A dual-chamber leadless pacemaker in d-TGA patient after senning procedure

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Introduction

Although the arterial switch operation is now the standard of care for dextro-transposition of the great arteries (d-TGA), many adults underwent Mustard and Senning (atrial baffle) procedures as children. Approximately 30% of these patients require pacemakers by adulthood.[1] Arrhythmias requiring pacemakers or implantable cardioverter-defibrillators are among the most common long-term complications for these patients.[2] Transvenous pacemaker leads may cause thrombosis and partial or total occlusion of the subclavian vein or baffle in 30% to 50% of patients with prior Mustard surgery. Furthermore, stenting of the baffle in case of occlusion or leak requires transvenous lead extraction. We present a unique case of using a dual-chamber leadless pacemaker in a patient with d-TGA who previously had a transvenous pacemaker for sinus node dysfunction but experienced pocket pain and recurrent infection, necessitating multiple pocket revision surgeries and lead extraction.

KEY TEACHING POINTS

- This is the first case report demonstrating the feasibility of dual-chamber pacemaker implantation in a patient with d-TGA with an atrial switch.
- Special considerations during preoperative and perioperative evaluation include navigating the delivery system through anatomic challenges, ensuring baffle patency, addressing distance between pacemaker components, and mitigating potential phrenic nerve stimulation from the atrial lead.
- Testing for phrenic nerve capture before deploying the atrial leadless pacemaker is essential.
- Preprocedural and periprocedural imaging, such as CT scan, cardiac magnetic resonance imaging, or intracardiac echocardiography, is crucial in anatomically complex cases.

Case report

A 39-year-old man with d-TGA underwent a Senning procedure at 7 months of age and received a dual-chamber epicardial pacemaker at 6 years because of sinus node dysfunction. At 18 years, the abdominal generator of the epicardial system was removed, and a transvenous dual-chamber pacemaker was implanted in the left prepectoral area. At 31 years, because of the elective replacement indicator, he underwent a generator exchange, during which a malfunctioning atrial lead was extracted and a new atrial lead implanted.

Two weeks postprocedure, the patient presented with serous drainage from the pacemaker pocket incision, requiring pocket exploration and fluid evacuation. Six months later, he experienced pocket swelling and tenderness. Despite negative pocket ultrasound and blood cultures, persistent pain led to a pocket revision with scar debridement and relocation to a subpectoral pocket 1 year later.

Three years later, the patient developed tenderness and swelling at the old thoracotomy incision site, 4 inches below the pacemaker incision, resulting in the removal of purulent material and revealing a connection between the abscess cavity and the pacemaker pocket. Subsequent debridement and revision to an intramuscular pocket were performed. Persistent pain and discomfort with arm movement led to another pocket revision to a subpectoral location a few months later.

Two years later, the patient again developed pocket pain, with chest radiographic evidence of device migration, necessitating a fourth pocket revision. Post-revision, he continued to experience pain, tenderness, and keloid formation. Cardiac magnetic resonance imaging (MRI showed a hypertrophied systemic right ventricle with mildly reduced systolic function and a widely patent pulmonary venous baffle.

KEYWORDS Leadless pacemaker; d-TGA; Dual-chamber leadless pacemaker; Adult congenital heart disease; Transposition of the great arteries

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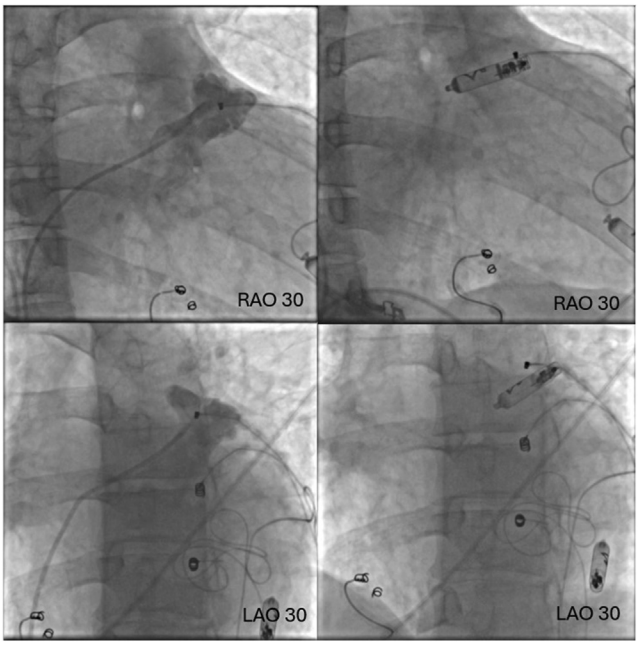

Figure 1 Left atrial appendage angiogram and the final location of the leadless pacemaker.

