Transcarotid implantation of a leadless pacemaker in a patient with Fontan circulation

Peter Calvert, MBChB(Hons), MRCP,*† Cheng Yeo, MD, FCAI,* Archana Rao, MD, MRCP,* Simon Neequaye, MSC, BSc(Hons), FRCS,*‡ David Mayhew, MBBS, BSc(Hons), FRCA,* Reza Ashrafi, MBBS, MRCP*

*From the *Liverpool Heart & Chest Hospital, Liverpool, United Kingdom, †Liverpool Centre for Cardiovascular Science, University of Liverpool, Liverpool, United Kingdom, and ‡Liverpool University Hospitals Foundation Trust, Royal Liverpool Hospital, Liverpool, United Kingdom.*

Introduction

Cardiac pacing is a well-established therapy for symptomatic bradycardia and high-grade atrioventricular (AV) block. Permanent transvenous pacing traditionally has necessitated the implantation of long-term leads, raising the risk of complications such as infection, thrombosis, and vessel stenosis. Leadless pacemakers have been welcomed as a way to reduce such complications in selected patients.

Pacemaker implantation can be challenging in patients with adult congenital heart disease (ACHD), where transvenous access to cardiac mass may have been surgically rerouted. We present a rare and complex case of a patient with Fontan circulation receiving a leadless pacemaker implant, which was successfully delivered via the carotid artery.

Case report

The patient provided consent for the writing of this case report.

Background and history

Our patient was a 26-year-old man with complex ACHD. His cardiac history included mitral atresia with a hypoplastic left ventricle, transposition of the great arteries, hypoplastic aortic arch with coarctation, and a small ventricular septal defect.

As a neonate, he had undergone coarctation repair with atrial septectomy and pulmonary artery banding. This was complicated by complete AV block and an epicardial pacemaker was placed via a median sternotomy. Following this, he underwent a Damus-Kay-Stansel procedure and a Glenn shunt. At 6 years of age, he underwent completion of the Fontan circulation with a 22m Gore-Tex extracardiac conduit and implantation of new epicardial atrial and ventricular pacing wires, all performed via median sternotomy.

Over the next few years, he suffered from multiple device infections, which presented as superficial wound infections, deep-seated wound infection with sinuses, and at least 1 episode of systemic illness. Over the years he had multiple courses of antibiotics, wound revisions, exploratory left thoracotomies, and finally full sternotomy to remove an old lead fragment. His atrial lead failed after 3 years, and he was left 100% VVI-paced in sinus rhythm. At age 24, an attempt at transbaffle pacing was successful at inserting a new atrial lead, but a ventricular lead could not be positioned. Repeat sternotomy was required to implant a new epicardial ventricular lead; however, this culminated in an episode of mediastinitis. Within 18 months, the epicardial ventricular lead threshold had increased to 5 V at 0.4 ms, with rapid battery depletion.

KEY TEACHING POINTS

- In complex congenital heart disease alternative access strategies to traditional pacing maybe required and this may require multidisciplinary and hybrid approaches.
- Leadless pacemakers offer a promising option in patients with complex congenital heart disease owing to the lack of a need for long-term lead placement and reported lower incidence of infection in the structurally normal heart cohort.
- When performing large-bore sheath procedures in complex cases, transesophageal monitoring is helpful in procedural guidance and for monitoring of complications.

KEYWORDS Leadless pacemaker; Fontan; Single ventricle; Transcarotid; Adult congenital heart disease

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The case was discussed in the congenital multidisciplinary team (MDT) meeting. The option of repeat surgical lead placement was considered; however, given the previous mediastinitis, the multiple thoracotomies, and the opinions of the surgical team, who noted heavily scarred tissue during his previous procedure, this was not considered feasible. Consideration was therefore given to placing a leadless pacemaker. The patient had a systemic right ventricle (RV), with a hypoplastic left ventricle considered too small to place a device into specifically. Furthermore, the optimal vascular access route with which to place a leadless device required consideration. These factors were discussed at the MDT meeting. The traditional femoral venous route would arrive at the Fontan circuit. A transbaffle approach was considered; however, the previous transbaffle puncture had been challenging and it was felt that a fenestration of the size required for a leadless device would not be feasible. The distance from the femoral artery was too far for currently available sheaths. The curve of the sheath also made axillary arterial access unfavorable. Finally, the left carotid artery was considered the most viable option. This was subsequently discussed and agreed upon with the patient.

