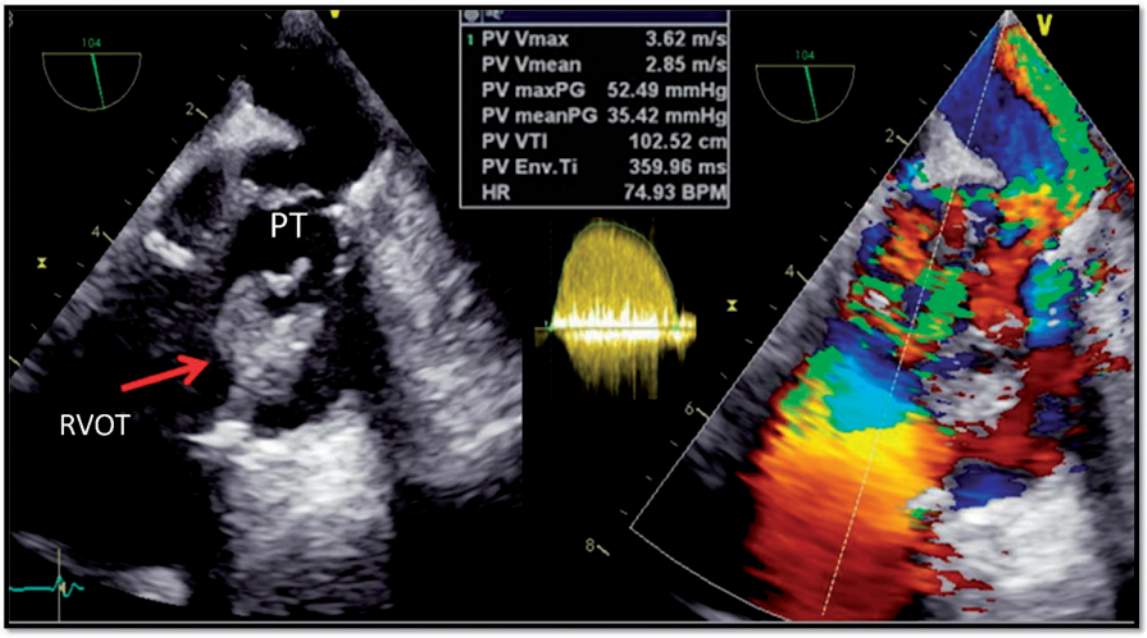


**Figure 2** Transoesophageal echocardiogram. Red arrow pointing to a mobile mass arising from the arterial side of the pulmonary prosthesis, extending into the right pulmonary artery and causing right ventricle obstruction; PT, pulmonary trunk; RVOT, right ventricular outflow tract.

