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CASE REPORT
Adult congenital heart disease

A multidisciplinary approach to re-intervention in an Ebstein patient: a case report

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Background Ebstein's anomaly is a rare condition due to incomplete delamination of the tricuspid valve (TV) leaflets with downward displacement of the proximal leaflet attachments. It is associated with a smaller functional right ventricle (RV) and tricuspid regurgitation (TR) that is typically treated with TV replacement or repair. However, future re-intervention poses challenges. We describe a multidisciplinary team approach to re-intervention in a pacing-dependant Ebstein patient with severe bioprosthetic TV regurgitation.

Case summary A 49-year-old female patient underwent bioprosthetic TV replacement for severe TR in Ebstein's. Post-operatively, she developed complete atrioventricular (AV) block necessitating the implantation of a permanent pacemaker which included a coronary sinus (CS) lead as the ventricular lead. Five years later, she presented with syncope due to a failing ventricular pacing lead, and a new RV lead was positioned across the TV bioprosthesis due to the lack of CS options. Two years later, she presented with breathlessness and lethargy with severe TR identified on transthoracic echocardiography. She successfully underwent a percutaneous leadless pacemaker implant, extraction of existing pacing system, and implantation of valve-in-valve TV.

Discussion Patients with Ebstein's anomaly typically undergo TV repair or replacement. Following surgical intervention, owing to the anatomical location, patients can develop AV block requiring a pacemaker. Pacemaker implantation may involve a CS lead to avoid placing a lead across the new TV in efforts to avoid lead induced TR. Over time, these patients not uncommonly require re-intervention that can be challenging especially in pacing-dependant patients with leads across the TV.

Keywords Case report • Ebstein's anomaly • Tricuspid regurgitation • Leadless pacemaker

ESC Curriculum 4.5 Tricuspid regurgitation • 5.4 Atrial flutter • 5.9 Pacemakers • 5.7 Bradycardia • 9.7 Adult congenital heart disease

Learning points

- Leadless pacemakers could be considered for Ebstein patients with previous tricuspid valve (TV) replacement requiring re-intervention.
- Percutaneous tricuspid valve-in-valve replacement may be considered as an option in patients with isolated TV regurgitation who are at increased risk of surgical re-intervention.
- A multi-disciplinary percutaneous approach with a patient-individualized care plan can be successfully utilized for re-intervention in the complex Ebstein patient.

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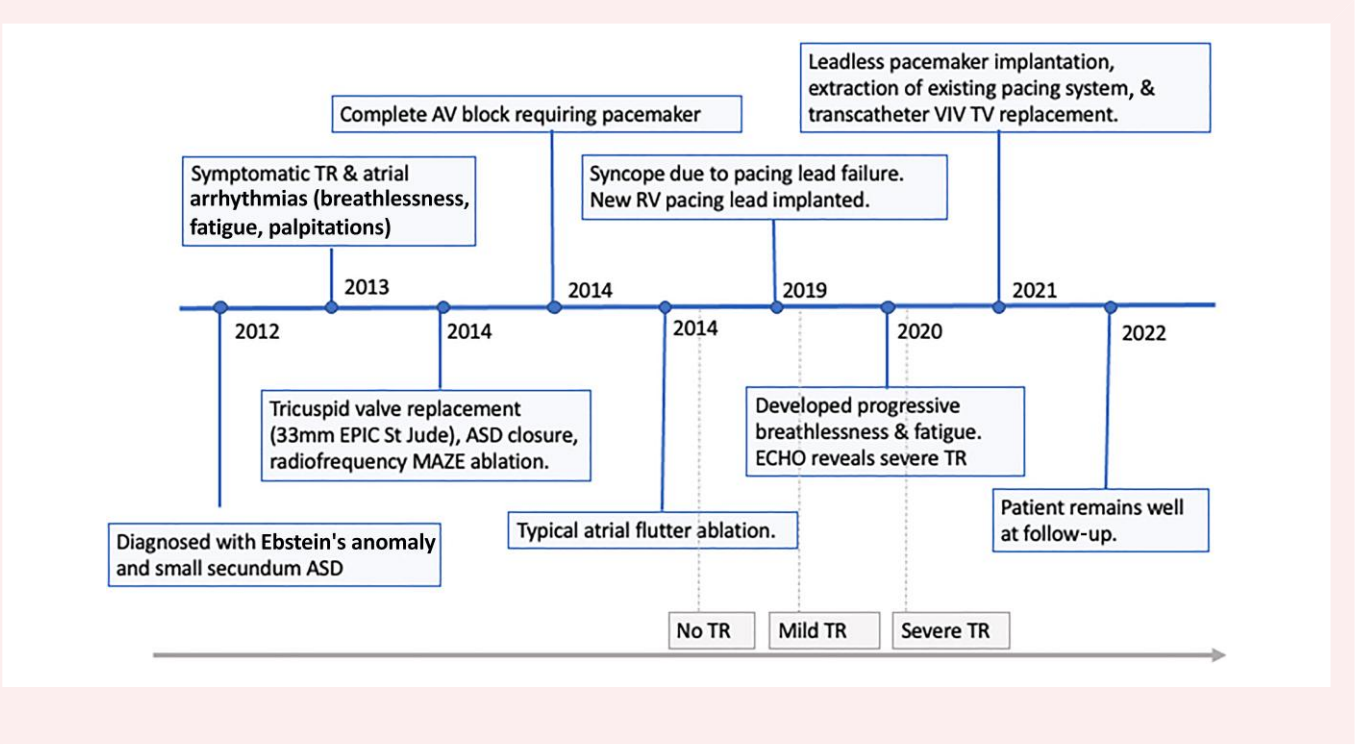
Introduction

