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Device Extraction in Adults with Congenital Heart Disease

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Abstract

Background—Device extraction is a critical component in the treatment of patients with device-related infections. Due to complex anatomic considerations, device extraction in adults with congenital heart disease presents unique challenges to the electrophysiologist.

Methods—Here, we present a series of device-extraction cases performed in patients with transposition of the great arteries status post either Mustard or Senning surgical procedures that subsequently had permanent pacemakers placed and ultimately developed device-related infections.

Results—All of these patients eventually underwent successful laser extractions of their infected devices resulting in complete removal of all hardware and resolution of their infections.

Conclusions—These cases illustrate that endovascular device extraction has been safely and effectively performed in adult patients with congenital heart disease, though further studies are needed to determine the procedural risks and success rates of this procedure in this patient population.

Keywords

congenital heart disease; device extraction; transposition of the great arteries; pacemakers; device infections

Introduction

Over the past several years, the indications for permanent pacemakers and implantable cardioverter-defibrillators (ICDs) have grown, resulting in a subsequent increase in the number of implanted devices.^{1,2} Unfortunately, this increase in the number of implanted devices has been associated with a marked increase in the rate of cardiac device infections.³ The exact incidence of device-related infections is somewhat difficult to define due to the variability in the definitions employed and frequent latency between device implantation and manifestation of infection.⁴ However, reported incidence rates of cardiac device-related infections have ranged from 0.13% to 19.9% for permanent pacemakers and from 0.0% to 3.2% for ICDs.^{5–7}

Adults with congenital heart disease also represent a growing population of patients.^{8,9} Arrhythmias are a common late complication in these patients. Sudden cardiac death is the most common cause of mortality in adults with congenital heart disease,¹⁰ most often as the result of ventricular tachycardia/ventricular fibrillation, but bradyarrhythmias are also prevalent in this patient population.¹¹ The prevalence of arrhythmias in adults with congenital heart disease has resulted in an increasing requirement for permanent pacemakers and ICDs. Due to their complex anatomy, vascular limitations, and intracardiac shunts, some adults with congenital heart disease require epicardial lead placement; however, successful transvenous lead placement is often possible in patients with congenital heart disease.^{12,13} Due to the relative youth of this patient population at the time of their cardiac device implantation, adults with congenital heart disease who require permanent pacemaker or ICD implantation are at risk for potential device- and lead-related complications that may eventually require device extraction. These patients often will require multiple device replacements or device upgrade surgeries that are associated with infection rates of 2–7%.⁴ Additionally, both younger age at the time of device implantation and the presence of congenital heart disease have been associated with an increased risk of lead failure.¹⁴

Even in patients with structurally normal hearts, laser lead extraction can be associated with a nontrivial risk of major complications.^{15–17} Though several small studies have been published, little is known about the feasibility of laser lead extraction in adults with congenital heart disease.^{13,18} Due to complex anatomic consideration in these patients, endovascular lead extraction in adults with congenital heart disease presents unique challenges to the electrophysiologist. This report describes a series of three cases where infected devices were completely removed from adult patients with transposition of the great arteries (TGA) who had undergone surgical repair via Mustard¹ or Senning² procedures and subsequently underwent permanent pacemaker implantation. These cases illustrate that in the right setting, endovascular lead extraction can be safely and effectively performed in patients with complex congenital heart disease.

Case Histories

Case 1

The first patient was a 24-year-old female with D-transposition of the great arteries. At age 10 months, she had undergone a Senning atrial switch palliative repair and initially did very well until age 16 when she had a syncopal episode. The patient was subsequently diagnosed with sick sinus syndrome and underwent permanent pacemaker placement 2 years later. Approximately 1 month prior to presentation to our center, the patient noted erythema, local swelling, and discomfort over her pacemaker pocket. Subsequently, she noted a bleb formation just superior to her pacemaker incision. Additionally, the patient noted frequent night sweats but denied fevers and rigors. The patient was initially started on a course of oral keflex without resolution of her symptoms. An ultrasound showed a fluid collection over the pacemaker. Reportedly, an attempt to aspirate this fluid was made, at an outside hospital, and yellow purulent fluid was withdrawn. Subsequently she was started on oral Ciprofloxacin and Bactrim for 1 week again without resolution of her symptoms.

