DEVICE THERAPY

COMPLEX CASE STUDY

Prolapse of Pacemaker Leads Resulting in Complete IVC Obstruction in an Adult Congenital Patient

STEVEN K. CARLSON, MD, PHILIP CHANG, MD, FHRS and RAHUL N. DOSHI, MD, FACC, FHRS

Keck School of Medicine of the University of Southern California, Los Angeles, California

ABSTRACT Cardiac implantable electronic devices (CIEDs) are increasingly being used in pediatric cardiac patients. Transvenous implantation offers advantages over an epicardial approach; however, lead displacement and/or dysfunction due to somatic growth remain a concern. Introduction of intravenous lead slack at implant has been used to allow for growth. We report a unique presentation of complications secondary to excessive retained lead slack. A 23-year-old patient with Down syndrome and surgically repaired complete atrioventricular canal defect underwent placement of a dual-chamber transvenous pacemaker at age 10 years, having had prior epicardial pacemaker support for postoperative complete heart block. She was referred for evaluation of abdominal and lower extremity swelling. Workup revealed liver cirrhosis. Imaging revealed prolapse of both transvenous leads into the inferior vena cava (IVC), resulting in complete obstruction at the level of the hepatic veins and large azygous collateralization. Complete device extraction was performed with subsequent dilation of the IVC to restore anterograde blood flow into the right atrium. This case highlights an unusual set of complications related to lead prolapse. Expected release of lead slack did not occur, likely due to overestimation of the potential growth of this patient with Down syndrome. This case exemplifies the potential for new and unusual CIED-related complications that electrophysiologists should be aware of while caring for the growing population of pediatric and congenital heart disease patients with CIEDs.

KEYWORDS adult congenital, liver cirrhosis, pacemaker extraction, venous obstruction.

Introduction

Cardiac implantable electronic devices (CIEDs) are now routinely used in the treatment of bradyarrhythmias and tachyarrhythmias in both pediatric and young adult patients with congenital heart disease. Transvenous CIED implantation offers several advantages over an epicardial approach; however, lead displacement due to growth of the child remains a concern. One method to compensate for somatic growth is the introduction of intravenous and endocardial lead slack. Failure of the lead slack to release, causing lead failure has been described.1 We report a unique presentation of complications secondary to excessive retained lead slack in a young adult with congenital heart disease.

Case report

A 23-year-old patient with a history of Down syndrome and surgically repaired complete atrioventricular canal defect underwent placement of a dual-chamber transvenous pacemaker at age 10, having had prior epicardial pacemaker support for postoperative complete heart block. She was referred for evaluation of recent-onset abdominal and lower extremity swelling. Physical examination revealed more than one lower extremity edema to the thighs and dullness to percussion over the abdomen. Laboratory data were significant for elevated liver function tests: AST (aspartate aminotransferase) 217,
ALT (alanine aminotransferase) 215, alkaline phosphatase 523, total bilirubin of 0.8, and albumin 2.1 U/L. A basic metabolic panel including renal function was normal. Abdominal ultrasound revealed a coarse liver parenchymal texture with intraluminal narrowing in the inferior vena cava (IVC) caudad to the insertion of the hepatic veins.

Echocardiogram showed normal left ventricular and right ventricular structure and function. No residual shunting in the region of membranous ventricular septum/interatrial septum. The IVC was noted to be small with collapse suggesting a right atrial pressure of 0–5 mm/Hg.

**Pacemaker interrogation**

Pacemaker interrogation consisted of a dual-chamber pacemaker in the DDD mode: 100% ventricular paced; atrial lead impedance 322; ventricular lead impedance 200 ohms (noted to be unchanged from previous interrogations). Atrial sensing at 0.1 mV, had decreased from 0.75 mV at the last interrogation. The sensing configuration of the atrial lead changed from bipolar to unipolar, which resulted in improved sensing. Ventricular lead sensing was not performed because of pacemaker dependence.

The patient was referred for transjugular liver biopsy. Imaging revealed prolapse of both the atrial and ventricular leads into the IVC (Figure 1). In addition, the retrohepatic vena cava demonstrated high-grade stenosis adjacent to the pacemaker leads with extensive intrahepatic collaterals from the hepatic vein into the infrahepatic IVC, which then drains to the right atrium via the azygos vein (Figure 2). Biopsy confirmed stage 4/4 cirrhosis.

The patient was subsequently referred for device extraction with IVC angioplasty. The initial venogram confirmed severe obstruction of the IVC (Figure 3). A temporary pacing wire was placed from the right jugular vein given pacemaker dependence. Laser lead extraction was performed with a 14 French powered laser sheath (Spectranetics, Colorado Springs, CO). The ventricular lead was extracted first. Progressive dissection freed the lead down to the IVC–RA (right atrium) segment, at which point the lead’s outer insulation broke, leaving only the inner coil and locking stylet for support and countertraction. The remainder of the lead was removed with primarily blunt dissection. The atrial lead contributed the most prominent prolapse into the IVC. At the IVC–RA junction, manual dissection with the outer sheath was primarily used to progressively peel the lead from the IVC wall. Eventually,
the most inferior portion of the loop was freed and the remainder of the lead was removed using a combination of laser pulses and outer sheath dissection.

