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CASE REPORT
Adult congenital heart disease

Fulminant biventricular multivalvular infective endocarditis in complex congenital heart disease: a case report

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Background Infective endocarditis (IE) is a major issue during follow-up of adults with congenital heart disease (ACHD), leading to significant mortality.

Case summary A 37-year-old woman with transposition of great arteries and previous Mustard operation developed a drug-resistant pneumonia shortly after a pacemaker implant procedure performed at a local hospital. After referral to the ACHD centre, the patient was diagnosed with multivalvular IE with biventricular involvement by methicillin-resistant *Staphylococcus aureus*. On admission, the patient was already in acute respiratory distress and presented both systemic and pulmonary embolization. Despite adequate treatment was promptly started, the patient developed multiorgan failure.

Discussion This case depicts a particularly aggressive of infective endocarditis forms caused with biventricular involvement and multiple embolization. Patients with congenital heart disease are at high risk of IE with adverse impact on the prognosis. Early recognition and treatment are the keys to improve prognosis. Therefore, suspicion should be high, especially following invasive procedure, which should be preferably performed at ACHD specialized centres.

Keywords Congenital heart disease • Infective endocarditis • Biventricular infective endocarditis • ACHD • Case report

ESC Curriculum 2.2 Echocardiography • 4.11 Endocarditis • 4.9 Multivalvular disease • 5.9 Pacemakers • 9.7 Adult congenital heart disease

Learning points

- Methicillin-resistant *Staphylococcus aureus* may give particularly aggressive forms of infective endocarditis (IE) with multivalvular biventricular presentation and multiple embolization.
- Suspicion of IE should be immediately raised in case of persistent fever following an invasive procedure in complex patients with congenital heart disease.

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Introduction

Infective endocarditis (IE) is a frequent complication during follow-up of adults with congenital heart disease (ACHD) and is related to significant mortality.[1] We report a rare case of aggressive IE following pacemaker (PMK) implantation in a young patient with transposition of the great arteries (TGA) and previous Mustard operation.

Timeline

Infancy	Surgical correction of complex congenital heart disease with atrial switch procedure, ventricular septal defect closure, and implant of a valved conduit between left ventricle and pulmonary artery
May 2021	High-grade atrioventricular block→implant of a transvenous pacemaker (PMK)
July 2021	Drug-resistant pneumonia, treated with i.v. antibiotics in a local hospital
August 2021	New hospitalization due to recurrent fever spikes
1 October 2021	Adults with congenital heart disease centre referral: septic shock and severe acute respiratory failure
4 October 2021	Evidence of multiple vegetations in both subpulmonary left ventricle, on pacemaker leads, on the mitral valve, and in the systemic right ventricle→ diagnosis of infective endocarditis methicillin-resistant *Staphylococcus aureus* related
8 October 2021	Urgent neurosurgical procedure for evacuation of a large cerebral haemorrhage
10 October 2021	Percutaneous extraction of infected PMK leads
20 October 2021	Death due to multiorgan failure