Preoperative work-up

A right-sided pacemaker with transbaffle atrial and epicardial ventricular leads remained in place and a device check showed 100% ventricular pacing with no underlying escape rhythm. Cardiac catheterization found a conduit pressure of 15 mm Hg and wedge pressure of 10 mm Hg.

Transthoracic echocardiogram showed a systemic RV with preserved function, mild-to-moderate tricuspid regurgitation, and no thrombus within the Fontan circuit. A computed tomography scan demonstrated normal carotid anatomy and patent circle of Willis.

Anticoagulation with warfarin was withheld preoperatively for 72 hours. His international normalized ratio was less than 2 for the procedure.

Procedural details

The procedure was carried out in a hybrid theatre. External defibrillator/pacing pads were placed prior to induction of general anesthesia with remifentanil and propofol. Anesthesia was maintained with sevoflurane and remifentanil.

An arterial pressure line was placed in the right radial artery and a central venous line was placed in the right internal jugular vein. The left neck was spared for surgical access to the carotid artery. A femoral arterial sheath was placed for further vascular access and to facilitate a postprocedural aortic arch angiogram. Preprocedural transesophageal echocardiography (TEE) showed no thrombus within the Fontan conduit.

Cerebral saturations were monitored with near-infrared spectroscopy. A target mean arterial pressure $\geq$65 mm Hg necessitated judicious fluid administration and commencement of a phenylephrine infusion.

The procedure began with surgical exposure of the left carotid artery using the technique applied during carotid endarterectomy. Following systemic heparinization (target activated clotting time $\geq$200 seconds) and with sloops around the carotid artery, a puncture was performed using an 18G needle, and a 6F splittable sheath was placed over a 0.035 guidewire. Using a 6F AL2 diagnostic catheter, the main outflow valve was crossed using fluoroscopic guidance and an 0.035 J tip Amplatzer superstiff (Boston Scientific, Clonmel, Co. Tipperary, Ireland) wire unfurled in the systemic ventricle and the diagnostic catheter removed. TEE and fluoroscopy showed good wire trajectory through the middle of the valve and with pressure hemostasis the 6F sheath was removed and a Coons 22F dilator (Cook Medical, Castletroy, Co. Limerick, Ireland) taken over the superstiff wire to dilate the carotid artery.

The Coons dilator was up-sized to a 26F Gore dry sheath (W. L. Gore & Associates, Livingston, Scotland, UK). Following this, an AV-Micra (Medtronic Inc, Minneapolis, MN) leadless pacemaker was deployed into the systemic RV under fluoroscopic guidance without complication (Figure 1). TEE here showed stable sheath position with a smooth trajectory through the outflow and AV valves and no obvious leaflet prolapse due to the sheath. An initial position apically had poor pacing parameters and with a small amount of anticlockwise catheter torque, a second septal position had better pacing parameters.

The sensed R wave was 4.9 mV with an impedance of 830 Ω and threshold of 0.38 V at 0.24 ms. The paced QRS duration was 90 ms, in comparison to previous epicardial paced QRS of 160 ms (Figure 2A and 2B). His previous device was then switched off, as he had no atrial pacing requirement, with a native sinus rate measured on exercise tests up to 170 beats per minute, with reasonable tracking from the AV-Micra. Periprocedural TEE showed good placement of the device within the ventricle, and on removal of the delivery apparatus no new significant valvular regurgitation was observed.

Sheaths were removed and the carotid site was closed surgically. Aortic arch and carotid angiography were normal following this. The patient remained hemodynamically stable throughout the procedure and postoperative recovery was uneventful. Anticoagulation was restarted the following morning with edoxaban 60 mg once a day, to obviate international normalized ratio monitoring and minimize the burden to the patient, and he was discharged home after 24 hours of monitoring.

Long-term outcome

Since the procedure, the AV-Micra device has functioned normally, with a pacing threshold of 0.13 V at 0.24 ms. AV synchrony was achieved roughly 60%–70% of the time with an A4 amplitude of 0.5 m/s based on device-reported AV synchrony and on review of the 12-lead electrocardiograms. As the patient has spent most of his life with VVI pacing only, this was considered a substantial improvement. The patient has since returned to work and has been clinically stable, with no major adverse effects or infection issues, in

