Leadless pacemaker implantation in a subpulmonic left ventricle in a patient with congenitally corrected transposition of the great arteries

Ravi S. Shah, MD,* William R. Davidson Jr., MD,* Soraya M. Samii, MD, PhD*

*From the *Heart & Vascular Institute, Penn State Health, Milton S. Hershey Medical Center, Hershey, Pennsylvania.*

Introduction

Leadless pacemakers are designed for implantation in a subpulmonic, morphologic right ventricle in patients with normal cardiac anatomy. In selected scenarios the subpulmonic ventricle may be a morphologic left ventricle. We present a case of a 38-year-old male patient with congenitally corrected transposition of the great arteries (ccTGA) with complete heart block who developed a pocket infection after implantable cardioverter-defibrillator (ICD) implantation. He had significant superior vena cava (SVC) stenosis, making vascular access for future lead placement difficult. An AV Micra™ (Medtronic Inc., Minneapolis, MN) leadless pacemaker was successfully implanted in his subpulmonic left ventricle.

KEY TEACHING POINTS

- Patients with congenitally corrected transposition of the great arteries (ccTGA) are predisposed to developing heart block.
- Patients with ccTGA and conduction abnormalities have a unique circumstance where device leads must be implanted in a morphologic left ventricle.
- Implantation of a leadless pacemaker in a morphologic but subpulmonic left ventricle was shown to be feasible.

Case report

The patient is a 38-year-old man with ACHD-AP (Adult Congenital Heart Disease Anatomic and Physiologic) class IIIC adult congenital heart disease with ccTGA and dextrocardia, born with a malalignment conal ventricular septal defect, membranous ventricular septal defect, infundibular and pulmonic stenosis, subvalvular aortic stenosis, and double outlet right ventricle. Initial corrective surgery at age 8 included aortic homograft placement from the subpulmonic left ventricle to pulmonary artery and closure of ventricular septal defects. He developed complete heart block during the surgery and had a permanent transvenous ventricular pacemaker placed prior to discharge. He developed severe tricuspid insufficiency with right systemic ventricle dysfunction as well as left subpulmonic ventricle–to–pulmonary artery conduit stenosis. This required reoperation and placement of mechanical tricuspid and pulmonary valves at age 16. He also had placement of a dual-chamber epicardial pacemaker system at that time and explantation of prior transvenous system. Two years later, he required a new atrial lead, resulting in creation of a hybrid pacemaker with a transvenous atrial lead and epicardial ventricular lead. There was debate about designating his anatomy as mesocardia or dextrocardia, but imaging reliably showed a cardiac apex pointed rightward, consistent with dextrocardia.

Eight years later, at age 26, the patient underwent pacemaker generator change for elective battery replacement indication. Six years later, at age 32, he underwent another generator change owing to reaching elective battery replacement indication. Four years later, at age 36, he had nonsustained polymorphic ventricular tachycardia and malfunction of the epicardial right ventricular pacing lead with intermittent lead noise. With this lead fracture, he was referred for device revision. He had developed a severely dilated systemic right ventricle with poor function, yet remained in NYHA class II heart failure and was working full-time. He was referred for implantable cardioverter-defibrillator (ICD) placement owing to a single, asymptomatic run of polymorphic ventricular tachycardia, nonischemic cardiomyopathy, and complete heart block. He had a single-chamber transvenous ICD implanted on therapeutic warfarin, given his

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Address reprint requests and correspondence: Dr Ravi Shah, Heart & Vascular Institute, Penn State Health, Milton S. Hershey Medical Center, 500 University Dr, Hershey, PA 17033. E-mail address: Rshah6@pennstatehealth.psu.edu.

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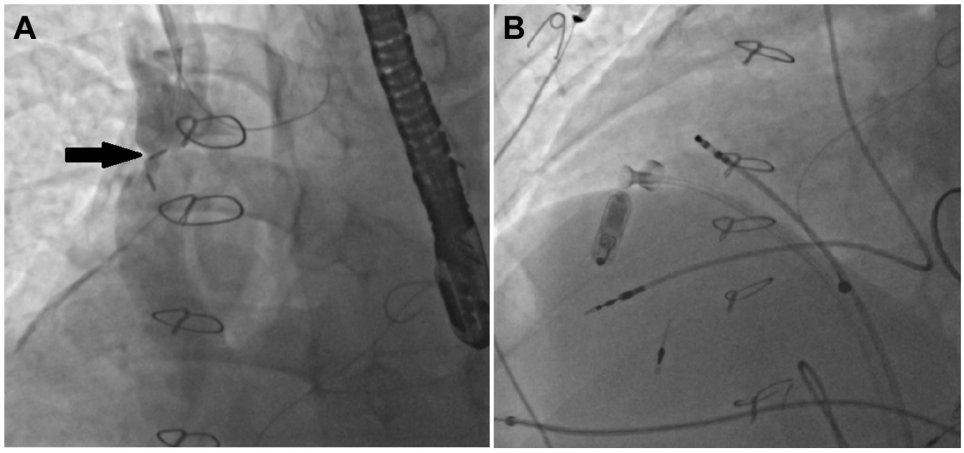

