

Transvenous approach to pacemaker lead implantation for sinus node dysfunction after extracardiac lateral tunnel Fontan conduit placement



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Introduction

Sinus node dysfunction (SND) affects 9% of patients with single-ventricle physiology that are palliated via the Fontan procedure, and pacemaker implantation has become the most common cardiac procedure in this population.^{1,2} It has been shown that in addition to the increased procedural risk inherent to open surgery, the epicardial approach is associated with higher pacing thresholds, lower sensing thresholds, and faster battery depletion compared with transvenous (TV) placement.³ Unfortunately, the commonly utilized extracardiac lateral tunnel (ECLT) technique, where a prosthetic or

autologous pericardial conduit connects the inferior systemic venous return to the pulmonary arterial circulation, precludes TV access to the atria (Figure 1).^{4,5}

Newcombe et al⁶ reported a successful TV lead placement for SND in a patient with an ECLT Fontan constructed with autologous pericardium. Here, the authors were able to map atrial tissue within the Fontan baffle. For patients without paceable tissue in the Fontan pathway, 2 studies have recently described a transpulmonary technique whereby the patient's atrial endocardium was accessed via TV puncture into a branch pulmonary artery, followed by advancement of the pacing lead into the common atrium.^{7,8} We report 2 additional cases where TV pacemaker implantation was attempted in 2 patients with both SND and ECLT Fontan anatomy.

Case report

Case 1

The patient was a 20-year-old man with a history of heterotaxy, unbalanced atrioventricular canal, double-outlet right ventricle, interrupted inferior vena cava with azygos continuation to the left-sided superior vena cava, and hepatic venous drainage to the right atrium. He underwent a left bidirectional Glenn (Kawashima) procedure at 21 months of age, with residual hepatic venous drainage to the inferior common atrium. Finally, he underwent an ECLT Fontan and atrioventricular valvuloplasty at 10 years of age, with tunneling of his hepatic veins to the pulmonary arterial confluence via an 18-mm Gore-Tex tube graft. He presented at 20 years old with 6 months of progressive fatigue and palpitations, and was found to have sinus pauses > 3 seconds on Holter monitoring and a maximum heart rate of 102 beats per minute on exercise testing. He was taken for hemodynamic catheterization, during which time he also underwent a pacing study that documented several potential pacing sites in the posterior inferior aspect of the Fontan baffle, near the anastomosis with the hepatic veins.

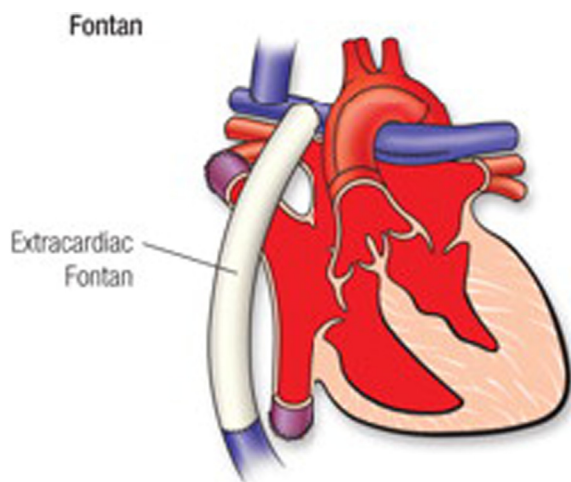


Figure 1 Example of an extracardiac lateral tunnel Fontan circuit. Reprinted with permission from Wolters Kluwer.⁵

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KEY TEACHING POINTS

- Sinus node dysfunction affects 9% of patients with single ventricle physiology that are palliated to a Fontan circulation.
- Pacemaker implantation is the most common procedure performed in patients with Fontan circulation.
- Transvenous lead implantation spares patients the morbidity and complications of open surgery, yet requires systemic anticoagulation.
- Transvenous lead implantation in patients with extracardiac lateral tunnel Fontans is possible, though technically challenging. Ppaceable tissue may be found within the baffle itself or via transpulmonary access to the native atrium.

