Salvage of an Epicardial Lead in a Pacemaker-Dependent Patient with Fontan Palliation Using an IS-1 Extender.

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Abstract

We present a case report of severed epicardial atrial lead salvage using an IS-1 lead extender. A 37-year-old male with single ventricle physiology, Fontan palliation, sinus node dysfunction, recurrent atrial tachycardias and atrial fibrillation resulting in failing Fontan physiology presented with failure of the atrial pacing lead. The patient was initially paced with an epicardial system that had to be removed due to pocket infection, and the epicardial leads were cut and abandoned. Given his significant sinus node dysfunction he required atrial pacing to allow for rhythm control. The failing Fontan physiology of the patient precluded him from undergoing surgery for epicardial lead placement or a complex intravascular lead placement procedure (although anatomically feasible). We considered the option of salvaging the existing epicardial atrial leads to provide atrial pacing, allowing for rhythm control and improvement of his failing Fontan physiology as a bridge to a more permanent pacing solution. This case report is important because it demonstrates how a lead extender can be used to salvage a severed pacemaker lead. This may be useful for patients in whom implantation of new leads is not promptly feasible due to patient anatomy and/or clinical status.

Introduction

Sinus node dysfunction and the need for atrial pacing is common in patients with Fontan palliation.[1-3] Pacemaker implantation in patients with Fontan palliation is challenging as, with modern Fontan procedures, the venous circulation is directed to the pulmonary arteries with a conduit, bypassing the systemic atrium.[2-5] This anatomy makes placement of a pacing lead via the typical venous routes nearly impossible. Epicardial leads are often placed at the time of the Fontan procedure to manage the sinus node dysfunction that is common in these patients. However, when the pacing lead(s) fail, the options to provide atrial rate support are very limited. We present a case report of a severed epicardial atrial lead that was salvaged using an IS-1 to IS-1 lead extender. The patient's worsening failing Fontan physiology precluded him undergoing a fifth time re-do sternotomy for epicardial lead placement or a complex intravascular lead placement procedure. The aim of this case report is to show how lead extenders can be used to salvage severed pacemaker leads in patients that need pacing and in whom implantation of new leads is not promptly feasible due to anatomical limitations and/or clinical status.

Case Report

Patient History

The patient is a 37 year-old man who was born with double inlet left ventricle, right ventricular hypoplasia, and left transposition of the great arteries with the aorta arising from the outlet chamber. At the age of 2 months old, he underwent a staged single ventricle repair that included initial pulmonary arterial banding. At age 6.5 years an atriopulmonary Fontan palliation with patch closure of the right-sided atrioventricular valve and suture closure of the atrial septum was performed. The right atrium was directly anastomosed to the main pulmonary artery (classic Fontan). At age 21 he developed recurrent and symptomatic episodes of atrial

tachycardia. He failed several catheter ablation attempts and, despite multiple cardioversions, maintenance of sinus rhythm was not feasible. Other than the aforementioned well-tolerated atrial tachycardias, he did remarkably well after his Fontan procedure until age 28 when he developed decreasing exercise tolerance, recurrent atrial dysrhythmias, and ascites: all findings concerning for failing Fontan physiology. At age 30 he underwent cavo-pulmonary Fontan revision using a non-fenestrated, intracardiac, 20 mm Gore-Tex conduit connecting the inferior vena cava to the pulmonary artery. During this operation he also underwent a surgical maze procedure with radiofrequency lesions placed from the free right atriotomy across the atrial septum to the coronary sinus, and from the atrial pulmonary connection to approximately the 10 o'clock location of the tricuspid valve patch. Additionally, surgical cavo-tricuspid isthmus ablation with lesions placed from the right atrioventricular valve patch at 4 o'clock to the inferior vena cava orifice was performed. His rhythm in the months preceding his Fontan revision was alternating between atrial tachycardia and sinus bradycardia with junctional escape consistent with sinus node dysfunction. To manage his bradycardia, a dual chamber epicardial pacemaker system was implanted at the time of the Fontan revision. Two sew-on coaxial bipolar epicardial atrial leads (Medtronic, 35 cm, 4968 CapSure Epi) were placed along the right border of the right atrium and two epicardial screw-in bipolar leads were placed on the inferior wall of the right ventricle. The leads were tunneled to the right upper quadrant of the abdomen and placed in a subcutaneous pocket where the best of each of these two leads was chosen and connected to a dual-chamber pacemaker. The unused leads were capped and left in the pocket. Unfortunately, two months after implantation he developed a pacemaker pocket infection with methicillin-sensitive*Staphylococcus aureus* for which the pacemaker was extracted. The epicardial leads were unable to be removed with counterclockwise rotation and thus were pulled gently and then cut flush to the wound edges allowing the lead remnants to retract to the depth of the wound. A vacuum-assisted drainage system was inserted into the wound and he was placed on the appropriate antibiotic regimen. His heart rate alternated between normal sinus with rates of 60-70 with junctional rhythm 40-50 and thus a decision was made not to pursue re-implantation of a pacemaker.

