

Systemic ventricular implantation of a leadless pacemaker in a patient with a univentricular heart and atrioventricular node calcification



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Introduction

The development of a fully encapsulated leadless pacing system with active fixation provides an alternative treatment option for patients requiring pacemaker implantation. To date it has predominantly been used in patients with structurally normal hearts.¹ There is limited experience regarding patients with congenital cardiac disease. Since patients with complex cardiac lesions are surviving into adulthood, incidences of conduction disturbances will continue to increase.

The implantation of transvenous endocardial pacemakers in this patient cohort may be complicated by the anatomical substrate, venous access, and thromboembolic considerations. Epicardial systems are frequently employed, but concerns exist regarding their longevity and the risk of lead fracture. The development of a leadless Micra Transcatheter Pacing System (Medtronic, Minneapolis, MN) may overcome some of these challenges. To date, the short-term safety and efficacy of the leadless device has been established in a large adult population with structurally normal hearts.¹ Minimal data exists on the use of the Micra system in patients with congenital heart disease, with only 1 reported case in a patient with single-ventricle physiology.^{2,3}

We present a patient with Ebstein anomaly, palliated with a Blalock-Taussig shunt, who developed late-onset complete heart block secondary to atrioventricular node calcification (Figure 1). He was managed with leadless pacemaker implantation into the systemic ventricle.

Case report

We present the case of a 46-year-old man with Ebstein anomaly, resulting in functional tricuspid atresia. The rudimentary

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KEY TEACHING POINTS

- Patients with complex congenital cardiac lesions require special consideration when deciding on the optimal pacemaker system. Implantation of a leadless pacemaker with good thresholds is feasible in patients with univentricular physiology.
- The use of multiple imaging modalities helps to guide safe pacemaker implantation and optimization of thresholds.
- Anticoagulation prophylaxis is essential in patients with endovascular pacemaker systems and an obligatory right-to-left shunt, although many of these patients may be treated prior to implantation.

right ventricle (RV) was hypoplastic with severe tricuspid regurgitation (Figure 2). An atrial septal defect was present with right-to-left shunting. He was originally palliated with a classic right Blalock-Taussig shunt at age 3 (1976). Shunt occlusion gradually occurred and, at age 44, a left modified Blalock-Taussig shunt was performed.

A severe stenosis in the right pulmonary artery was stented at the hilum in 2016, through the left modified Blalock-Taussig shunt. During follow-up review, atrial tachycardia was noted in late 2017 and 15 months later he developed complete heart block with junctional rhythm with rates of 50–60 beats per minute. Cardiac magnetic resonance imaging was performed, and severe fibrotic infiltration of the heart was reported.

Concerns were raised regarding the risk of thrombus formation from endocardial lead contact and friction with the myocardium and venous system, with a potential high risk of embolization into the systemic circulation through the obligatory right-to-left shunt.

With limited published data with regard to the risk of embolization in conventional pacing vs leadless pacing, we

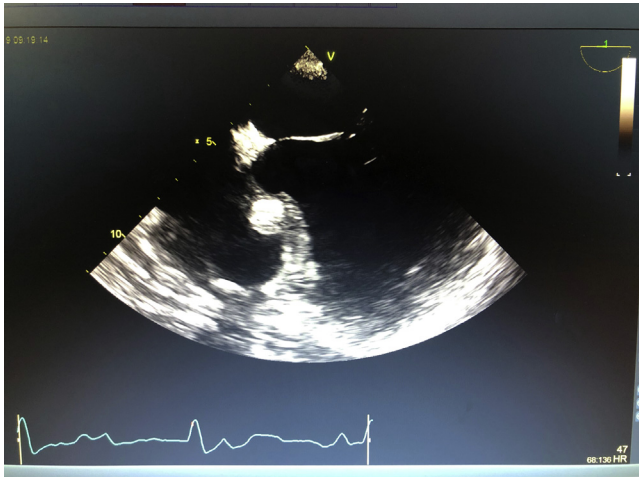


Figure 1 Transesophageal echocardiography demonstrating nodular calcification.

could only speculate that the overall risk may be lower with a leadless system, where the risk of lead friction is removed.

A surgical epicardial dual-chamber pacemaker was also considered but was declined by the patient and surgeon secondary to extreme erythrocytosis (hemoglobin of 27 g/dL) and resultant bleeding and thrombotic risk.

The decision was made to implant the device in the left ventricle (LV) owing to severe fibrosis in the RV. We believed the LV would likely have more viable myocardium for engagement of the tines and better threshold measurements. We recognize the lack of data on the technicalities of implanting a leadless device in the less trabeculated LV. There is also the potential risk of embolization within the systemic ventricle. The patient was carefully counseled on all options available with a full and open discussion. The patient declined a surgical approach and was keen to proceed with leadless pacemaker implantation.