Given his symptoms and objective evidence of SND, he was taken to the catheterization lab for TV atrial pacemaker implantation. Venous access was obtained via the left subclavian vein. Hand contrast injections were used to visualize the Fontan pathway and potential pacing sites identified in the previous catheterization. Numerous sites were sampled with the permanent pacing lead and high thresholds were noted in the inferior portion of the Fontan baffle. More lateral areas resulted in diaphragmatic capture. It was only when the lead was secured to the posterior inferior aspect of the baffle near the presumed anastomosis with hepatic veins that an adequate capture threshold was noted. Stable lead position was confirmed fluoroscopically. Unfortunately, pacemaker interrogation noted loss of capture several hours after the procedure and chest radiograph (CXR) showed lead dislodgement. The procedure was unsuccessfully reattempted the following day owing to a combination of poor pacing/sensing thresholds, continued lead dislodgement, and inadvertent capture of the phrenic nerve. It was decided to abort the procedure and reattempt at a later time, given the patient's preference to avoid another open surgical procedure. One month later he returned to the catheterization laboratory, where adequate pacing/sensing thresholds were found in the posterolateral aspect of the inferior hepatic baffle (Figure 2). A Medtronic SelectSecure 49-cm bipolar lead was fixated at this site and connected to a Medtronic Adapta ADSR01 generator. Capture threshold was found to be 1.50 V at 0.40 msec. He was discharged home on warfarin with target international normalized ratio of 2.0–3.0.

At most recent follow-up (4 months from implantation), the patient continues to have adequate lead function, with a capture threshold of 1.0 V at 0.40 msec, and stable lead position on CXR. Unfortunately, he has missed several follow-up appointments, which is concerning, given his systemic anticoagulation and need for international normalized ratio monitoring.

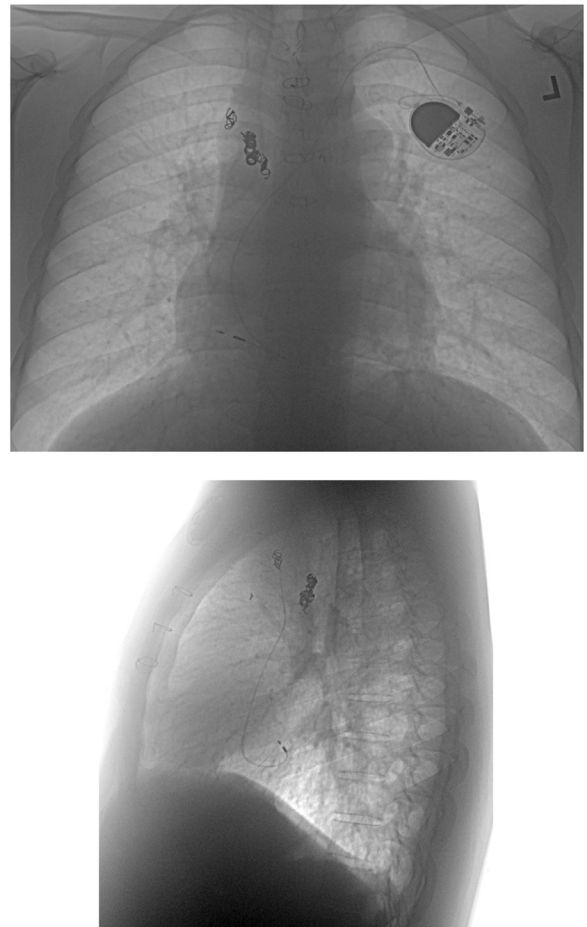


Figure 2 Posterior-anterior and left lateral chest radiographs of final lead position for Case 1.

Case 2

The patient was a 14-year-old boy with a history of tricuspid atresia, ventricular septal defect, D-transposition of the great arteries, and pulmonary stenosis. He underwent bidirectional Glenn placement at 1 year of age and ECLT Fontan at 2 years of age. Subsequently, he developed SND requiring epicardial pacemaker implantation at 7 years of age at an outside facility. At presentation, his pacemaker showed progressive evidence of lead failure with atrial and ventricular non-capture resulting in 2-second pauses, in addition to declining impedance in the atrial lead. A 24-hour Holter monitor further demonstrated these findings, with multiple examples of atrial and ventricular non-capture.

Given these findings and with the generator approaching its elective replacement indicator, it was decided to proceed to pacemaker revision with both generator change and lead replacement. In an effort to try and limit patient morbidity, a TV approach was first attempted. Access was obtained via the right internal jugular vein, purposefully done to test for pacing sites prior to creating a pocket or violating the left subclavian vein. Contrast hand injections showed an unobstructed Fontan pathway and bilateral pulmonary arteries. With a deflectable decapolar catheter, atrial capture was demonstrated at the left pulmonary artery/Fontan junction,

where the distal tip of the catheter effaced the roof of the atria. A Boston Scientific Fineline II pacing lead (Model 4469) was then inserted via the right internal jugular vein. Despite trying numerous locations and screwing the lead deep into the atrial tissue, the lowest capture threshold in this region was 3.1 V at 0.5 msec. Given the inability to pace the atrium at acceptable outputs, sheaths and leads were removed and the procedure was aborted.