Case presentation

A 37-year-old woman with a congenital diagnosis of transposition of TGA, ventricular septal defect, and pulmonary stenosis, who had undergone surgical correction during infancy with atrial switch procedure (Mustard operation), ventricular septal defect closure, and implant of a valved conduit between the left ventricle and the pulmonary artery, was not under specialized ACHD care. During follow-up, she developed a high-grade atrioventricular block, requiring a permanent transvenous PMK implant, which was performed at the local hospital. Shortly after the procedure, she presented with drug-resistant pneumonia, which was treated with i.v. antibiotics. After apparent full remission, the patient was discharged home, but she reported early recurrence of fever spikes, requiring hospitalization. Considering the history of complex congenital heart defect and the evidence of significantly raised inflammatory markers, the patient was referred to our ACHD centre for further diagnostic work-up and treatment. On admission, she was in critical conditions due to septic shock and severe acute respiratory failure requiring mechanical ventilation. She was therefore transferred to the intensive care unit. After stabilization of vital parameters, transthoracic and transoesophageal echocardiography with three-dimensional acquisitions demonstrated multiple mobile vegetations in the subpulmonary left ventricle attached to the PMK lead and to the mitral valve leaflets. Another mobile mass with maximal length of 1.7 cm was revealed in the right ventricular outflow tract, just below the aortic valve (*Figure 1* and *Videos S1–S5*). Moreover, an intraventricular abscess in the systemic right ventricle was detected as an anechoic round-shaped mass attached to a papillary muscle (*Figure 2* and *Video S6*). The patient showed signs of both systemic and pulmonary septic embolization: systemic embolization was initially suspected due to necrosis of the extremities (*Figure 3*). Total-body computed tomography (CT) showed small pleural-based peripheral thickenings with triangular morphology referable to areas of pulmonary infarction, hypodense ischemic areas in the spleen and in both kidneys. Cerebral CT revealed a large haemorrhage involving the left fronto-parieto-occipital region, with intraventricular extension into both frontal horns of lateral ventricles, into the third and fourth ventricles, with effacement of cortical sulci and midline shift of 11 mm (*Figure 4*). Blood cultures were positive for methicillin-resistant *Staphylococcus aureus* (MRSA), confirming the diagnosis of IE, according to modified Duke's criteria.[2] The patient was started on antibiotic therapy with daptomycin. Cardiac surgery was not deemed feasible due to haemodynamic instability. Infected PMK lead was extracted with a percutaneous procedure, and the patient received a temporary device implant. After CT, urgent surgical haematoma evacuation was successfully performed. Nevertheless, the patient developed severe refractory multiorgan failure and died few days later.

Discussion

We report a rare case of MRSA-related IE with biventricular involvement, in an ACHD patient following PMK implant. Infective endocarditis caused by *S. aureus* was previously mainly found in i.v. drug users. However, this has recently become one of the most common pathogens among healthcare-associated infections, leading to a higher rate of systemic and pulmonary embolization and increased mortality.[3] Healthcare-associated IE by *S. aureus* may be related to the presence of intracardiac catheter in up to 25% of cases[4] and are more likely caused by a MRSA strain.[3] Infective endocarditis with multivalvular involvement is a particularly aggressive form of infection, with higher risk of complications without a timely and adequate treatment.[5] Our case depicts an extremely uncommon presentation with biventricular vegetations and intracardiac abscess.

Complex ACHD patients are particularly prone to develop IE due to both presence of intracardiac prosthetic material and multiple invasive procedures required during follow-up. The incidence of IE has recently been estimated at 4 cases/100 000 patient/year, which is about a 30-fold increased risk compared to the general population.[6] Due to their peculiar anatomy, ACHD patients show also an increased risk of IE involving the right heart. Therefore, this special population requires specialized care for an appropriate follow-up by ACHD experts with a specific training in imaging and managing complex cardiac lesions. Patients with congenital heart disease should be advised and educated on antibiotic prophylaxis, which is recommended before invasive procedures including dental interventions for those at highest risk of IE, namely those with cyanotic defects and those with prosthetic material in the first 6 months following the procedure or lifelong in case of residual shunting or valvular regurgitation.[2]Whenever a complex, invasive procedure is necessary, it should be preferably performed at the ACHD centre, where all measures to minimize the risks of bacterial exposure must be undertaken and where a specialized team can offer the best course of treatment. Infective endocarditis should be immediately suspected in case of persistent fever following any interventions and should be ruled out with a thorough diagnostic work-up.