Ebstein's anomaly (EA) affects one to five in 200, 000 people due to incomplete delamination of primarily the septal and inferior tricuspid valve (TV) leaflets.[1] Commonly, apical displacement of the proximal septal and posterior TV leaflet attachments occurs from the atrioventricular (AV) ring leading to 'atrialization' of the right ventricle (RV) with a smaller functional RV associated with tricuspid regurgitation (TR).[1] Treatment is determined by regurgitation severity and patient symptoms whilst taking into consideration associated abnormalities. The feasibility of the TV for repair or replacement is important in treatment planning. Here, we describe successful re-intervention in an EA patient with the implantation of a leadless pacemaker (PPM), extraction of an existing transvenous pacing system, and valve-in-valve (VIV) TV replacement.

Timeline



Case presentation

Eight years prior to presentation, a 49-year-old female presented with increasing breathlessness along with frequent palpitations. Transthoracic echocardiography (TTE) revealed EA with apical displacement of the TV leaflets by 10 mm/m^2 alongside severe TR. She was also noted to have a small atrial septal defect (ASD). Electrocardiogram (ECG) monitoring revealed atrial arrhythmias. In keeping with European Society of Cardiology (ESC) recommendations for severe symptomatic TR in EA (Class IC recommendation), she underwent surgery.[2] During the operation, the TV was felt not to be repairable to a durable level, and instead she had a combined procedure that included TV replacement with a 33 mm bioprosthetic valve, ASD closure, and a radiofrequency Maze procedure. Post-operatively, she developed complete AV block necessitating a PPM. A transvenous dual chamber PPM was implanted with the right atrial lead in the standard position. However, the ventricular lead was positioned in the coronary sinus (CS) to avoid placing a transvalvular lead. She represented post-operatively with further palpitations due to typical atrial flutter that was treated with radiofrequency ablation. Two months after, she was well with TTE confirming normally functioning TV with no TR.

She remained well until she represented 5 years later with recurrent syncope. ECG monitoring revealed capture failure on the CS pacing lead with a sudden large elevation in ventricular lead impedance and threshold. Prior to the pacing system revision, repeat TTE demonstrated moderate TR. Lead revision was challenging owing to a CS stenosis just before the bifurcation of the anterolateral branches. Proximal to the stenosis, poor stability, intermittent capture, and twitch at low outputs prevented a new CS lead. As safe ventricular pacing was imperative, a thinner SelectSecure 3830 (Medtronic Inc., Minneapolis, MN, USA) lead was placed in the lower RV septum. Subsequent TTE at 2 months reassuringly revealed unchanged moderate TR. Medical therapy included a novel oral anticoagulant (NOAC) and diuretic therapy to maximize valve longevity and reduce the TR.[3]

At follow-up, 4 months later, she noted breathlessness, abdominal bloating, and fatigue preventing her from attending the gym. On examination, there was a small amount of pedal oedema, and the jugular venous pressure was raised to the jaw line at 45°. Subsequent TTE revealed severe TR with preserved RV size and function (*Figure 1*). The mean gradient across the TV was 8 mmHg. She was initially given increased loop diuretic therapy whilst further assessments were performed.

