

Leadless pacemaker placement in an 18-kilogram child: Procedural approach and technical considerations



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Introduction

Permanent cardiac pacing in pediatric patients can be challenging secondary to patient size and associated congenital cardiac anomalies. Leadless pacemakers were developed to avoid lead-related complications associated with transvenous and epicardial pacemaker systems, and they provide an alternative approach in the pediatric population. To date, the Micra Transcatheter Pacing System (TPS) (Medtronic, Minneapolis, MN)¹ is the only available Food and Drug Administration–approved system. Herein we describe a case of a leadless pacemaker implant in an 18-kilogram pediatric patient.

Case report

An 8-year-old, 18-kilogram male child (height of 120 cm), with a history of congenital central hypoventilation syndrome (CCHS) and Hirschsprung disease related to a heterozygous 27 polyalanine repeat mutation (20/27) in exon 3 of the *PHOX2B* gene, was referred for pacemaker placement. His past medical history included tracheostomy placement with ventilator dependence, multiple abdominal surgeries, and numerous peripherally inserted central catheters (PICC) throughout his lifetime. He presented to an outside hospital with a small bowel obstruction and underwent an emergent open adhesiolysis. During his hospital course he developed episodes of severe bradycardia with ventricular rates of 20 beats per minute, as well as sinus arrest with pauses lasting 15 seconds, requiring cardiopulmonary resuscitation. He was transferred to our institution for placement of a pacemaker. Of note, prior to transfer, a PICC was placed in his right brachiocephalic vein for total parenteral nutrition.

KEYWORDS Bradycardia; Leadless pacemaker; Pediatric cardiology; Sinus pause; Transcatheter pacing system
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Pacemaker implantation was recommended owing to severe bradycardia with a high likelihood of future malignant events.² After informed parental consent, the patient was taken for attempted transvenous dual-chamber pacemaker placement. The patient was placed under general anesthesia with inhaled sevoflurane. A left peripheral venogram demonstrated complete occlusion of the left subclavian vein with an extensive venous collateral network. Percutaneous access was attempted from the left axillary vein, but a wire could not be advanced through the collateral network. Given his recent abdominal surgery and elevated risk with an epicardial system, as well as his continued need for a PICC, excluding the possibility of a right-sided transvenous implant, a Micra TPS was discussed with both the patient's family and the cardiothoracic surgical team.

Ultrasound evaluation demonstrated that his femoral veins measured 7 mm in diameter, while his right internal jugular (RIJ) vein measured 12 mm in diameter. Given the large size of the Micra TPS delivery sheath (ie, 27F, or 9 mm in diameter), the opportunity for vascular complications was felt to be excessive from a femoral vein approach and an RIJ approach was preferred. A surgical cut-down was elected in order to perform a controlled venotomy with improved hemostasis. An incision was made in the supraclavicular region over the medial head of the sternocleidomastoid muscle, with subsequent division of the platysma muscle and separation of the 2 sternocleidomastoid heads. The patient was next heparinized with a 100 mg/kg bolus. The RIJ was encircled with #2 silk and tourniquets. Using the Seldinger technique the RIJ vein was accessed and an Amplatz Super Stiff wire (Boston Scientific, Marlborough, MA) was advanced to the right femoral vein. A transverse venotomy was made in the RIJ after snaring both proximally and distally, at which point the 27F Micra Introducer (Medtronic) was advanced to the level of the diaphragm under fluoroscopic guidance (Figure 1).

The dilator was removed and the sheath was positioned at the superior vena cava–right atrial junction. The Micra

KEY TEACHING POINTS

- Leadless pacing via a transcatheter pacing system can be used in small pediatric patients when traditional pacing systems are contraindicated.
- Transesophageal guidance can facilitate optimal leadless device positioning and allow monitoring of the tricuspid valve apparatus during implant.
- Preservation of vein integrity may require careful preoperative evaluation and, in some cases, use of a surgical cut-down approach.

Delivery Catheter (Medtronic) was advanced toward the right ventricular apex and then directed toward the septum with a buckle noted on the shaft of the delivery system. Position was confirmed by transesophageal echocardiogram (TEE). The leadless pacemaker was deployed and pacing and sensing characteristics were verified. A pull test was performed with widening of at least 2 nitinol wires, confirming stable position. Once the tether was cut and carefully removed (Figure 2), lead characteristics were reconfirmed, revealing an R-wave amplitude of 9.7 mV and pacing threshold of 0.38 V at 0.24 ms.