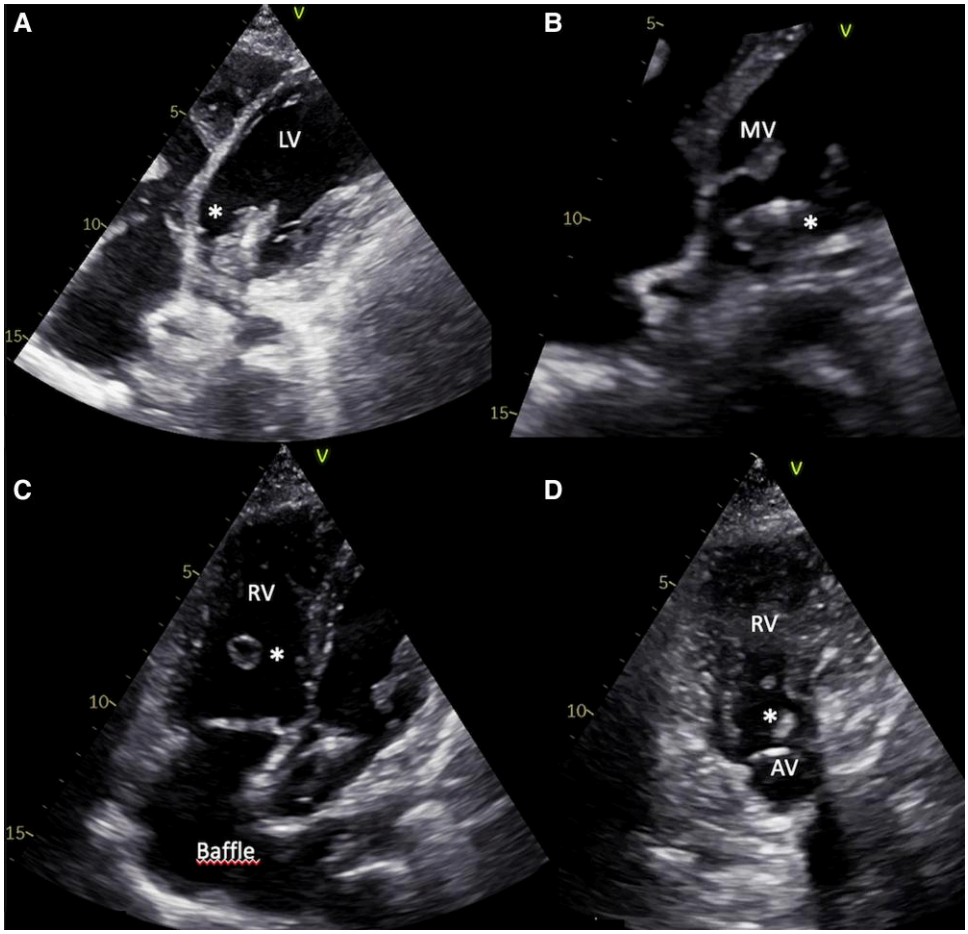

Figure 1 Transthoracic echocardiography demonstrating multiple intracardiac vegetations attached to the pacemaker lead inside the subpulmonary left ventricle (*A*), with involvement of the mitral valve leaflets (*B*). A rounded shaped formation with anechoic centre was noted in the systemic left ventricle (*C*), and a mobile mass was revealed in the right ventricular outflow tract, below the aortic valve (*D*). AV, aortic valve; MV, mitral valve; LV, left ventricle; RV, right ventricle; asterisks, vegetations/abscess.

Echocardiography is the first-line imaging technique and provides valuable diagnostic clues. Transoesophageal echocardiography performed by expert operators with understanding of underlying anatomy may allow a better visualization of some cardiac structures, such as subvalvular apparatus or PMK leads. Cardiac magnetic resonance may provide additional data on intracardiac abscess, but, unfortunately, could not be performed in our patient due to clinical instability.

Early referral to the ACHD centre and a prompt diagnosis to initiate the appropriate treatment are paramount to improve the prognosis and could be lifesaving in complex ACHD patients. It should be noted that, in the case presented, the patient was already in critical conditions upon arrival at our institution leading to adverse outcome, despite targeted therapy.

Conclusions

Patients with congenital heart disease are at high risk of IE, especially following invasive procedures, which should be performed with strictly sterile conditions in an ACHD specialized centre. Early recognition could improve the prognosis, especially in case of particularly aggressive forms caused by MRSA with biventricular involvement and multiple embolization.