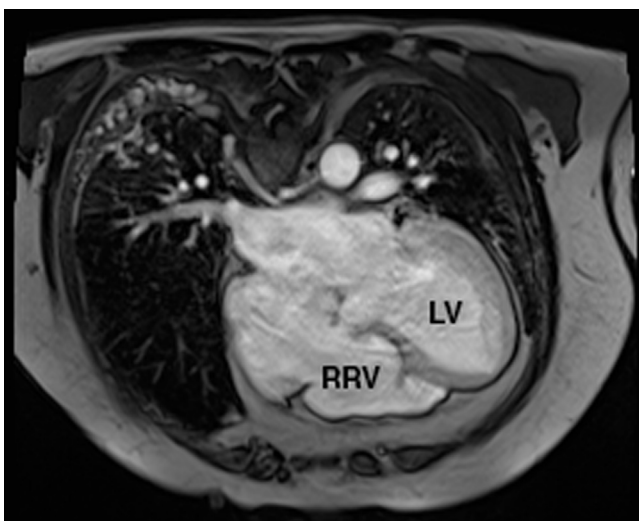


Figure 2 Representative images from cardiac magnetic resonance imaging highlighting the complex congenital anatomy of the rudimentary right ventricle and left ventricle.

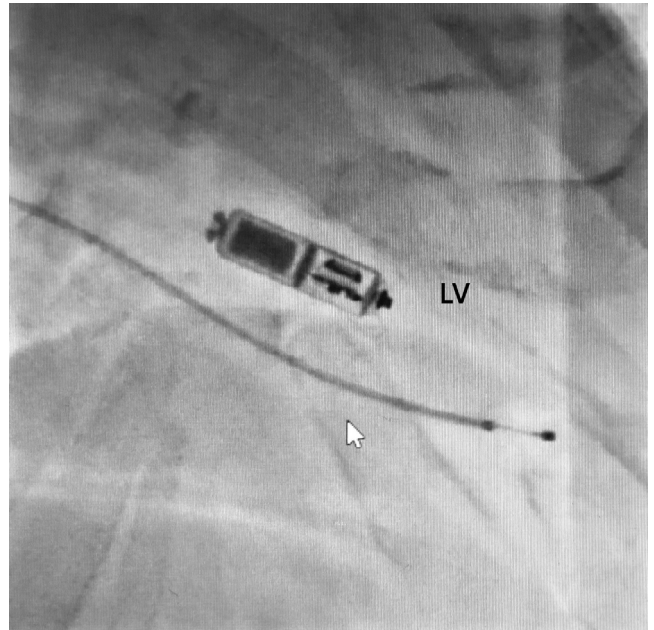


Figure 3 Micra (Medtronic, Minneapolis, MN) implant position in left ventricle with temporary wire in the rudimentary right ventricle.

The procedure was performed under general anesthetic to provide for transesophageal echo (TOE) guidance. The procedure was carried out using fluoroscopy and TOE. TOE showed an atrial communication with right-to-left shunt and hypoplastic RV. A 12-mm-diameter calcified mass was noted close to the atrioventricular node ([Figure 1](#)).

The right groin was then prepared and punctured, and two purse string sutures placed. The femoral vein was dilated to 23 French and the Micra introducer inserted. The Micra delivery system catheter was easily passed through the septal defect into the left atrium and further to the LV. During manipulation he developed prolonged asystole, and a temporary wire was introduced into the rudimentary RV via the left femoral vein.

The device was positioned in the left ventricular apex with initial high threshold noted. The device was captured and re-deployed into the rudimentary RV with adequate separation to the tricuspid valve, but high thresholds were again present.

The device was passed across the atrial septum a second time with implantation into the left ventricular apex. Pull test showed good engagement of the tines and satisfactory parameters were achieved. Pacemaker check revealed an R wave of 7.1 mV, threshold of 0.75 V @ 0.24 ms, and impedance of 980 ohms. The device was released and rechecked for stability of the parameters.

The delivery system was removed with purse-string closure and effective hemostasis. The patient was recommenced on rivaroxaban anticoagulation following the procedure and remains well. Follow-up threshold of the Micra pacer was 1.13 V @ 0.24 ms ([Figure 3](#)).

Discussion

We have demonstrated that implantation of a leadless pacemaker with good thresholds is feasible in patients with

univentricular physiology. It is possible that the use of this device may increase as patients with congenital heart disease survive longer, with an increased incidence of conduction disturbance secondary to disease progression and multiple interventions.⁴

Patients with congenital heart disease and rhythm disturbances are a challenging patient group owing to their complex anatomy and embolic risk. Conventional pacing with either epicardial or endocardial systems for patients with univentricular physiology may be suboptimal owing to endovascular thrombosis risk and need for multiple re-do procedures. The leadless device avoids the risk of lead fracture, lead displacement, and endovascular thrombus formation with venous occlusion. Owing to the short pulse width and battery technology, the longevity of the device may exceed conventional devices, resulting in fewer revisions.^{1,5}

Intracardiac thrombus risk is not eliminated with this device, and concern still exists regarding the potential for systemic emboli and stroke in patients with univentricular physiology. Anticoagulation prophylaxis is essential in all patients with endovascular pacemaker systems (including leadless pacemakers) and an obligatory right-to-left shunt in order to minimize this risk. There are no data supporting the use of one anticoagulant over another. Our patient was already managed with rivaroxaban, which we continued post implantation.

The use of multiple imaging modalities may guide safe pacemaker implantation and was recently advised in a case report by Ferrero and colleagues.² In our patient, TOE provided visual and spatial awareness, which was crucial to the success of the implant.

Conclusion

Implantation of a leadless device within the systemic ventricle is feasible in patients with complex cardiac anatomy. Long-term thrombotic risks in this context have not been described and these patients will require close follow-up. Leadless pacing offers an alternative approach and may have advantages in specific patients.

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