He continued to experience recurrent atrial tachycardias, as well as atrial fibrillation, and was managed medically with verapamil and therapeutic anti-coagulation. His atrial fibrillation burden increased, but due to sinus node dysfunction his antiarrhythmic regimen was unable to be escalated. The lack of organized atrial contraction and atrioventricular synchrony resulted in a significant deterioration of his functional status and recurrent exacerbations of failing Fontan physiology as evidenced by right heart failure and worsening of protein losing enteropathy. The patient required 6 hospitalizations over the course of 4 months for diuresis and management of his worsening protein-losing enteropathy. There was an immediate need to establish adequate atrial pacing support that would allow for cardioversion, restoration of AV synchrony and escalation of anti-arrhythmic medications. The clinical challenge was that the patient was too sick (from failing Fontan physiology) to undergo a fifth re-do sternotomy for placement of a new epicardial pacemaker system or percutaneous transvenous lead implantation. Transvenous access to his atria would require a complex procedure involving puncture of the Fontan conduit to access atrial tissue or puncture of the left pulmonary artery floor with placement of a lead into the right atrium, and the patient was deemed a high-risk candidate for the prolonged duration of anesthesia that would be required for such a procedure. As a result, an attempt was made to salvage the existing atrial leads. The endpoint of the procedure was to establish atrial pacing and AV synchrony in the short term in order to stabilize the patient enough to improve his clinical status and candidacy for a more invasive lead implantation procedure. The team worked together with the atrial lead manufacturer engineers to assure the feasibility of lead salvage. A shared-decision was made between the clinical team and the patient to attempt to salvage the existing epicardial atrial leads by using an IS-1 to IS-1 lead extender.

Lead Salvage Procedure

Prior to the procedure, prophylactic Cephazolin and Vancomycin were administered intravenously. The patient was brought to the electrophysiology laboratory in a post-absorptive state. The right upper quadrant and right sub-costal areas were inspected under fluoroscopy and the location of the previously cut and abandoned leads was marked. Following infiltration with 1% lidocaine/0.5% bupivicane mixture, an incision was made at the right upper abdominal area at the location overlaying the tip of the previously cut and

abandoned leads. Using sharp dissection, cautery and blunt dissection, the incision was extended down to the rectus abdominalis fascia, and a pocket was then fashioned caudad and sized to the device. The leads were exposed in the sub-diaphragmatic area. Hemostasis was obtained using cautery.

The two coaxial bipolar atrial epicardial atrial leads were exposed: lead #1 was longer and exposed about 3 cm in the pocket and lead #2 was shorter and exposed about 2.5 cm in the pocket. The distal 3 mm of outer insulation was removed from the end of the cut lead and the outer conductor coil (anode) was identified. The inner coil was not visible in this preparation and considering the short length of the lead a decision was made not to cut the lead more proximally. The lead was inserted, with the anode exposed, into the female end of a bipolar IS-1 lead extender (Oscor, BIS/BIS, 17.4cm, quadrifilar, silicone-covered, connector) such that the conductor coil passed through to the end of the receptacle, passing through both the anodal and cathodal set screws. Both set screws were tightened sufficiently to obtain contact onto the outer conductor coil, but not tightly enough to crush or damage the coil. Good unipolar sensing was confirmed (patient was in atrial fibrillation). Subsequently the patient was cardioverted with a 200J synchronized biphasic shock from atrial fibrillation into junctional rhythm with retrograde atrial conduction and ventricular far-field sensing from Lead #1. Pacing was performed using a unipolar polarity, and capture thresholds were acceptable (1.25 mV at 0.4 ms). The lead extender / prior assembly was then tied with two 0-silk sutures and silicone surgical adhesive was used to seal the lead extender / lead assembly at the lead insertion site. The same process was attempted for Lead #2 but during traction there was a major insulation breach noted on this lead which rendered it unusable for pacing (right atrial capture occurred only at very high thresholds with constant and forceful abdominal muscle capture). Lead #2 was re-capped and abandoned. The pocket was inspected for foreign bodies, irrigated copiously with normal saline solution and inspected again for hemostasis. The lead extender IS-1 pin was inserted into a single chamber pacemaker device, the screws were tightened, and the lead extender was tugged to be sure the connection was secure. The generator was inserted into the pocket with excess lead behind it. The pocket was then closed in multiple layers of absorbable suture, steri-strips were applied, and a sterile dressing was placed over the incision. After implantation the unipolar right atrial lead impendence was 152 Ohms, sensed P-wave was 1.0 mV, and capture threshold were 1.25 V at 0.4 ms. The pacemaker was programmed as AAIR 80-120 bpm. The patient was started on Sotalol 80 mg twice daily and was discharged home in stable condition on post-operative day two. He was seen as an outpatient in follow up one week after discharge. He was in an atrially paced rhythm with consistent atrial capture and intact atrioventricular conduction. His functional status had significantly improved with an obvious decrease in abdominal girth, peripheral edema and improvement of his protein and albumin levels. Approximately 4 months after his procedure, his pro-BNP had decreased from 941pg/mL on the day before the procedure to 102pg/mL. His albumin had increased from 1.5g/dL to normal at 4.6g/dL and his total protein increased from 3.1g/dL to normal at 6.7g/dL. All of this was achieved with routine outpatient Fontan management. This was the longest period of time which the patient had spent out of the hospital in the prior year. His future care plan is to pursue durable atrial pacing (new epicardial or transvenous leads) as he continues to improve by maintaining atrial contraction and atrioventricular synchrony (and thus operative/anesthesia risk improves).