At a multi-disciplinary team (MDT) meeting that included cardiologists, cardiothoracic surgeons, and cardiac anaesthetists, it was felt that the lead was likely splinting one of the TV leaflets down increasing the TR. Whilst surgical intervention and staged procedures were discussed, both the team and patient favoured a percutaneous combined procedure approach. Factors that influenced this decision included her young age as well as the fact this would be her second sternotomy. It was agreed to evaluate the severity of the TR following lead extraction before intervening on the valve in a combined percutaneous procedure

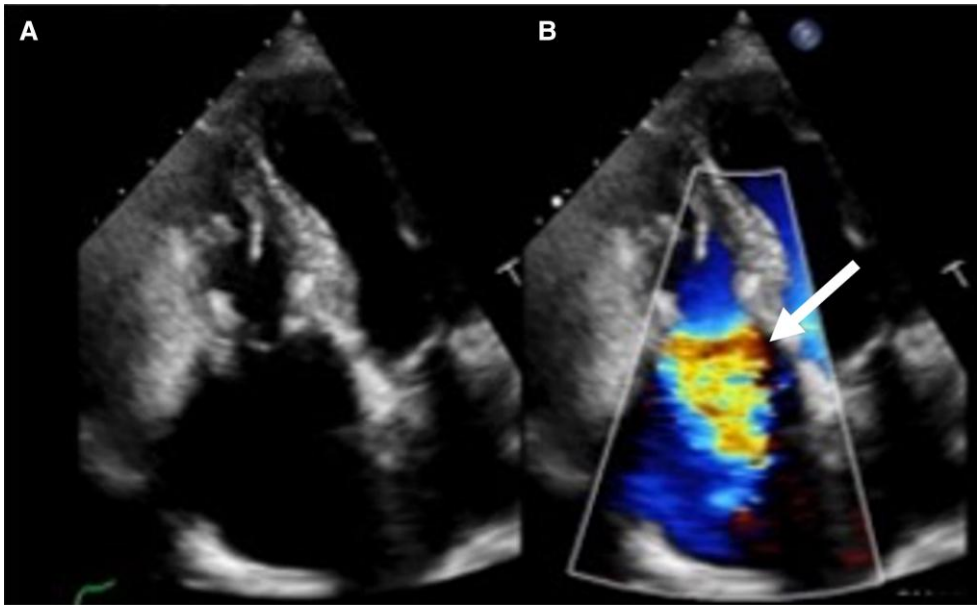


Figure 1 Apical four-chamber echocardiographic view demonstrating severe tricuspid regurgitation (see white arrow) pre-intervention without (*A*) and with colour flow (*B*) across the TV.

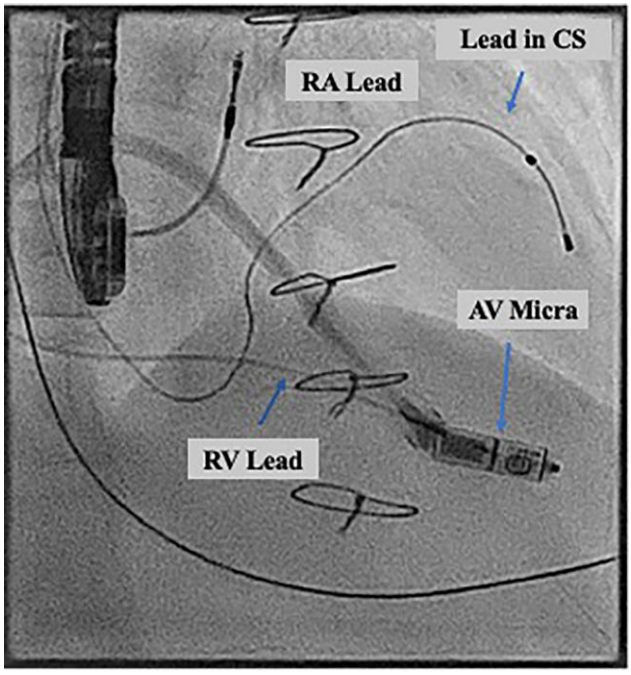


Figure 2 Leadless pacemaker (atrioventricular Micra) positioning and deployment. The existing right ventricle, right atrium, and coronary sinus pacing wires along with the transoesophageal echocardiography probe are also shown. AV, atrioventricular; CS, coronary sinus; RA, right atrium; RV, right ventricle.

using leadless pacemaker implantation, extraction of the existing pacing system, and percutaneous tricuspid VIV replacement with the use of intraoperative transoesophageal echocardiography (TOE).