Figure 1 A: Left anterior oblique (LAO) 30° view showing significant superior vena cava stenosis (*black arrow*), as seen on venography during implantable cardioverter-defibrillator lead extraction. B: LAO 20° view showing the AV Micra (Medtronic Inc., Minneapolis, MN) deployed against the left ventricular septum without difficulty in securing its position.

mechanical valves. The ICD lead was anchored to the left ventricular, subpulmonic endocardium. This procedure was prolonged owing to difficulty advancing the lead through the SVC and manipulating the lead in the left ventricle. Given the prolonged procedural time, a TYRX antimicrobial pouch was used to reduce his risk for infection. The permanent pacemaker was left in place, as he was on therapeutic anticoagulation (international normalized ratio 2.5) during the case. He unfortunately presented with an infected ICD pocket 3 months later.

The patient had explantation of the transvenous ICD system and pacing system using a 12F laser and 11F cutting sheath. The permanent pacemaker was also removed because the course of the atrial lead was superimposed to the infected pocket. Evidence of SVC stenosis at the right atrial junction was confirmed during the extraction (Figure 1A). A temporary-permanent pacer was placed via the right internal jugular vein through the SVC for a period of 4 days until cultures were confirmed negative while the patient was on intravenous antibiotics. The significant SVC/ right atrium stenosis presented a therapeutic challenge for placement of future transvenous leads, with risk of SVC syndrome.

A decision was made to implant a leadless pacemaker, and not a transvenous ICD, given difficult vascular access, pacemaker dependence, and recent infection. Although there was risk of pacing-induced cardiomyopathy, dual-chamber

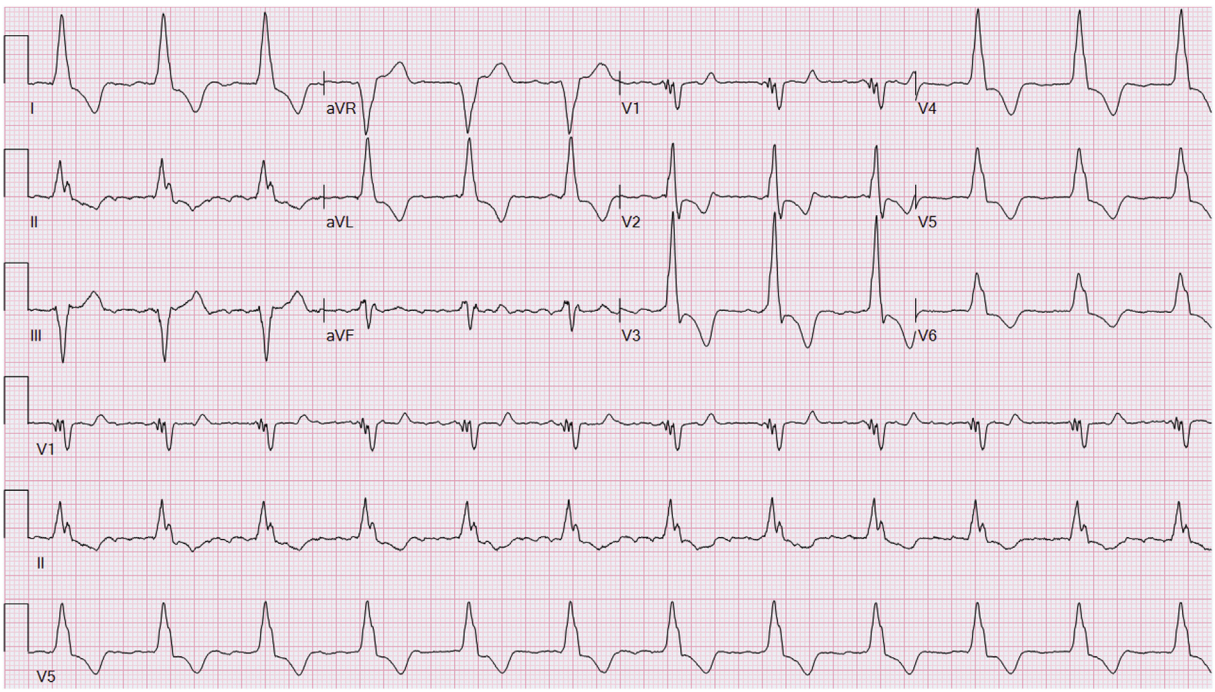

Figure 2 Electrocardiogram post Micra (Medtronic Inc., Minneapolis, MN) implant showing a ventricular-paced rhythm with underlying atrial fibrillation.