Discussion

We present this case report of a severed epicardial atrial lead salvage using an IS-1 lead extender in a 37-year-old patient with Fontan palliation, sinus node dysfunction, recurrent atrial tachycardias and atrial fibrillation resulting in failing Fontan physiology. By salvaging the existing epicardial atrial leads we were able to provide atrial pacing that allowed for rhythm control and marked improvement of his failing Fontan physiology. In this case report we demonstrate how a lead extender can be used to salvage a severed pacemaker lead in patients in need for pacing, in whom implantation of new leads is not promptly feasible due to patient anatomy and/or clinical status. Although our initial intent was to use the IS-1 extension in a bipolar configuration, we were able to retrieve only the anode of the existing leads. We also show that an IS-1 lead extender can be used even if only the anode of a severed lead can be recovered, with adequate unipolar sensing and pacing.

A unipolar extender would have worked in this case as well, however we had planned to attempt a bipolar salvage and did not have a unipolar extender available, Note that with these epicardial leads, the lead body is coaxial bipolar in construction, however it bifurcates into what are essentially two unipolar electrodes that are individually attached to the myocardium (Figure 1C). Although one electrode is used as the anode and electrically connected to the ring of the IS-1, in our salvage procedure we utilized this electrode as the pacing cathode. Yet another option is to use an LV-1 to IS-1 lead adapter. This had the benefit of a much smaller opening at the female end of the adapter, allowing for a better size match between the lead body and the adapter.

The technique that we present here allows for potential salvage of pacemaker leads and avoidance of surgery or complex invasive procedures. Risks of this technique include increased susceptibility of unipolar leads to oversensing and/or adventitious pacing of the diaphragm, chest wall or abdominal muscles. In fact, the insulation breach that we caused in our effort to salvage Lead #2 resulted in forceful abdominal muscle contractions at the energy output required for atrial capture. Furthermore, the long-term durability of the repair that we present here is unknown. For this reason, we do not recommend this technique for patients who are pacemaker-dependent, nor as a sustainable solution for any patient. However, it can be a useful technique for patients whose clinical condition precludes them from being candidates for new lead implantation, and whose condition is expected to improve over time.

The risks, benefits and alternatives must be weighed and presented to all patients in a shared decision-making process. For our patient the alternative to lead salvage was implantation of new epicardial or transvenous leads. However, considering his failing Fontan physiology the risks of these procedures, especially prolonged positive pressure ventilation in the setting of Fontan physiology, were substantial. Due to the brevity of the procedure, the risk of lead salvage was considerably lower, while the benefits could be immediate and significant. We also anticipated that the failing Fontan physiology, which was the main contributor to his operative risk, would improve with rhythm, and atrial pacing and re-establishment of AV synchrony. Last, we want to highlight the important role that the lead manufacturer had in this decision making. Engineers of the lead manufacturer reviewed the lead model, radiographic pictures of the leads, as well as the available connectors and reassured the clinical team that such a repair is feasible.