The patient was admitted to our center with suspected pacemaker pocket infection in September 2005. The pocket was fluctuant and a thin blister with yellow discoloration was noted over an opening at the incision site with the pacemaker generator visible below the blister. Culture swabs of fluid expressed from this blister grew gram-positive cocci in clusters. The patient was noted to be relatively device dependent, and thus a temporary screw-in pacemaker lead was placed via the right internal jugular vein through the superior/inferior vena cava (SVC/IVC) baffle into the morphological left ventricle where it was actively fixated in place. This was attached to an old pacemaker pulse generator that was

secured to the patient's chest and remained in place until the patient subsequently underwent permanent pacemaker placement 3 days later.

A transesophageal echocardiogram (TEE) was done intraoperatively to rule out major vegetations on the leads prior to extraction and to help guide the extraction. No baffle obstruction was noted. The old pacemaker pulse generator had nearly eroded through the skin and was visible in the left deltopectoral groove (a similar device erosion is shown in Fig. 1). The pocket was opened and it contained evidence of liquefaction and necrosis. The capsule was completely excised. Cultures were obtained from the pocket but remained negative. The leads were dissected out using blunt dissection and electrocautery down to the suture sleeves that were cut and removed. The leads were cut and lead-locking stylets were placed in the central lumen of both leads. Then under fluoroscopic and TEE guidance, a Spectranetics Excimer Laser Sheath (Spectranetics, Inc., Colorado Springs, CO, USA) was slowly advanced, initially over the ventricular lead under fluoroscopic guidance. Exuberant scarring was noted around the leads, particularly at the region of venous entry and at the lead tip. Multiple applications of laser energy (Excimer xenonchloride, 308-nm wavelength) were delivered during advancement and the lead was removed without incident. Subsequently, the atrial lead was removed in a similar fashion with gentle countertraction on the lead and locking stylet. There was no hemodynamic compromise and the patient tolerated the procedure well. The pocket was primarily closed loosely and the skin was closed with staples to allow drainage. The patient underwent placement of a new dual-chamber pacemaker 3 days later, after blood cultures were negative for 48 hours, without incident. She remains stable and is doing well without any further signs of infection.

Case 2

This patient was a 27-year-old woman with diagnoses of D-transposition of the great arteries, ventricular septal defect (VSD), and sub-valvular pulmonic stenosis. The patient underwent a balloon septostomy as a newborn, followed by a Senning atrial switch procedure at 1 year of age. In 1993, the patient was referred to our center for resection of her subpulmonic stenosis, repair of her aneurysmal VSD, and left ventricular myomectomy with removal of a left ventricular outflow tract membrane. Additionally, an epicardial pacemaker was placed abdominally at that time due to history of sick sinus syndrome. This pacemaker was subsequently revised and placed in the left shoulder region with transvenous leads in 1999. In June of 2006, the patient noted soreness over her left upper chest area near her pacemaker incision site. In September of 2006, the patient was admitted to an outside hospital due to swelling over her implanted pacemaker. A fluid collection was noted over the pacemaker and this fluid was aspirated and 25 cc of turbid fluid were removed. Analysis of the fluid revealed 44,000 white blood cells with 93% neutrophils. Grams stain and cultures on this aspirated fluid as well as systemic blood cultures were negative. The patient was started on Daptomycin and transferred to our center for further care. The patient was found to be paced <1% of the time. Therefore, the pacemaker generator was removed and the pocket was thoroughly irrigated; however, the leads were not initially extracted given the negative cultures and the perceived difficulty of removing a chronic lead from across a baffle. The patient received a 3-month course of intravenous antibiotics (ceftriaxone and vancomycin) and was subsequently switched to oral antibiotics. Serial TEEs were performed, which did not demonstrate any obvious vegetations on the leads. The patient ultimately received an additional 3 months of oral Bactrim therapy. Within 3 weeks of completing the course of Bactrim, the patient called with recurrent tenderness, erythema, and warmth over her pacemaker site. Blood cultures were obtained; the patient was restarted on Bactrim, and admitted to our center for consideration of device extraction. Repeat blood cultures were negative and a repeat TEE revealed no obvious vegetations on the pacemaker leads.