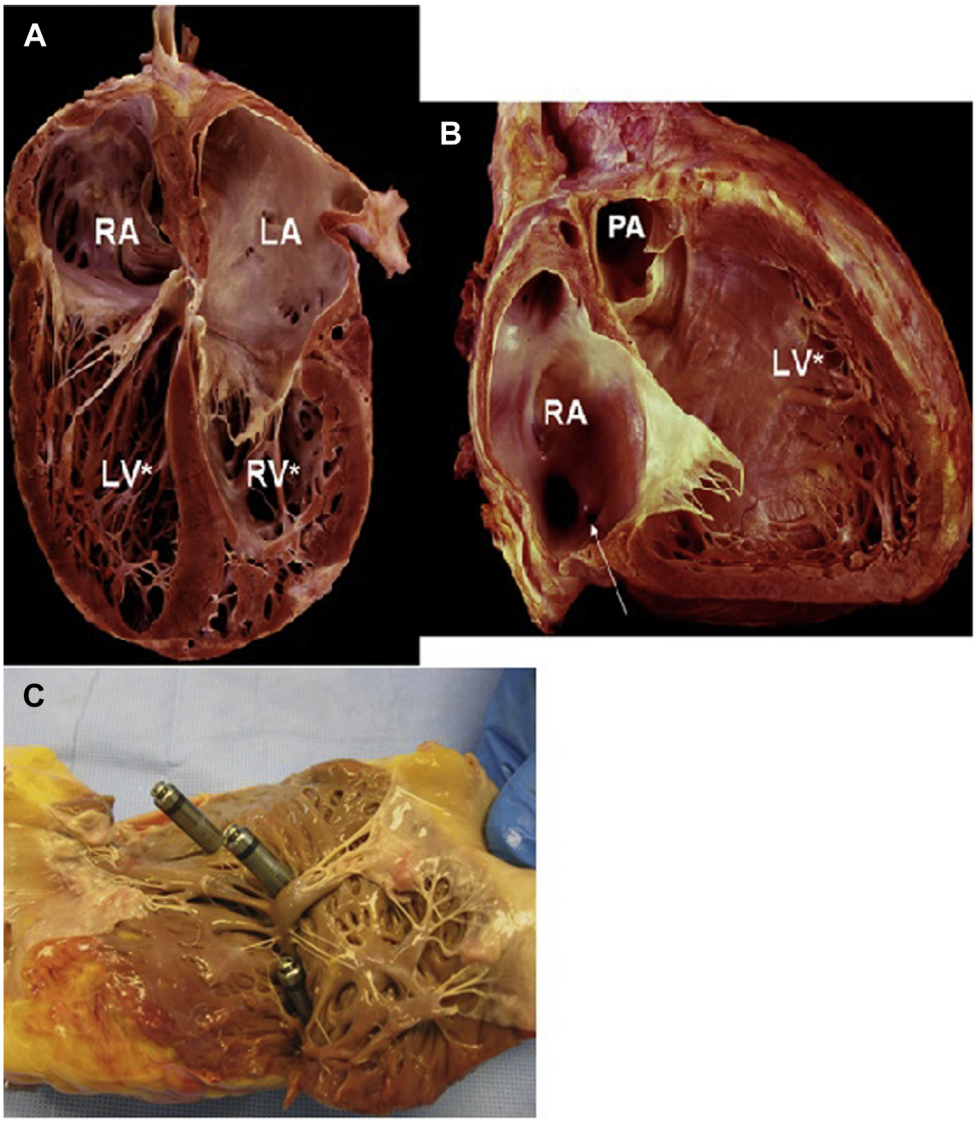

Figure 3 Anatomic section from a patient with congenitally corrected transposition of the great arteries (**A** and **B**) compared to a normal heart (**C**). **A:** Four-chamber view with the right atrioventricular valve located cephalad to the atrioventricular valve. **B:** Long-axis view with the arrow pointing to the coronary sinus ostium. **C:** Typical trabeculation in a morphologic right ventricle with multiple Micras (Medtronic Inc., Minneapolis, MN) implanted. CS = coronary sinus; LA = left atrium; LV = morphologic left ventricle; PA = pulmonary artery; RA = right atrium; RV = morphologic right ventricle. Used with copyright permission.[14,15]