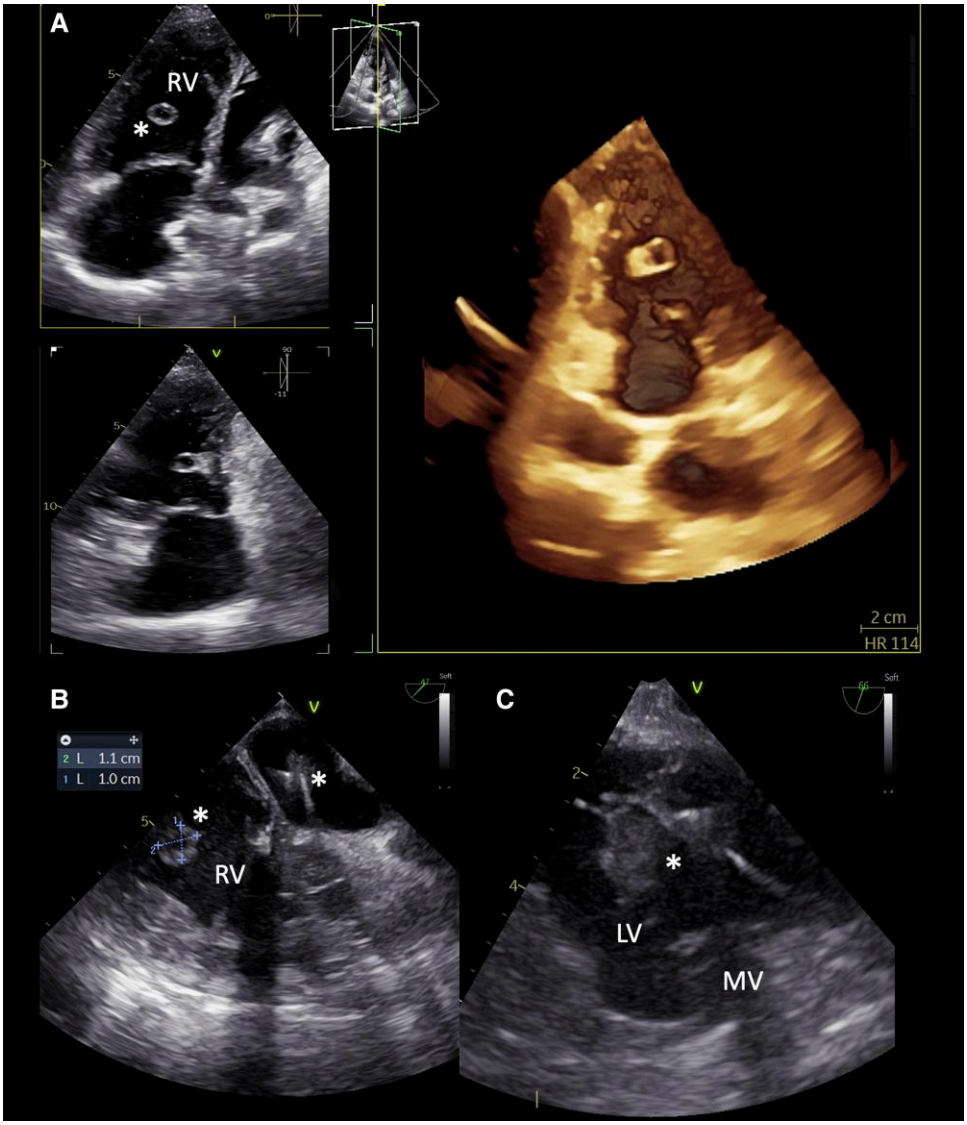

Figure 2 Advanced echocardiography and transoesophageal echocardiography allow a better definition of intracardiac masses. *A*) Multiplane and 3D echocardiography clarified the extension of the rounded shaped formation, showing that it was in continuity with the papillary muscle. *B*) Transoesophageal echocardiography midoesophageal four-chamber view showing bilateral presence of vegetations. *C*) Transoesophageal echocardiography transgastric view with a 60° angle, focused on the left ventricle allowed complete visualization of the pacemaker lead with multiple vegetations. MV, mitral valve; LV, left ventricle; RV, right ventricle; asterisks, vegetations/abscess.