Eventually, he underwent removal of the generator and laser lead extraction of the atrial and ventricular leads. Because the patient was pacemaker-dependent (with underlying symptomatic sinus bradycardia at a rate of 40 beats/min), a leadless pacemaker (Aveir LP, Abbott Medical, Abbott Park, IL) was implanted using transesophageal echocardiogram and standard fluoroscopy guidance. Subpulmonic left ventricular angiography, performed using intravenous contrast via a pigtail catheter, delineated the apex of the subpulmonic left ventricle. An apical septal site was chosen, with pacing and sensing thresholds of 0.75 V at 0.4 ms and 11 mV, respectively, and a normal impedance of 1170 ohms. Although the patient tolerated VVI pacing well during follow-up, upgrade to a dual-chamber system was planned once it became available. This upgrade aimed to maintain AV synchrony, given the patient's intact AV conduction, and to minimize left ventricular pacing to reduce the risk of pacing-induced cardiomyopathy.

A year later, the patient underwent an upgrade to the Aveir DR system. A pigtail catheter was advanced via the baffle to the left atrial appendage, and an atrial appendage angiogram was performed in the right anterior oblique and left anterior oblique views. Before deployment, phrenic capture testing was performed, and at least 4 attempted locations were found to have significant phrenic captures with low output. At the final location, left phrenic nerve capture was noted at 5.0 V at 0.4 ms, with an initial threshold of 1.5 V at 1.5 ms. The Aveir LP was deployed into the superior base of the atrial appendage (Figure 1). Final pacing threshold and sensing were acceptable at 0.25 V at 0.4 ms and 1 mV, respectively, with normal impedance at 370 Ω. No phrenic nerve capture was demonstrated after releasing the pacemaker. The distance between the atrial and ventricular leadless pacemakers was confirmed to be less than twice the device length (Figure 2). Communication between the atrial pacemaker and ventricular pacemaker was successfully established. The patient was monitored overnight. Subsequent interrogation showed a reduction in the pacing threshold. The atrial lead exhibited a pacing threshold of 0.25 V at 0.4 ms and an impedance of 370 Ω. Atrial escape was absent with AAI 30. Ventricular lead testing indicated a sensing greater than 18 mV, an impedance of 630 Ω, and a pacing threshold of 0.25 V at 0.4 ms. The device was programmed in DDD mode with a rate range of 60 to 130 beats/min and a paced/sensed AV delay of 225/200 ms. The atrial lead output was adjusted to 2.0 V at 0.4 ms with a sensitivity of 0.5 mV, while the ventricular lead output was set to 1.25 V at 0.4 ms with a sensitivity of 2.0 mV. The timeline of his procedures is summarized in Table 1. Electrocardiogram showed appropriate atrial pacing rhythm with intrinsic incomplete right bundle branch block (Figure 3). The patient recovered well after the procedure.

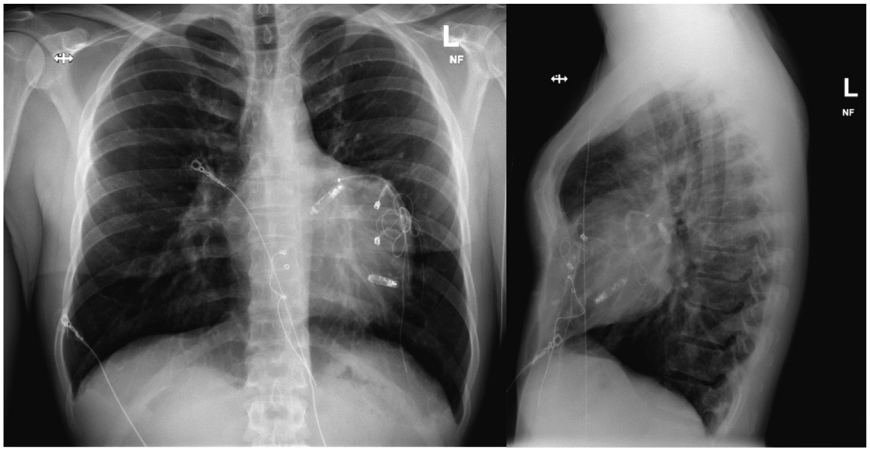

Figure 2 PA and lateral chest radiograph.