With ultrasound guidance, femoral venous access was obtained and upsized to a 26F GORE® Dry seal sheath (WM Gore, USA). Utilizing the delivery kit, the Micra AV™ PPM (Medtronic Inc., Minneapolis, MN,

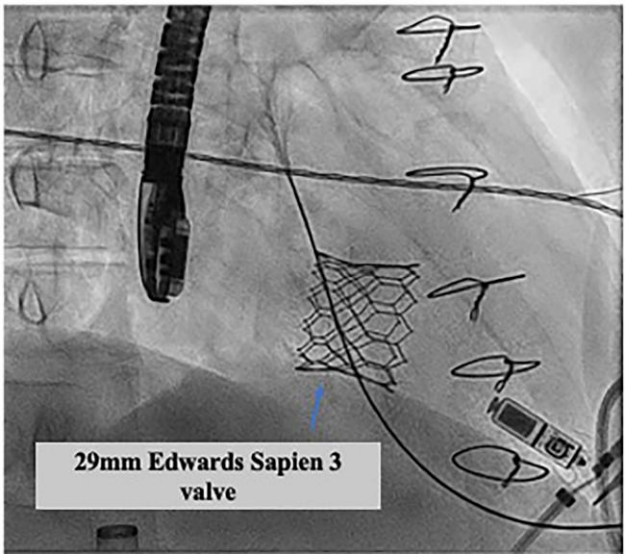


Figure 3 Leadless pacemaker (atrioventricular Micra) and tricuspid valve-in-valve replacement.

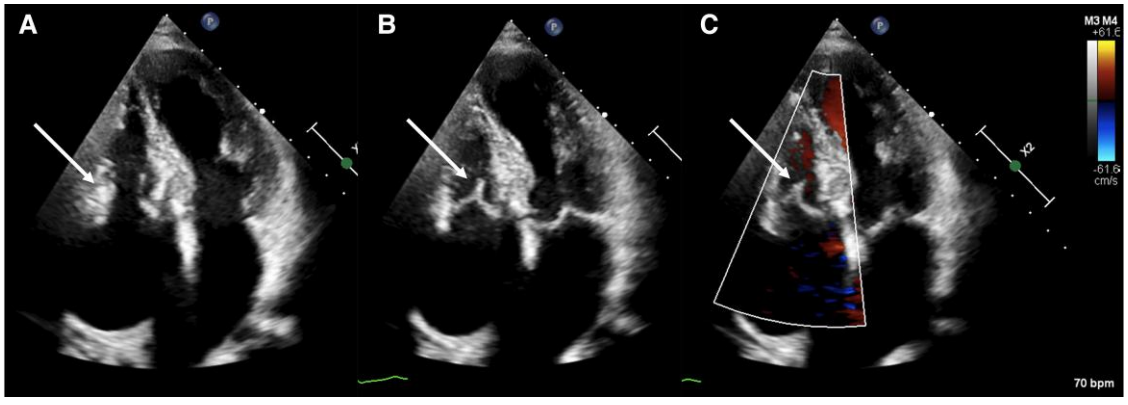


Figure 4 Apical four-chamber echocardiographic view demonstrating new valve-in-valve tricuspid valve in diastole (*A*), systole (*B*) and with colour flow across the tricuspid valve valve in systole (*C*) with no significant tricuspid regurgitation seen (see white arrow).

USA) was first advanced across the TV and positioned in the RV apex (*Figure 2*). Prior to deployment, standard testing and stability tests were performed (see Supplementary Material online, *Video S1*). The device was then deployed and parameters confirmed. The transvenous device was programmed off, and she was paced via the leadless system.

The transvenous system extraction began by making an incision over the device, removing the generator, and freeing the leads. Using in order, gentle traction, locking stylets, and Evolution cutting sheaths (Cook Medical, USA), the RV, atrial, and CS leads were fully extracted. The pocket was closed in layers. Transoesophageal echocardiography demonstrated no complications following extraction, but the TR remained severe despite removal of the pacing lead. As agreed, we then proceeded to TV replacement.