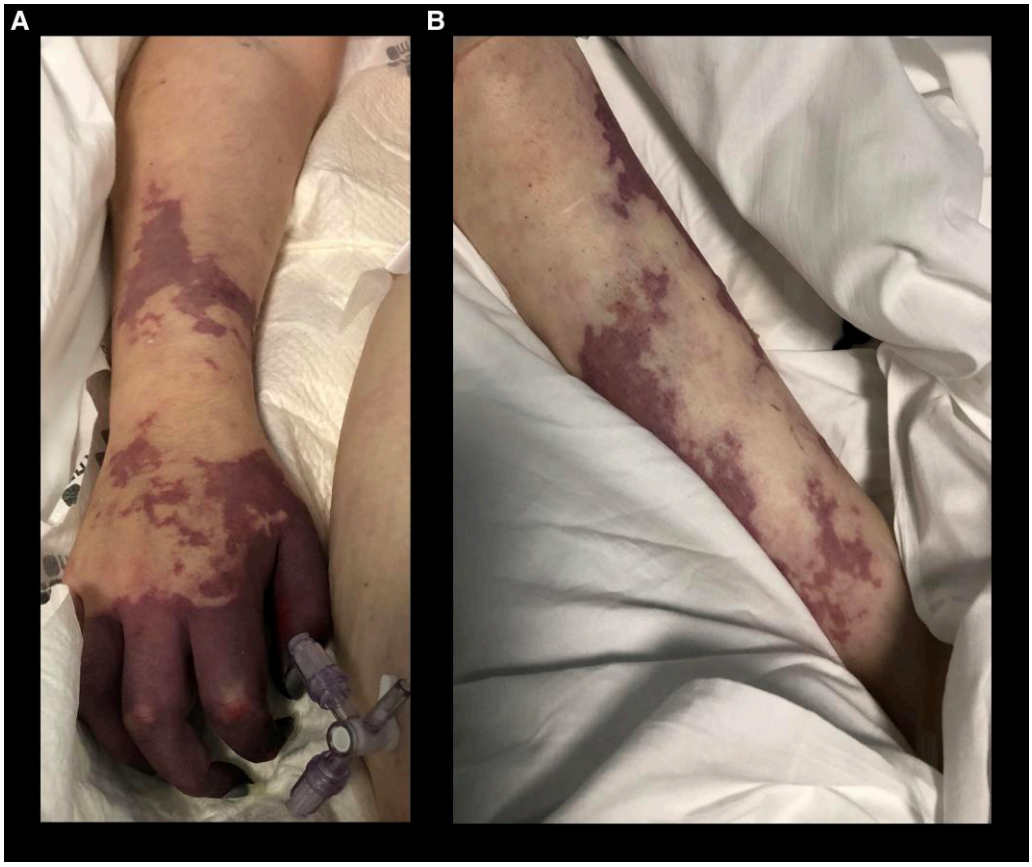

Figure 3 Necrosis of the upper (*A*) and lower (*B*) extremities.