The patient was taken to the operating room for extraction of the indwelling leads. When the leads were cut, a brownish pus-like material was expressed. This fluid was sent for culture and eventually grew out *Staphylococcus Aureus* that was sensitive to bactrim. A TEE was again performed intra-procedurally to rule out major vegetations along the leads and to guide lead removal. Using blunt dissection and electrocautery, dissection was performed along the leads down to the suture sleeves that were cut and removed and then the leads were freed down to the clavicle. Locking stylets (2 LLD, Spectranetics Inc.) were placed down both leads. Then, a 14-French Excimer Laser Sheath (Spectranetics Inc.) was advanced slowly over the lead under fluoroscopic guidance into the right atrium. However, dense calcifications encased the lead as it entered and coursed through the baffle. As we tried to advance the laser sheath into the baffle with countertraction on the lead and locking stylet, multiple locking stylets were broken. A Libertor stylet (Cook Medical, Bloomington, IN, USA) was then placed down the ventricular lead and we were able to slowly advance a larger, 16-French Excimer Laser sheath across the baffle. The ventricular lead was densely adhered to the baffle surface with heavily calcified scar along most of its course. Multiple laser applications were made within the baffle; however, this did not appear to greatly assist with lead removal. Eventually, the sheath was advanced across the entire baffle mostly with blunt dissection and countertraction on the lead. Once the sheath was in the ventricle, the lead was quickly extracted and removed. Next, the atrial lead was extracted using the same 16-French Excimer laser sheath. Both leads were completely removed and all remaining capsule was excised from the pacemaker pocket. The pocket was thoroughly irrigated with antibiotic solution and loosely closed. The skin was closed with staples to allow drainage from the pocket. The patient completed an additional 2-week course of Bactrim and has remained infection free and asymptomatic without the need for a new pacemaker since that time.

Case 3

The final patient was a 23-year-old male with a history of repaired complex congenital heart disease who was admitted to our center for endocarditis and possible valve replacement. The patient was diagnosed with D-transposition of the great arteries at birth. He subsequently underwent a balloon atrial septostomy in the first few days of life. At 9 months of age he underwent an atrial switch procedure. He did very well after this surgery and in general was a very active child. However, due to complaints of easy fatigability a Holter monitor was done at the age of 15, which revealed sinus bradycardia with ventricular rates as low as 35 beats per minute, and a transvenous pacemaker was subsequently placed. The pacemaker pulse generator was changed when the patient was 20 years old. In August of 2007, the patient noted an insect bite on his left hand. He went to a local emergency room and was noted to be febrile and was then transferred to a regional medical center. The patient's left arm was noted to be markedly swollen and Doppler ultrasound revealed a left subclavian thrombus. The patient was placed on intravenous heparin. Blood cultures grew out methicillin-sensitive *S. Aureus*. The patient's condition deteriorated and he required intubation and was placed on broad-spectrum intravenous antibiotics, including vancomycin and ciprofloxacin. The patient was transferred to our center for further evaluation and was placed on Oxacillin. The patient was not pacemaker dependent and interrogation of his pacemaker revealed that the ventricular lead was not capturing and the generator was near end of life.

The patient was referred for pacemaker extraction. Again, a TEE was performed intra-procedurally to rule out major vegetations along the leads and to guide lead removal. A small vegetation was noted on the mitral valve near the ventricular lead, baffles were patent, and no evidence of valve deterioration/perforation or destruction was noted. A 4-cm incision was made in the left deltopectoral groove over the old leads and pacemaker pulse generator.

Using blunt dissection and electrocautery, we dissected along the leads all the way down to the suture sleeves that were cut and removed and then the leads were freed all the way to the clavicle. Both leads were cut and a locking stylet was placed down the central lumen of each lead. Then, a 16-French laser sheath was advanced over the ventricular lead across the baffle and into the ventricle. Again, significant scarring was noted at the venous entry site and lead tip, requiring multiple laser pulses under the clavicle and in the ventricle near the lead tip. Eventually, the lead was freed and removed. Then, the laser sheath was advanced over the atrial lead without difficulty and the lead tip was freed. The lead started to unravel and broke as the active fixation screw reached the level of the clavicle. However, additional sutures were placed around this lead fragment and we were able to extract this remaining lead segment through the laser sheath. The capsule was completely excised and removed. The patient tolerated the procedure well. He received a full 6-week course of Oxacillin. The patient remains free of infection and has not required new pacemaker placement to date.