With improvement in surgical techniques, an increasing number of patients with single-ventricle physiology palliated with Fontan procedures survive to adulthood.[6, 7] The median 30-year survival for patients that have undergone Fontan palliation is 43%, (N=1,052).[8] 90% of patients with Fontan palliation who have survived to age [?]16 will survive at 30 years and 80% at 40 years of age (N=683).[6] Patients with Fontan palliation commonly require pacemaker implantation (6-13% of cases[2, 6]), and in some cohorts pacemaker insertion or revision is the most common reason for re-operation (up to 20% of cases).[8] The most common indication for pacemaker implantation is sinus node dysfunction (64% of cases).[2] Extensive and multiple surgeries involving placement of intra-atrial baffles contribute to sinus node dysfunction and the need for atrial pacing.[1-3] Patients with intracardiac Fontan and double-inlet left ventricles (such as the patient presented in this case report) are more likely to need a permanent pacemaker.[2] Other characteristics associated with higher risk for need of pacemaker implantation are mitral atresia, double outlet right ventricle and heterotaxy syndrome.[2]

Delivering atrial pacemaker leads in patients with Fontan palliation is challenging since, with modern Fontan procedures, the venous circulation is directed to the pulmonary arteries with a conduit, bypassing the atrium. Atrial leads can be delivered epicardially with surgical approaches or, in select cases of intra-cardiac conduits, transvenously with complex interventions that involve puncture of the Fontan conduit.[2-5] Case reports are available that describe transvenous lead implantation in patients with extracardiac Fontan conduits.[4, 5, 9, 10] Any attempt for transvenous lead delivery in patients with Fontan palliation requires an experienced operator with an in-depth understanding of the patient's anatomy and the details of previous surgeries. A discussion on transvenous lead delivery in patients with Fontan palliation is beyond the scope of this case report.

Conclusions

We presented a case report of severed epicardial atrial lead salvage using an IS-1 lead extender, in a 37-

year-old patient with Fontan palliation and failing Fontan physiology. Lead extenders can be used to salvage severed pacemaker leads in patients for whom implantation of new leads is not promptly feasible due to their anatomy and/or clinical status. Bipolar IS-1 lead extenders can be used even if only the anode of a severed lead is recoverable, with adequate unipolar sensing and pacing. As the long-term durability of the repair that we present here is unknown we do not recommend it for patients who are pacemaker-dependent, or as a durable solution for any patient, but rather as a temporizing intervention.

Key Teaching Points

- Sinus node dysfunction and the need for atrial pacing is common in patients with Fontan palliation, but pacemaker implantation is challenging as the venous circulation is directed in the pulmonary arteries bypassing the systemic atrium.
- Lead extenders can be used to salvage severed pacemaker leads in patients for whom implantation of new leads is not promptly feasible due to their anatomy and/or clinical status.
- Bipolar IS-1 lead extenders can be used even if only the anode of a severed lead can be recovered, with adequate unipolar sensing and pacing.
- Lead salvage with IS-1 lead extenders is not recommended for patients who are pacemaker-dependent, or as a durable solution for any patient, but rather as a temporizing intervention.

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Figures

Figure 1. 2-view chest X-ray and electrocardiogram of the patient prior to the procedure. **(A)** Chest X-ray shows the severed epicardial leads. There are 2 bipolar atrial and 2 ventricular screw leads in place. There is mild cardiomegaly, trace right and small left pleural effusion. Postsurgical changes of median sternotomy wires.**(B)** electrocardiogram shows atrial fibrillation with normal ventricular response, right axis deviation and non-specific ST-T wave changes. **(C)** Example of the CapSure epicardial leads. CapSure epicardial leads, have a lead body that is coaxial bipolar in construction, however it bifurcates into what are essentially two unipolar electrodes that are individually attached to the myocardium (Figure C source: Medtronic Academy website. Reproduced with permission from Medtronic).

Figure 2. Snapshots during the procedure. **(A)** Surgical exposure of the two severed epicardial atrial bipolar leads. One 0-silk suture was used to secure the tip of each lead. **(B)** The atrial lead has been cut and prepared to be inserted in the IS-1 lead extender. Only the anode was retrieved in this preparation. **(C)** Surgical adhesive glue used to seal off the ends of the lead extender / lead assembly. **(D)** The lead extender / lead assembly is completed and ready to be connected to the generator. **(E)** The Oscor, BIS/BIS, quadrifilar, silicone-covered, lead extender with the bipolar receptacle and bipolar connector, as well as the set screws annotated.

Figure 3. 2-view chest X-ray and electrocardiogram of the patient after the lead salvage procedure. **(A)** Chest X-ray shows a single-chamber pacemaker implanted at the right upper quadrant. The pacemaker is connected to one of the atrial epicardial leads via the BIS/BIS lead extender (red arrow). The other leads remain in same position. No visible pneumothorax. Minimal bilateral pleural effusions, mild cardiomegaly, postsurgical changes of median sternotomy wires, unchanged from the pre-procedure films. **(B)** electrocardiogram shows atrial pacing with consistent atrial capture and intact atrioventricular conduction, right axis deviation and non-specific ST-T wave changes.