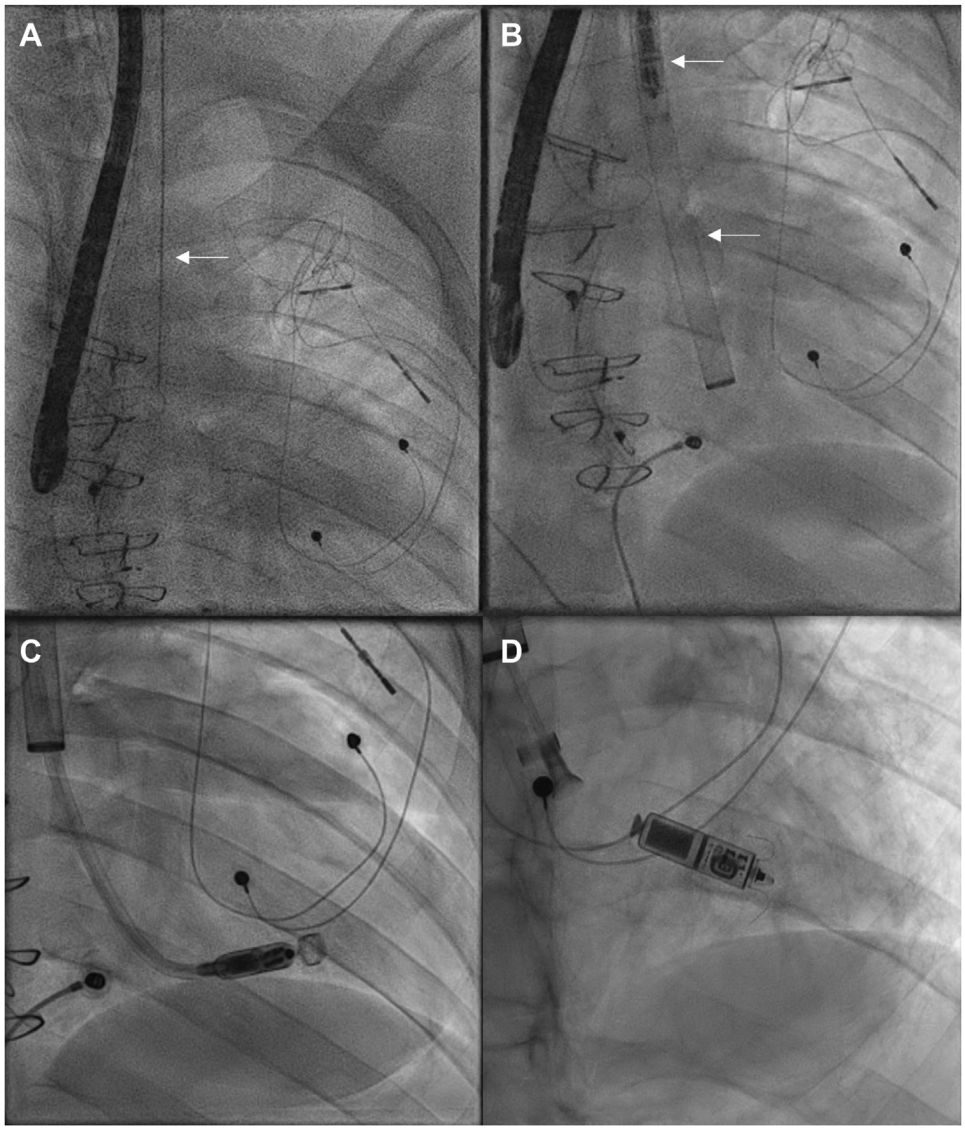

Figure 1 Fluoroscopic images from implantation of an AV-Micra (Medtronic Inc, Minneapolis, MN) device implanted via transcarotid access. **A:** Guidewire is advanced via the carotid artery (*arrow*). **B:** A 26F Gore dry sheath is advanced to the heart with the AV-Micra device in transit (*arrows*). **C:** AV-Micra device during deployment. **D:** AV-Micra device following deployment. A pull-back test is performed to ensure stability.

contrast to his many previous procedures. A postprocedural transthoracic echocardiogram has shown stable device function with no obvious thrombus around the device or nearby.

Discussion

We present a unique case of a young patient with Fontan circulation requiring long-term pacing for complete AV block, with successful deployment of a leadless pacemaker via the carotid artery. Below, we discuss the principal considerations when managing such complex cases in the context of our experience with this patient.