He then underwent dual-chamber epicardial lead placement (with 60-cm Medtronic 4968 leads) and a generator change (Medtronic Adapta ASDR01) 6 days later with an uncomplicated intraoperative and initial postoperative course. Atrial capture threshold was 2.0 V at 0.40 msec, and he was discharged on postoperative day 3. Unfortunately, he presented 6 days after discharge to clinic with pallor and tachypnea. CXR was obtained, which showed a left-sided pleural effusion occupying roughly 25% of the left hemithorax. After an outpatient trial of enteral furosemide failed to resolve symptoms, he was admitted to the hospital for parenteral furosemide and standing ibuprofen. He was discharged 4 days later on maintenance enteral furosemide in addition to a 2-liter fluid restriction per day. At clinic follow-up 3 months from discharge a CXR showed only a faint residual left-sided effusion and enteral furosemide was discontinued. His atrial capture threshold nearly 1 year from implantation remains adequate at 2.0 V at 0.4 msec.

Discussion

These cases serve as 2 additional examples of TV pacemaker implantation attempts in patients with ECLT Fontans, and highlight the benefits, risks, and complexities one must consider when deciding on a transvenous vs epicardial approach.

Patients in whom transvenous leads are being considered should have their Fontan pathway mapped before a pacemaker pocket is created or a permanent lead inserted. Mapping of the baffle can be considered during an interventional catheterization procedure that does not involve electrophysiological interventions, which could also delineate the proximity of the pulmonary arterial vasculature to the atrium via angiography. Depending on how the baffles are created and the material of which they are made, isolated islands of paceable tissue may be available within the baffle itself, or, in some cases, via transpulmonary access. In patients without a need for heart catheterization, cardiac computed tomography can be considered to better understand the anatomical relationship of the Fontan pathway with the common atrium and determine whether transvenous pacing is potentially feasible.⁸

"Hunting" for paceable tissue is a relatively straightforward task. Use of a deflectable catheter via the internal jugular vein allows for sites to be tested prior to the pacemaker site being violated (ie, without the creation of a pacemaker pocket or access to the axillary or subclavian vein). Once viable tissue is found, use of a permanent pacing lead via the internal jugular vein can confirm whether there

are acceptable pacing and sensing thresholds, and whether there is capture of the phrenic nerve. This approach allows the physician to quickly rule out, or confirm, the ability to place a transvenous lead without significant risk to the patient.

The likelihood of success may primarily be dictated by how the surgeon created the Fontan baffle. In Case 1, the presence of an interrupted inferior vena cava resulted in the inferior limb of the Fontan being anastomosed to the hepatic veins. This likely left a remnant of atrial tissue that could be paced. Although this tissue is not typically present in ECLT Fontans, this case may highlight the utility of leaving a cuff of atrial tissue in the inferior portion of all extracardiac baffles in case atrial pacing is required in the future. This could negate the need for transpulmonary atrial lead placement, along with its inherent risk of cardiac perforation, and systemic emboli and stroke.⁸

Unfortunately, even with paceable tissue present, our cases illustrate the technical difficulty of securing a lead in the appropriate location. Lead dislocation is more likely to occur given the complex course the lead travels from the innominate vein and through the Fontan. This is often a tortuous route, with numerous turns as the lead passes from the innominate vein, into the superior vena cava, through a native branch pulmonary artery, and then into the baffle itself. Once there, the baffle itself is relatively narrow, adding to the difficulty in maneuvering the lead into the correct position. The tortuous lead course often leads to difficulty screwing the lead into the myocardium. After securing the lead, the distal electrode may then have limited contact with the endocardial surface of the atrium, resulting in unacceptably high atrial capture thresholds.