After completion of the implant, the 27F Micra Introducer was withdrawn and the RIJ repaired using a 6-0 polypropylene continuous suture. The remaining wound was closed in layers, with excellent hemostasis. The patient recovered in the pediatric intensive care unit, where a chest radiograph and echocardiogram were obtained on postoperative day 1 showing stable device position and no pericardial effusion. The Micra pacemaker was also interrogated; interrogation showed an R-wave amplitude of 10.5 mV and a pacing threshold of 0.75 V at 0.24 ms.

The patient remained in the hospital 2 days postoperatively prior to being discharged home. Follow-up at 6 months after device implantation showed an R-wave amplitude measuring 13.3 mV and stable pacing threshold of 0.5 V at 0.24 ms. Transthoracic echocardiogram at follow-up showed no evidence of tricuspid valve regurgitation.

Discussion

Safety and effectiveness of leadless pacemaker systems are well defined in adults with structurally normal hearts.^{3,4} The use of leadless pacemakers in the pediatric population remains limited,⁵ and to our knowledge, the 18-kilogram patient described in this report is the smallest patient to date to receive a leadless device.

The patient was affected by CCHS, which is most commonly the result of a heterozygous polyalanine repeat in exon 3 of the *PHOX2B* gene. The normal 20-alanine repeat is expanded to 24–33, with increased phenotype severity associated with larger repeats.⁶ More rarely, CCHS can be the result of a non-PARM variant in exon 1, 2, or 3, resulting in a missense, nonsense, frameshift, or stop codon mutation.⁶ Non-PARM variants are also commonly associated with more severe phenotype presentations. The present patient had a 20/27 PARM, which has previously been associated with an increased risk for prolonged asystole and is considered to be a risk of sudden death.^{2,7} Thus, given his presentation with 15-second pauses and higher-risk genetics, pacemaker implantation was warranted.

The decision to proceed with this implantation was a shared decision-making process between the patient's parents, the cardiothoracic team, and the pediatric electrophysiology team. Because of his history of multiple abdominal surgeries as well as recent small bowel obstruction with subsequent surgical adhesiolysis, he was thought to be a poor candidate for an epicardial system. A transvenous system was not possible owing to his left subclavian vein obstruction and a PICC in his right brachiocephalic vein. Given his need

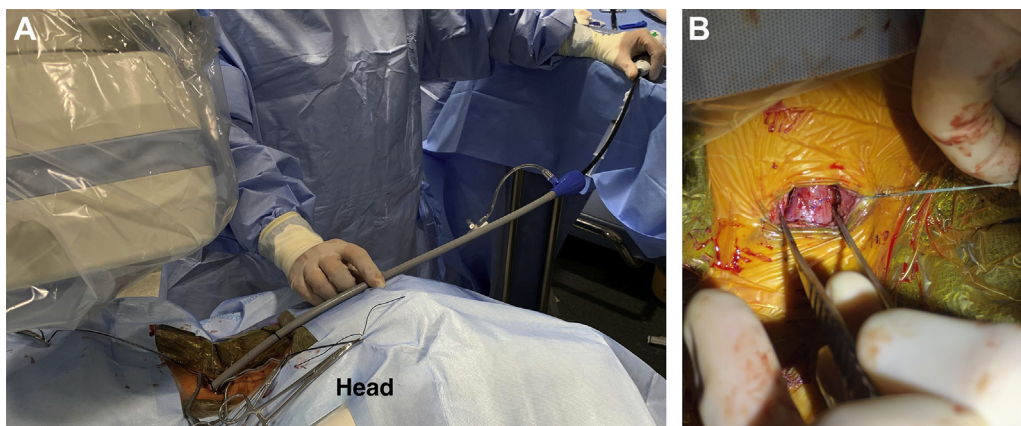


Figure 1 Micra (Medtronic, Minneapolis, MN) Introducer and Delivery Catheter set-up. **A:** Introducer was marked with black suture at level that reached the inferior vena cava. **B:** Close-up image of the right internal jugular vein after venotomy repair.

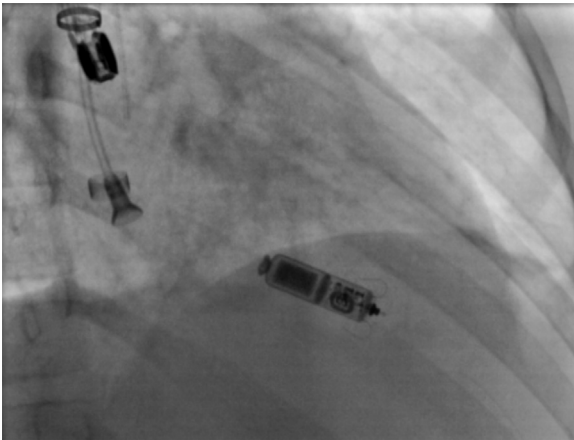


Figure 2 Anterior-posterior fluoroscopic image of the final Micra Transcatheter Pacing System (Medtronic, Minneapolis, MN) position after tether cord was cut and removed.

for only intermittent ventricular pacing, a leadless system was felt to be the best option. Possible complications, as described in the LEADLESS II study,⁸ were discussed with the parents. These include, but are not limited to, device dislodgement, cardiac perforation, vascular injury, and elevated pacing threshold at follow-up.