Decision to pace and type of device

Although pacing is generally avoided in younger patients, it is unfortunately frequently required in Fontan patients owing to either significant sinoatrial dysfunction or AV block.[1] Ventricular pacing should be minimized, as this increases the risk of long-term morbidity and mortality in Fontan patients.[2] Atrial pacing is preferred to maintain normal AV synchrony and normal ventricular activation; in Fontan patients, this has been shown to improve cardiac output, left atrial pressure, and pulmonary blood flow.[3] Where this is not possible—such as in our case, owing to longstanding complete AV block—maintaining AV synchrony remains important. This is typically achieved with an atrial pacing wire, but newer leadless devices such as the AV-Micra are able to sense atrial systole mechanically, resulting in effective AV synchrony without an atrial lead.

Transvenous pacing may be achieved safely and effectively in many cases, but not in all complex ACHD patients. Short- or long-term complications occur in as many as 1 in 6 patients, primarily owing to lead failure or infection.[4] The impact upon patients and the associated healthcare costs are

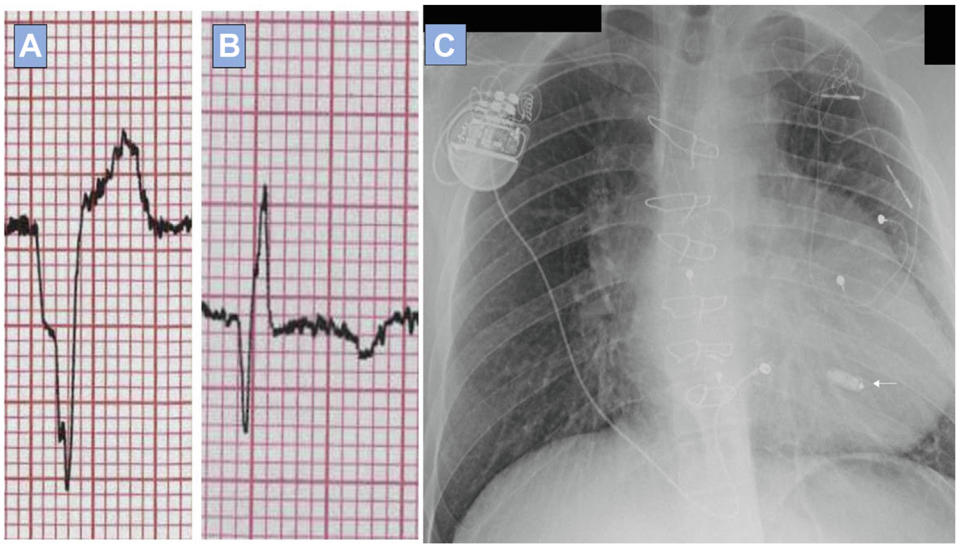

Figure 2 **A:** Preprocedural paced electrocardiogram (ECG). **B:** Postprocedural paced ECG with much-improved QRS width. **C:** Postoperative chest radiograph. The arrow shows the AV-Micra (Medtronic Inc, Minneapolis, MN) device; also shown is the right-sided epicardial pacemaker and left-sided discarded epicardial pacing leads.

substantial. In the Fontan population, implantation can be challenging owing to anatomical constraints. Epicardial systems are frequently used in these patients; however, as this requires sternotomy or thoracotomy, epicardial leads are usually placed during surgery for a different indication. Additionally, while studies show similar performance between epicardial and endocardial pacing in Fontan circulation, there are trends toward increased lead failure and reduced generator longevity with epicardial systems.[5]

Leadless pacemakers are a relatively new development, with low rates of serious complications and very few reported infections.[6] Furthermore, lead failure is negated and circulatory access sites are preserved for future interventional procedures. Evidence for use of these devices in the Fontan population is currently limited, however.

Anatomical access and thromboembolic risk

In extracardiac or lateral Fontan patients, several endocardial pacing approaches have been demonstrated. These include transhepatic, transbaffle, use of collateral vessels, and hybrid techniques.[7] There are also case reports of transpulmonary atrial pacing[8] and ventricular pacing via persistent left superior vena cava.[9] Given the relative rarity of such patients, large-scale trial data are scarce. All of these approaches require long-term pacing leads placed in the systemic ventricle; hence there is a risk of serious complications. Systemic thromboembolism is of particular concern in Fontan circulation, owing to reduced blood flow and hypercoagulability.[10] This can be reduced with long-term anticoagulation, which is already indicated in these patients to lower the risk of Fontan circuit thrombosis.