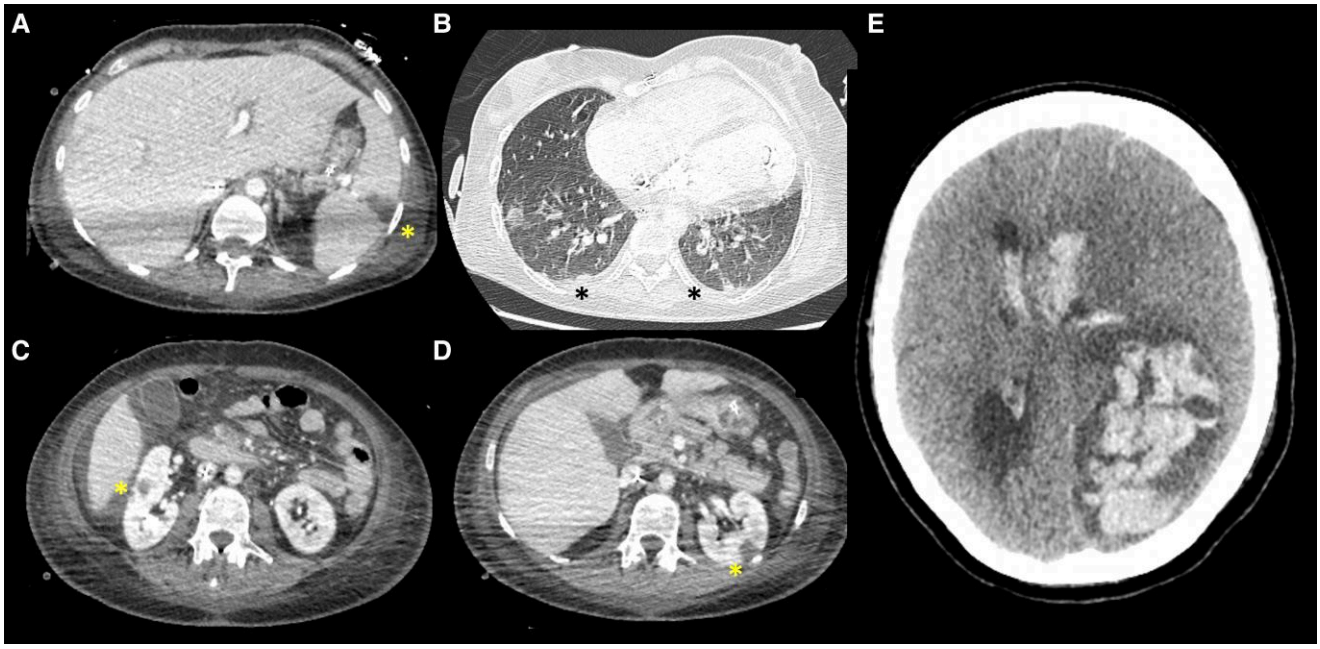

Figure 4 Total-body computed tomography demonstrating multiorgan embolization: hypodense ischaemic areas in the spleen (*A*), bilateral pleural-based peripheral thickenings with triangular morphology (*B*), ischaemic areas in both kidneys (*C* and *D*), and a large haemorrhage involving the left fronto-parieto-occipital region, with intraventricular extension into both frontal horns of lateral ventricles, with effacement of cortical sulci and midline shift of 11 mm (*E*). CT, computed tomography; asterisks, ischemic areas.

Lead author biography

Dr Giancarlo Scognamiglio, MD, PhD, is a Consultant Cardiologist and Lead of the Imaging Program in Adult Congenital Heart Disease at Monaldi Hospital in Naples, Italy. During his training, he specialized in cardiac imaging and congenital heart disease. He is author of chapters in four scientific books and more than 50 scientific papers on peer-reviewed journals. Dr Scognamiglio is Professor of Congenital Heart Disease in the Specialty Training Program in Cardiology at the University of Campania Luigi Vanvitelli, and he is a nucleus member of the ACHD working group of the European Society of Cardiology.

Supplementary material

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

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None declared.

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's relatives in line with COPE guidance.

Conflict of interest: None declared.

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Data availability

All data are available from the corresponding authors upon request.

References

1. Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, *et al.* 2020 ESC guidelines for the management of adult congenital heart disease. *Eur Heart J* 2021;**42**:563–645.
2. Habib G, Lancellotti P, Antunes MJ, Bongiorni MG, Casalta JP, Del Zotti F, *et al.* ESC Scientific Document Group. 2015 ESC guidelines for the management of infective endocarditis: the task force for the management of infective endocarditis of the European Society of Cardiology (ESC). Endorsed by: European Association for Cardio-Thoracic Surgery (EACTS), the European Association of Nuclear Medicine (EANM). *Eur Heart J* 2015;**36**:3075–3128.
3. Fowler VG Jr, Miro JM, Hoen B, Cabell CH, Abrutyn E, Rubinstein E, *et al. Staphylococcus aureus* endocarditis: a consequence of medical progress. *JAMA* 2005;**293**:3012–3021.
4. Røder BL, Wandall DA, Frimodt-Møller N, Espersen F, Skinhøj P, Rosdahl VT. Clinical features of *Staphylococcus aureus* endocarditis: a 10-year experience in Denmark. *Arch Intern Med* 1999;**159**:462–469.
5. Kim N, Lazar JM, Cunha BA, Liao W, Minnaganti V. Multi-valvular endocarditis. *Clin Microbiol Infect* 2000;**6**:207–212.
6. Kuijpers JM, Koolbergen DR, Groenink M, Peels KCH, Reichert CLA, Post MC, *et al.* Incidence, risk factors, and predictors of infective endocarditis in adult congenital heart disease: focus on the use of prosthetic material. *Eur Heart J* 2017;**38**:2048–2056.