In preparation for the procedure, the case was discussed with the interventional cardiologists at our institution, who have employed an RIJ vein approach for Melody valves (Medtronic), which utilize a 22F delivery system, in children smaller than 18 kg. Given the size of our patient, we postulated an increased risk for vessel trauma; thus a cut-down approach was recommended as the safest approach for ensuring adequate hemostasis following the procedure.

The internal jugular approach also provided a more direct route to the body of the right ventricle and allowed for easy positioning on a septal location.⁹ However, the relatively straight angle from the internal jugular vein to the ventricle made the movement typically seen on the Micra Delivery

Catheter during the pull test less apparent than normally seen with femoral access. We relied more heavily on transmitted cardiac pulsations on the tether cord to confirm that adequate force had been applied, after which the cineangiogram was reviewed to evaluate the widening of the nitinol wires. A limited number of prior reports have described successful implantation of leadless pacemakers via a jugular approach.^{9,10} The largest series described 19 adult patients, mean age 77.5 ± 9.6 years, all of whom received a MICRA TPS by percutaneous internal jugular approach. All patients in this series were pre-closed using either 2 Perclose ProGlide sutures (Abbott, Santa Clara, CA), or a figure-of-8 stitch. A cut-down approach was not described in this series.

The need for a surgical preparation of the field limited us to the use of single-plane fluoroscopy during the procedure. TEE guidance was thus used to aid localization and positioning of the device on the septal aspect of the right ventricle, paying close attention to the relative position of the Micra TPS to the septal tricuspid valve leaflet, as well as to monitor the amount of tip pressure applied while delivering the Micra TPS. The final position of the Micra TPS was apical septal at a site distal to the tricuspid valve apparatus as a result of the small right ventricular chamber size (Figure 3). There was no evidence of tricuspid valve regurgitation by TEE immediately following implantation or at follow-up. The first site of delivery had optimal lead parameters and a stable position; thus it was decided against repositioning to a higher septal site. To date, the lead parameters have remained stable in follow-up.

Conclusion

In this case we describe the implantation of a leadless pacemaker in an 18-kg patient by internal jugular vein approach. To our knowledge, this is the smallest patient implanted with a Micra TPS to date. His vascular anatomy and noncardiac medical needs made him an unsuitable candidate for traditional pacemaker systems, and his

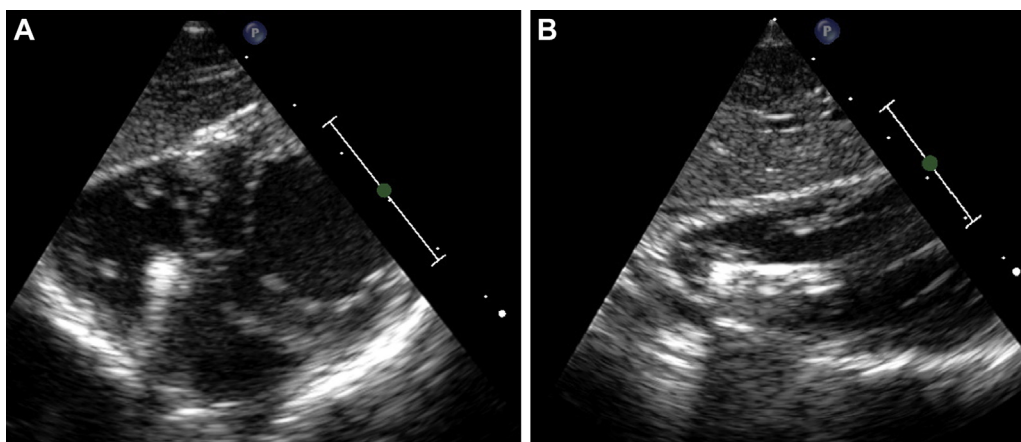


Figure 3 Transesophageal echocardiogram, short axis (A) and long axis (B), depicting apicoseptal position of the Micra Transcatheter Pacing System (Medtronic, Minneapolis, MN).

need for minimal pacing rendered him an ideal candidate for a leadless pacemaker system.

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