pacing was not an immediate option and an AV Micra was chosen. The procedure was performed on therapeutic warfarin, given his 2 mechanical valves. A quadripolar catheter was placed in the subpulmonic left ventricle for back-up pacing via the left femoral vein. An 8F sheath was placed in the right femoral vein and upsized to a 27F Micra sheath. Heparin (5000 units) was administered. The sheath was maneuvered from the right atrium to the left ventricle. The Micra was positioned against the left ventricular septum (Figure 1B). Multiple "tug" tests were performed and at least 3 tines were secured to the endocardium before the device was deployed. The mode was set to VVIR with threshold 0.3 V, pulse width 0.2 ms, and resistance 710 ohms. There were no issues with capture or attachment (Figure 2).

The patient was later screened for a subcutaneous ICD, but was deemed not to be a candidate owing to large T waves in the paced complexes. A transvenous ICD was not actively reconsidered, as the patient did not have new sustained ventricular tachycardia on pacemaker monitoring, nor did he have syncope or cardiac arrest.

The patient has had limited follow-up since Micra implant due to no-showing appointments. Remote device interrogation 6 months postimplant revealed normal device function without any episodes of ventricular arrhythmia and 98% pacing. Pacing and sensing thresholds were similar to 1 month postimplant measurements.

Discussion

Adult congenital heart disease patients with complex anatomy often have conduction abnormalities requiring hardware implantation for pacing and defibrillation. From our review,

this is only the second reported case of a Micra leadless pacemaker implanted in a patient with dextrocardia and the first in a morphologic left ventricle serving as the subpulmonic ventricle. In this case, the patient has congenitally corrected transposition (l-transposition), but patients with d-transposition of the great vessels after atrial baffle surgery may also have similar issues with venous stenosis and pacing needs. There is 1 prior report of Micra implantation in the morphologic left ventricle in a patient with a univentricular heart.[1]

Patients with ccTGA have discordant atrioventricular and ventriculoarterial anatomy. ccTGA is a rare form of congenital heart disease, with an incidence of 1 in 33,000 live births.[2] The position of the atrioventricular node(s) and course of the bundle branch fibers often vary with patients' unique anatomy in ccTGA.[3] As a result, these patients are known to have abnormal atrioventricular node function and up to half may develop AV block, with about 30% developing complete heart block. In a series of 107 patients, there was a 2% per year risk of progressing to complete heart block after diagnosis.[4]

In addition to having complex congenital heart disease, our patient is one of many who developed complications from an implantable device. Implantable pacemakers and implantable device complications are well studied. About 12% of patients experience complications within 2 months, and pocket-related complications account for about 5% of complications.[5] A possible solution for patients who are poor candidates for transvenous systems are the evolving leadless devices. Leadless pacemaker systems are expanding in their scope and ability to provide right and left ventricular endocardial pacing in adults and children.[6,7] Micra systems have not been associated with significant risk of infection and have an overall lower complication rate than conventional pacemakers.[8] Leadless pacemaker retrieval, either Nanostim or Micra, has been shown to be both feasible and safe.[9] Leadless pacemaker placement in unconventional scenarios has also been described and is important to consider in complex patients.[10]

The Micra Transcatheter Pacing System was designed for implantation in the right ventricle, with tissue penetration properties characterized from human hearts. Device implantation testing was performed in the right ventricular apex and right ventricular outflow tract in preclinical trials.[11] In this present case, the pacemaker was implanted in the morphologic left ventricle secondary to ccTGA. There has not been an evaluation of differences in pacing thresholds and fixation characteristics for left ventricular endocardial pacing for Micra devices. There are differences in the embryology and gene expression between the left and right ventricle, reflected as differences in histology, anatomy, and conduction properties.[12] In patients with normal anatomy, in comparison to the left ventricle, the subpulmonic right ventricle has thinner myocardium and is more compliant. The right ventricle also has coarser trabeculae than the left ventricle (Figure 3). Left ventricle endocardial pacing via transvenous leads has been shown in limited studies to be feasible and effective.[13] In this case, the tine deployment of the Micra TPS behaved the same way as a typical right ventricular device placement. Unfortunately, the patient was not a candidate for a transvenous ICD or subcutaneous ICD at this time.

Patients in the congenital heart disease population often have complex anatomy and complex medical histories that make the use of alternative pacing systems attractive. Further research needs to be done to fully characterize attachment and conduction properties of leadless pacemakers in morphologic left ventricles. However, this experience with implanting a Micra transcatheter pacemaker in a subpulmonic, morphologic left ventricle may allow us to consider leadless pacing therapy in patients with ccTGA or in d-transposition post atrial switch, where the left ventricle serves as the subpulmonic ventricle.

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