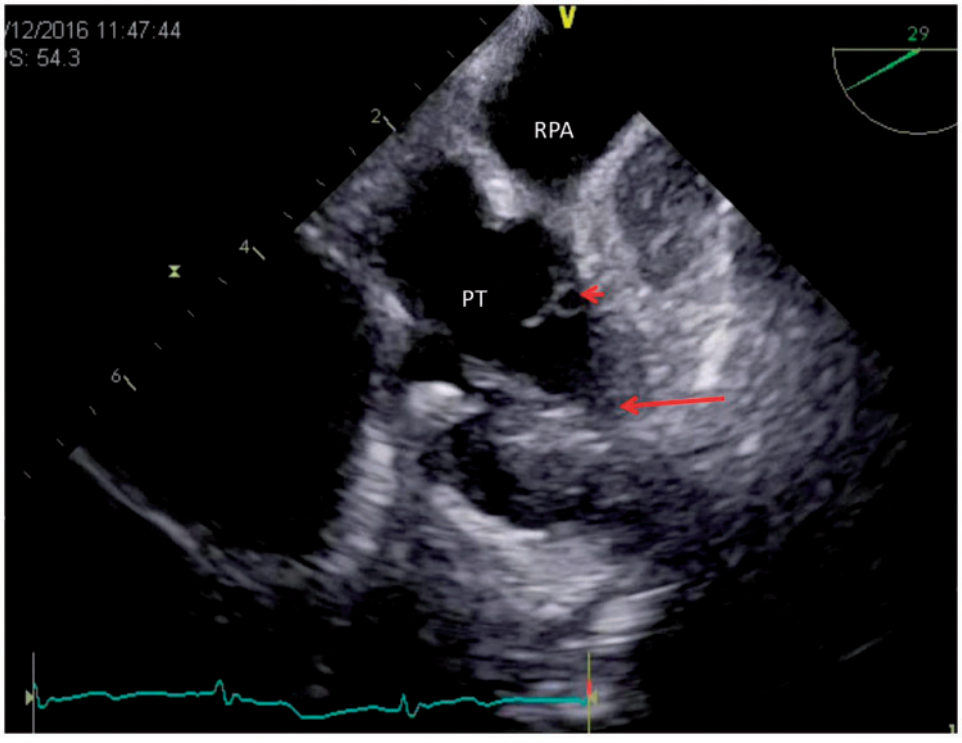


**Figure 3** Transoesophageal echocardiogram. Red arrow-head pointing to a filamentous structure arising from the PT and red arrow pointing to the mobile mass arising from the pulmonary prosthesis. PT, pulmonary trunk; RPA, right pulmonary artery.

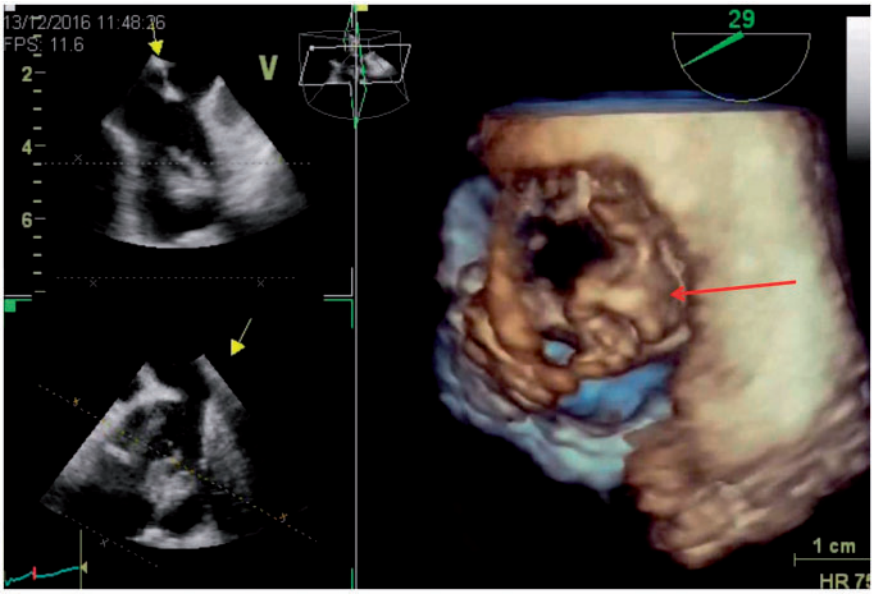


**Figure 4** 3D reconstruction from the transoesophageal echocardiogram. Red arrow pointing to the mass arising from the pulmonary prosthesis.

described cases were *Staphylococcus aureus*, coagulase-negative staphylococcus and streptococcal species.[3,4,6] An upper respiratory tract infection (RTI) might have been the underlying cause for bacteraemia in this case, since *S. mitis* is an uncommon but known cause for RTI[8,9] and the patient later referred a short period of fever, with hoarseness and cough 1 month previous to the hospital admission.

In current literature, right-sided endocarditis surgical indications and timing are not clear.[1] The majority of published cases refer to sustained fever and signs of haemodynamic impact as major indications for a surgical treatment,[4,6] but no major consensus have been published.

In conclusion, this case represents the importance of searching for clinical signs of right heart failure as well as the relevance of an adequate right heart imaging, especially when clinical suspicion of PVE is present. As general guidelines do not address this issue, determining surgical indications and timing is a challenge. Transthoracic and TOE combination, despite technically difficult, allowed a correct diagnosis and a timely treatment. According to our case and to available literature, we recommend considering surgical treatment when large vegetations, sustained fever and clinical signs of haemodynamic impact are present.

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**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

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