Despite the difficulties associated with TV leads in ECLT Fontans, they do provide a useful alternative to epicardial leads. A transvenous lead can spare the patient a repeat sternotomy or thoracotomy and the potential for pleural or pericardial effusions, perioperative bleeding, and prolonged hospital stays that are known to complicate the epicardial approach in this population.^{9,10} However, as illustrated in this case series, the acute benefits of a TV lead must be balanced with the patient's risk for venous thromboembolism and potential for nonadherence with anticoagulation regimens.¹¹ To our knowledge, no studies have demonstrated the safety and/or efficacy of the novel anticoagulants in preventing thrombosis in TV systems. Future investigations into this area would be beneficial.

Conclusion

In conclusion, TV lead placement in patients with Fontan circulations are a good alternative to open surgery and epicardial leads, especially with intra-atrial lateral tunnel baffles that have easily accessible atrial tissue to be paced. Our series provides an approach to mapping patients with ECLT to determine their candidacy for a transvenous lead, and provides evidence that TV lead implantation in ECLT Fontans is feasible, despite previously being viewed as a

contraindication owing to the lack of paceable tissue. This may be particularly true in extracardiac tunnels constructed with autologous pericardium, which theoretically allows for conduit growth with the patient.^{4,6} After discussion with the patient regarding risks and benefits of a TV vs epicardial system, including the long-term need for anticoagulation and potential for thrombosis in the former, we recommend that patients who choose to proceed with a TV device have their extracardiac tunnel mapped via the jugular vein prior to the creation of a pacemaker pocket. This adds minimal risk to the patient and potentially saves the patient from the complications inherent to epicardial pacemakers. Long-term follow-up and further studies would be needed to determine the risk of thromboembolic complications.

References

- Petko M, Myung RJ, Wernovsky G, Cohen MI, Rychik J, Nicolson SC, Gaynor JW, Spray TL. Surgical reinterventions following the Fontan procedure. *Eur J Cardiothorac Surg* 2003;24:255–259.
- Fishberger SB, Wernovsky G, Gentles TL, Gamble WJ, Gauvreau K, Burnett J, Mayer JE, Walsh EP. Long-term outcome in patients with pacemakers following the Fontan operation. *Am J Cardiol* 1996;77:887–889.
- Heinemann MK, Gass M, Breuer J, Ziemer G. DDD pacemaker implantation after Fontan-type operations. *Pacing Clin Electrophysiol* 2003;26:492–495.
- Hasaniya NW, Razzouk AJ, Mulla NF, Larsen RL, Bailey LL. In situ pericardial extracardiac lateral tunnel Fontan operation: fifteen-year experience. *J Thorac Cardiovasc Surg* 2010;140:1076–1083.
- d'Udekem Y, Iyengar AJ, Cochrane AD, Grigg LE, Ramsay JM, Wheaton GR, Penny DJ, Brizard CP. The Fontan procedure: contemporary techniques have improved long-term outcomes. *Circulation* 2007;116:1157–1164.
- Newcombe J, Gordon B, Razzouk A, Bailey L, Mandapati R. Extracardiac autologous pericardial tunnel Fontan allows implantation of an endocardial atrial lead for sinus node dysfunction. *Ann Thorac Surg* 2014;98:1094–1096.
- Moore JP, Shannon KM. Transpulmonary atrial pacing: an approach to transvenous pacemaker implantation after extracardiac conduit Fontan surgery. *J Cardiovasc Electrophysiol* 2014;25:1028–1031.
- Arif S, Clift PF, De Giovanni JV. Permanent trans-venous pacing in an extracardiac Fontan circulation. *Europace* 2015;19:110.
- Hansky B, Blanz U, Peuster M, Gueldner H, Sandica E, Crespo-Martinez E, Mathies W, Meyer H, Koerfer R. Endocardial pacing after Fontan-type procedures. *Pacing Clin Electrophysiol* 2005;28:140–148.
- Takahashi K, Cecchin F, Fortescue E, Berul CI, Alexander ME, Walsh EP, Fynn-Thompson F, Triedman JK. Permanent atrial pacing lead implant route after Fontan operation. *Pacing Clin Electrophysiol* 2009;32:779–785.
- Khairy P, Landzberg MJ, Gatzoulis MA, Mercier LA, Fernandes SM, Côté JM, Lavoie JP, Fournier A, Guerra PG, Frogoudaki A, Walsh EP, Dore A. Epicardial Versus Endocardial Pacing and Thromboembolic Events Investigators. Transvenous pacing leads and systemic thromboemboli in patients with intracardiac shunts: a multicenter study. *Circulation* 2006;113:2391–2397.