Once the pacemaker system was fully extracted, a repeat venogram confirmed continued complete obstruction of the IVC (Figure 4). Balloon venoplasty was then performed across the obstruction initially with a 12 × 20 mm Admiral Xtreme OTW balloon (Medtronic Inc., St. Paul, MN), followed by a 12 × 60 mm balloon. A post-inflation venogram revealed restoration of blood flow from the IVC to the RA (Figure 5). Using the retained subclavian venous access, new atrial and ventricular leads were then implanted in the standard fashion.

The patient had a short stay in the intensive care unit after the procedure after she developed hypoxia and hypotension. The etiology was thought to be secondary to the showering of a small amount of debris to the lungs after extraction. However, the patient did well and was discharged without any long-term complications. A repeat abdominal Doppler ultrasound performed 2 months after the extraction showed a narrowed lumen of the IVC but with normal directional flow and Doppler waveform within both the IVC and the hepatic veins. A repeat venogram performed 6 months after extraction confirmed continued patency of the IVC (Figure 6).

Discussion

Advances and improvements in the care of pediatric and congenital heart disease patients, including the incorporation of CIED implantation, have greatly improved survival, with the vast majority of patients expected to live into adulthood. However, along with improved survival comes a growing population of adult patients with unique CIED-related complications. As such, it will be important for the adult cardiologist to be able to recognize the long-term sequelae of CIED placement in the adult congenital heart population.

The presented case highlights an unusual set of complications related to chronic and excessive transvenous lead prolapse. Venous obstruction developed, which was further complicated by congestive liver dysfunction and eventual cirrhosis. While the excessive lead slack and prolapse likely contributed to the observed progressive lead dysfunction, it also had more devastating consequences with indolent but progressive end-organ impact.

In infants and small children, an epicardial approach to lead implantation remains the most common strategy and was for a long period the only option for CIED implantation. This method is more invasive, requiring surgical dissection for lead fixation, and has been associated with potential for trauma, lower threshold values, poor lead longevity, lead fracture, and the need for surgical replacement or removal. The timing of

Figure 4: Venogram immediately after removal of leads. Complete obstruction remains.

Figure 5: Venogram after balloon dilation shows anterograde flow in the RA.

Figure 6: Venogram 6 months after removal of leads shows improved anterograde flow in the RA.
transvenous lead implantation in pediatric patients largely depends on patient size and the implanting physician’s experience and preference, as well as the presence of suitable anatomy in the case of patients with various forms of congenital heart disease. Although transvenous leads are generally preferred over epicardial leads if given a choice, the problem of lead displacement caused by somatic growth in the pediatric patient remains a concern. There have been several methods proposed to alleviate this problem, with most factoring the potential future growth of the patient and introducing a redundant intravascular lead to compensate for this growth. Strategies for placement of the redundant lead have been described. Rosenthal and Bostock reported success in an infant with the use of an atrial loop placed just after birth that then fully uncoiled at 32 months allowing for a delay in replacement. Gasparini et al. presented results from three children aged 8–11 years who received an ICD in which a redundant loop was successfully introduced into the IVC. At the 16-month follow-up they noted partial release of the loops with expectation of further release as growth continued. Patients remained free from clinical events and device parameters remained stable. Other strategies have been described as well, including attaching the lead on the septum allowing the tip to “drop down” as the heart grows at the apex. Some have proposed using absorbable sutures to secure the anchor sleeves, with the expectation that the additional lead will enter the vessel from the pocket with growth.

Problems related to excess lead placement have been reported. Berul et al. described 11 pediatric patients who had developed a lead prolapsed through the pulmonary valve due to excess lead placement. A case of pacemaker malfunction due to a prolapsed IVC lead has also been described previously. In that case, a 10-year-old boy with Down syndrome required lead replacement 5 years after implantation due to loss of capture of his single ventricular lead. Similar to our presented case, the lead was prolapsed into the IVC with attachment to the endothelium. He had not developed IVC obstruction, however. It is important to note that both this case and the previously reported case of IVC lead retention occurred in patients with Down syndrome. Children with Down syndrome follow significantly different growth curves, and mean final heights for both boys and girls are 2.5 standard deviations below that of normal children. It is possible that the amount of lead slack necessary was overestimated and thus the expected release was never achieved. Also, both complications occurred well after implantation (5 and 10 years) suggesting endothelial fibrosis is a long-term process. This case emphasizes the importance of close follow-up for children with CIEDs, especially during the growth phase. Periodic imaging and device interrogation is necessary to ensure proper release of the lead as the child grows. Special attention must be given to children with congenital disease in relation to both cardiac abnormalities and expected growth. Finally, while surveillance of lead slack and device function may be normal, it is also important to incorporate reasonable methods of evaluating venous patency and to exercise awareness of the possibility of impact on other organs. In the case presented, liver dysfunction secondary to chronic venous obstruction due to excessive lead prolapse served as the motivator for lead removal.

References