Discussion

Limited data exist regarding device extraction in adults with congenital heart disease. Friedman and colleagues reported their experience of device removal using interlocking stylets and dilator sheaths through the subclavian vein in 13 patients, eight of whom had congenital heart disease.¹⁹ Additionally, Cooper et al. retrospectively analyzed their experience of laser lead extraction in seven young ICD patients with congenital heart disease.²⁰ However, this series differed greatly from our cases in that a majority of their procedures were performed for lead fracture or failure. Similarly, Moak and colleagues reported their results on a mixed population of children and young adults undergoing lead extraction, again most commonly due to lead fracture.¹⁸ Finally, Khairy and colleagues recently reported a 5-year prospective registry of laser lead extraction at their center that included 16 adults with congenital heart disease.¹³ These previous studies in conjunction with the three cases presented here illustrate that device extraction can be safely and successfully performed in patients with congenital heart disease.

Adults with congenital heart disease present unique concerns to the cardiac electrophysiologist attempting device extraction in this patient population. First, due to complex anatomy and often previous surgeries, venous access to the heart can be nontrivial. Baffle stenosis can occur in patients with preexisting pacemaker or defibrillator leads. In some cases, baffle stenosis around pacemaker or defibrillator leads can necessitate lead extraction, stenting of the narrowed baffle, and reimplantation.^{21,22} Additionally, significant scarring and calcification can develop along the leads, especially as they course through the baffle. Such scarring and calcification may limit the efficacy of laser applications in some areas, and dissection through such regions requires great patience and care from the operating physician. This was seen in one of our cases but not the other two (as illustrated in Fig. 2). Overall, the use of laser sheaths to assist in lead extraction has greatly increased the safety and success rates of device extractions.¹⁷ Device extraction in adults with congenital heart disease is also greatly improved by using the application of laser energy; however, dense scarring and calcification can slow progress through certain areas and may even require careful blunt dissection with the laser sheath and countertraction on the lead. Resolution of infection almost always requires complete device removal including device generator, leads, suture sleeves, and capsule.

Our center routinely performs TEEs prior to or during our device extractions. We have found this to be particularly useful in adults with congenital heart disease for several reasons. Preprocedural TEEs have been useful in confirming the diagnosis of device infection and in defining large vegetations on the leads that may be a contraindication to percutaneous extraction, particularly in patients with patent foramen ovale. On several

occasions, though not reported in this case series, we have identified significant vegetations, in the presence of patent foramen ovale, not noted on trans-thoracic echo, that lead us to refer the patient for open extraction rather than an endovascular procedure, given the associated risk of cerebral emboli. Additionally, in patients with complex congenital heart disease the pre-procedural TEE can aid in delineating the anatomy and assist in defining the optimal access to the heart.

One final point illustrated by our case series is that not all patients with congenital heart disease and infected devices will require immediate re-implantation of a pacemaker or ICD. Only one of our three cases required re-implantation of a new pacemaker on the contralateral side during the same admission due to relative device dependence. The other two patients have been followed closely and still have not required device reimplantation. We have found that placement of a temporary screw-in lead attached to an externally placed pacemaker generator can be an extremely useful tool in patients who are relatively device dependent (See Fig. 3). We call this approach placement of a tempo-permanent device. This allows patients to receive stable back-up pacing on the floor and remain ambulatory while receiving intravenous antibiotics and awaiting clearance for reimplantation of a permanent pacemaker or ICD.

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Figure 1.

This figure shows an infected device that has eroded through the skin. This photograph was taken just prior to removal of the device. Part of the pacemaker lead and tie-down sleeve are clearly visible at the skin surface.

Figure 2.

Panel A depicts an image from a transesophageal echocardiogram (TEE) with the probe placed in the midesophagus and demonstrates a patent superior vena caval (SVC) baffle with pacemaker (PM) lead passing freely through the baffle. This image was obtained from case 3 in our series. Panel B depicts a TEE image from case 2 in our series. This TEE image was obtained with the probe in the midesophagus and demonstrates the pacemaker leads coursing along the baffle. The lead is noted to be adherent to the baffle at points along its course. We noted dense scarring throughout the baffle, making extraction difficult and time consuming in this patient. Panel C shows a PA chest X-ray from patient 2 that demonstrates the ventricular lead coursing through the systemic venous baffle. The lead was densely adhered to the baffle requiring multiple laser applications and careful blunt dissection with the laser sheath for successful removal.

Figure 3.

PA chest X-ray demonstrating placement of a “tempo-permanent” pacemaker. This system consists of an active fixation ventricular pacemaker lead attached to an old pacemaker pulse generator that was secured to the patient's chest with a Tegaderm dressing. This system remained in place with the patient receiving intravenous antibiotics until cultures were negative for 48 hours, at which time a new permanent pacemaker was placed endovascularly.