An Amplatz Super Stiff Guidewire (Boston Scientific, USA) was deployed in the distal right pulmonary artery, and then a 30 mm Cristal balloon (Balt extrusion, Montmorency, France) was inflated across the valve for sizing. Next, a 29 mm Edwards Sapien 3 valve (Edwards Lifesciences Inc., Irvine, CA, USA) was advanced and deployed satisfactorily (see Supplementary Material online, *Video S2*) across the TV. Fluoroscopy demonstrated satisfactory valve position (*Figure 3*). This was further confirmed with TOE demonstrating a well-seated replacement with no TR. Femoral venous closure was achieved with the Perclose ProGlide™ (Abbott Vascular, Redwood City, CA, USA) and a Z-stitch.

Post-procedurally, the NOAC was switched to long-term aspirin. Initial follow-up at 3 months demonstrated a significant improvement in her symptoms and a well-seated TV on TTE with no TR and a valve

gradient of 4 mmHg. We were unable to achieve AV synchrony despite reprogramming the AV Micra, and it is currently programmed VVIR with the only issue being mild breathlessness at peak exertion at the gym. At 12 months, she remains well, still going to the gym regularly and TTE confirming a well-seated valve with a trace of TR (mean gradient 5.2 mmHg) (*Figure 4*).

Discussion

The TV abnormalities found in EA contribute to TR leading to right atrial enlargement with true right AV junction dilatation.[4,5] In up to 80% of EA patients, an interatrial communication can be observed.[6] Right atrial volume overload due to TR, with an interatrial communication, can lead to right to left shunting.[5,6] Typically, EA patients present with symptoms such as breathlessness or cyanosis related to the effects of significant TR, RV dysfunction, and right to left shunting.[1]

Tachyarrhythmias are common in EA patients due to accesory pathways (AP) on the posterior and septal aspects of the true right AV ring.[1,5] The high incidence of AP were attributed to the structural changes, but this is not completely understood yet. Ventricular tachycardia has also been observed originating from the atrialized portion of the RV where the tissue is abnormal.[5] Structural and haemodynamic abnormalities with EA predispose to atrial arrhythmias long term.[4,5,7]

Bradyarrhythmias are less common and tend to occur following intervention on the valve.[1,8] A small proportion of EA patients (3.7%) require permanent pacing predominantly for AV block.[9] Following TV replacement, the development of complete AV block necessitating PPM is increased (13%).[1,8] In patients with prosthetic TVs, leads can be positioned epicardially, via the CS or sutured outside the prosthesis to avoid placing a lead across the valve in order to reduce lead induced TR.[10]

Whilst leadless technology has made significant advancements and is able to provide AV synchrony using devices such as the AV Micra, this may not be successful in every patient and was not achievable in our patient.

The management in EA includes TV replacement or repair, ablation of AP, and the repair of associated defects.[1] Echocardiography can usually elucidate the extent of TV regurgitation and feasibility for repair.[4] The ESC recommends (Class 1C) TV repair for severe symptomatic TR in EA with closure of the ASD/PFO at the time of surgery if expected to be haemodynamically tolerated.[2] Surgical repair is recommended irrespectively if there is progressive right heart dilatation or reduced RV function (Class IIc).[2] In cases where the anatomy is unfavourable, TV replacement is recommended. During TV replacement, caution is taken due to the anatomical relationship to the AV node and right coronary artery (RCA)[1] with the true annulus marked by the RCA, which is at risk of kinking during plication annuloplasty procedures.[4] Following surgical intervention for EA, the TR severity typically decreases immediately, but over time, progressive recurrences of TR is common, which may require re-intervention.[11] Re-intervention rates did not differ between those who underwent TV replacement vs. repair.[12] Utilizing a totally percutaneous approach, we have safely achieved a good outcome for the patient and avoided re-sternotomy.

Conclusion

In conclusion, the EA is a challenging condition to manage in view of the degeneration of the TV over time and the need for pacing due to the nature of the surgical intervention. Whilst surgical re-intervention with re-sternotomy carries considerable risks,[13,14] a percutaneous approach involving leadless PPM and VIV intervention is a potential good strategy for re-intervention in the EA patient.

Lead author biography

I am currently a cardiology trainee with a subspecialty interest in cardiac electrophysiology and devices. I currently work at Liverpool Heart and Chest Hospital. I graduated from the University of Manchester in 2008 and was awarded my PhD from the University of Manchester in 2021.

Supplementary material

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

Acknowledgement

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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.

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Data availability

The data underlying this article are available in the article and in its online supplementary material.

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