In normal anatomy, the Micra is deployed in the subpulmonary RV via the femoral vein. In a Fontan patient, femoral venous access would arrive at the Fontan conduit, leading to the right pulmonary artery. While it is possible to create a fenestration through which a transvenous lead can pass to the ventricle, this was considered unfeasible in our case owing to the large size of the sheath (26F). Subsequently, we considered the femoral and axillary arteries; however, these were considered unfeasible, as described above. The left carotid artery provided a much straighter anatomical course, with the benefit of collateral cerebral blood flow via the contralateral carotid artery and the vertebrobasilar system. Indeed, transcarotid access has been used for transcatheter aortic valve implantation for some time, with similar safety to transfemoral access.[11]

Preoperative discussions and work-up

An MDT discussion is recommended for any complex ACHD case where an unusual procedure is planned. In our case, the MDT included expertise in ACHD, electrophysiology, pacing, vascular and cardiothoracic surgery, and cardiac anesthesia.

Once a decision is made, the importance of careful discussion with the patient cannot be understated. Shared decision-making in difficult cases is critical, and the patient should be made aware of the relatively unknown risks.

Thorough preoperative work-up provides 2 major benefits. Firstly, the identification of indications for sternotomy may shift the plan to replacement of epicardial leads. Secondly, a detailed work-up allows for effective risk stratification and planning, particularly from an anesthetic perspective. Attention should also be given to any Fontan-related complications, such as enteropathy or liver disease.

Preoperative imaging—which may include ultrasound, computed tomography, and/or magnetic resonance imaging—may provide valuable information both for risk stratification and access planning.

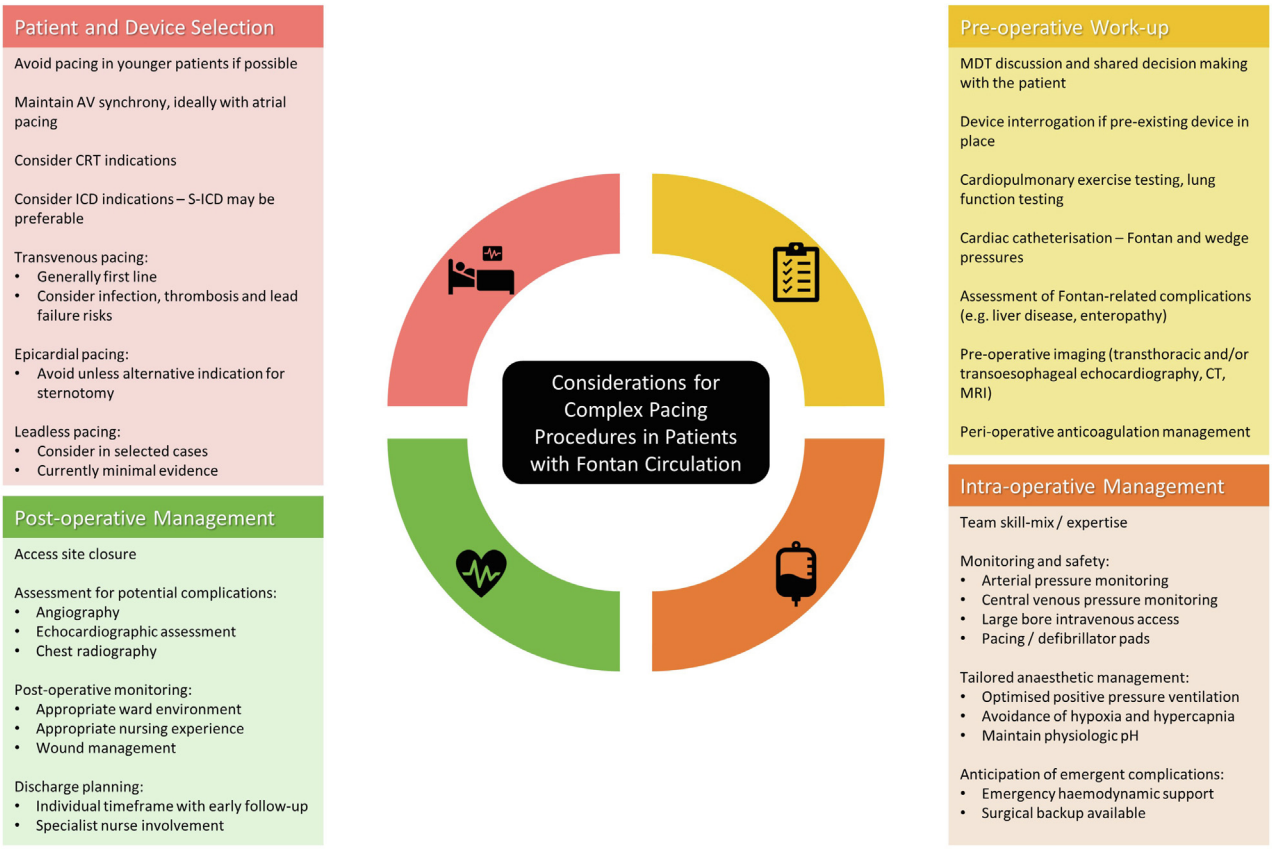

Figure 3 Considerations for complex pacing in patients with Fontan circulation. AV = atrioventricular; CRT = cardiac resynchronization therapy; CT = computed tomography; ICD = implantable cardioverter-defibrillator; MDT = multidisciplinary team; MRI = magnetic resonance imaging; S-ICD = subcutaneous implantable cardioverter-defibrillator.

Further considerations, which may alter the management plan, include indications for cardiac resynchronization therapy (CRT) and implantable cardioverter-defibrillator. The evidence for CRT in the Fontan population is limited and CRT implant may be very difficult to achieve.[12] Implantable cardioverter-defibrillator implant may be equally challenging[12]; therefore, consideration of a subcutaneous device may be required.

Intraprocedural management

In complex cases, a clear plan for the operation should be discussed among theatre staff prior to starting. Large-bore intravenous access should be available—ideally with a central line, which facilitates central venous pressure monitoring. Obviously, such lines should be kept distant from the intended site of pacing access.

The potential for emergent complications must be considered. TEE and transthoracic echocardiography should be available. The potential need for cardiopulmonary resuscitation should be anticipated and roles should be assigned prior to starting the procedure. Defibrillator pads should be applied, though care should be taken to avoid positioning these where they might obscure fluoroscopic guidance. Appropriate anticoagulation reversal agents and blood products should be readily available.

Anesthetic technique should be tailored such that the main determinants of forward flow in Fontan circulation (systemic venous pressure, pulmonary vascular resistance, AV valve function, cardiac rhythm, and ventricular function) are not compromised. Familiarity with anesthetic agents is more important than adhering to a fixed anesthetic regime. In our case, remifentanil—an ultra-short-acting opioid—was used. Remifentanil confers predictability and ease of titratability in response to laryngoscopy and surgical stimulus during carotid cut-down, obviating muscle relaxant top-ups. Remifentanil can be associated with bradycardia, so care should be taken if a pacing system is not already in place.

Caution should also be exercised when instituting positive pressure ventilation with respect to airway pressure and positive end-expiratory pressure, as this affects pulmonary vascular resistance. Similarly, hypercapnia, acidosis, and hypoxemia should be avoided. In our case, near-infrared spectroscopy was used to observe for cerebral desaturation. Although this is a potentially useful monitoring tool, the impact on overall clinical outcomes is uncertain.[13]

Intraoperative hypotension can be counteracted with pressor agents or preload challenging, guided by central venous pressure and arterial pressure monitoring. If carotid access is

used, obstruction of the brachiocephalic system during the procedure may be reflected on a right radial arterial line tracing.

Delivery of the device itself should follow standard manufacturer guidelines. Figure 1 shows intraprocedural deployment of the AV-Micra device in our case.

Postoperative management

In such complex and unusual cases, imaging to assess for subclinical complications is essential. In our case, an aortic arch angiogram was performed and was normal. Postoperative chest radiography (Figure 2C) demonstrated the device remained in the deployed position and excluded access complications such as pneumothorax.

Wound care is essential, and the patient should be advised of when to seek medical attention, especially as anticoagulation should restart as soon as possible.

Other published evidence

Two prior case reports have described leadless pacemaker implant in patients with single ventricular circulation. Both cases used femoral venous access in patients without Fontan palliation.[14,15]

To the best of our knowledge, our case represents 2 firsts: (1) the first use of a leadless pacemaker in the systemic ventricle of a patient with extracardiac Fontan circulation and (2) the first use of a leadless pacemaker implanted via transcarotid access.

Further evidence is required with regard to long-term outcomes using such devices in the Fontan population. Additionally, long-term planning for end of battery life will need careful consideration and MDT discussion to determine the optimal approach.

Conclusion

We present the first case of leadless pacing in a Fontan patient via the transcarotid route, along with a recommended approach to such patients. This technique appears safe and effective; however, further research will be required to determine long-term outcomes in this complex group of patients. Our suggested management considerations are summarized in Figure 3.

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