Table 1 Timeline of the procedures

Age	Procedures
7 months	Senning procedure
6 years	Epicardial dual-chamber pacemaker implant
18 years	Transvenous pacemaker system implant
31 years	Laser lead extraction of preexisting A lead because of high threshold, new RA lead placement, and generator exchange Pocket evacuation of serosanguinous fluid because of drainage
32 years	Scar debridement and revision to subpectoral pocket
35 years	Abscess connecting to the device pocket. Scar debridement and revision to an intramuscular pocket Revision of pacemaker pocket to subpectoral location
37 years	Scar debridement of pacemaker pocket and relocation of the pocket to new subpectoral pocket location because of device migration
38 years	Removal of pacemaker generator, laser lead extraction of A and V lead, and single-chamber leadless pacemaker implant
39 years	Device system upgrade to dual-chamber leadless pacemaker

Discussion

This case illustrates the feasibility of using a dual-chamber leadless pacemaker in d-TGA patients with an atrial switch who developed sinus node dysfunction requiring a pacemaker and experienced pocket complications from a transvenous pacemaker system. We also highlight the special considerations for implanting the dual-chamber leadless system in this population, including the imaging evaluation of the patency of the baffle and the periprocedural evaluation for phrenic nerve capture from left atrial leadless pacemaker placement.

Sinus node dysfunction has been observed in up to 65% of patients with d-TGA after an atrial switch procedure, with 45% of those surviving into adulthood requiring implantation of a pacemaker or implantable cardioverter-defibrillators.[1] The most common cardiac implantable electronic device complications in patients with congenital heart disease are lead malfunction and generator pocket infection.[3] Leadless pacing therefore may be a viable solution for select patients, either as a permanent system or as a bridge to future surgical procedures.

Since the first report of a leadless pacemaker in adult congenital heart disease was published in 2016,[4] a total of 5 cases have been reported for the use of a leadless pacemaker in d-TGA patients with an atrial switch.[5–9] All patients had a Micra VR (Medtronic, Minneapolis, MN) implant, with the indication of sinus node dysfunction in 3 patients and atrioventricular block in 2 patients. Of these, 3 patients had the leadless pacemaker implanted in the subpulmonic left ventricle. Ventriculography was performed in all cases, and intracardiac or transesophageal echocardiography was occasionally used to navigate the delivery system.[7] In addition to the anatomic challenges and the need to advance the delivery system through the baffle, less prominent trabeculae in the subpulmonic left ventricle compared with a normal right ventricle can pose an additional procedural challenge for leadless ventricular lead placement. The remaining 2 cases had the device implanted in the left atrial appendage, using left appendage angiography to guide the position of device placement.[5,6] Unlike our case, the authors did not report issues related to diaphragmatic stimulation from phrenic nerve capture. This case highlights the critical importance of periprocedural testing for phrenic capture before device deployment.

This case represents the first report of using a dual-chamber leadless pacemaker in d-TGA patients. Chronic subpulmonic left ventricular pacing is associated with systemic right ventricular failure.[10] Therefore, a dual-chamber leadless pacemaker system in patients with intact AV node conduction can reduce the risk of systemic right ventricular failure and may alleviate symptoms related to AV and ventricular desynchrony associated with VVI pacing. For proper communication between the atrial and ventricular leadless pacemakers, it is recommended that the distance between the 2 devices does not exceed twice the device length (76 mm = 38 mm × 2). Figure 2 demonstrates the typical "egg on a string" appearance of the cardiac silhouette in patients with d-TGA (narrow pedicle and enlarged heart). Because the Aveir device is recommended to be placed more apically compared with the Micra device, and because of the need for communication between the 2 devices, the distance between the 2 devices should be taken into account while placing the ventricular device because the atrial device can only be placed in the left atrial appendage.

The major advantage of a dual-chamber leadless pacemaker over a single-chamber device in these patients is the maintenance of atrioventricular synchrony. Baffle intervention is needed in 38% of patients with atrial switch surgeries, and transvenous leads often require extraction because of entrapment by stents, a problem avoided with leadless pacemakers. Although leadless pacemakers are a viable alternative for congenital patients, several limitations exist. The primary limitation is battery longevity in younger patients. Although the Aveir pacemakers were designed to allow retrieval using a dedicated retrieval catheter, the feasibility of extracting a chronically implanted device remains unproven, with evidence limited to an ovine model study and a single case report.[11,12] Implanting a new pacemaker in the same chamber when the first reaches the elective replacement indicator may be problematic unless the old pacemaker is extracted. The lack of atrial anti-tachycardia pacing functionality in leadless pacemakers limits their utility in patients at risk of atrial tachyarrhythmias. Patients who have undergone an atrial switch operation are especially susceptible to developing intra-atrial reentrant tachycardia, which can sometimes be treated with anti-tachycardia pacing. Additionally, infection and thrombus at the device site have been reported with leadless pacemakers. These limitations should

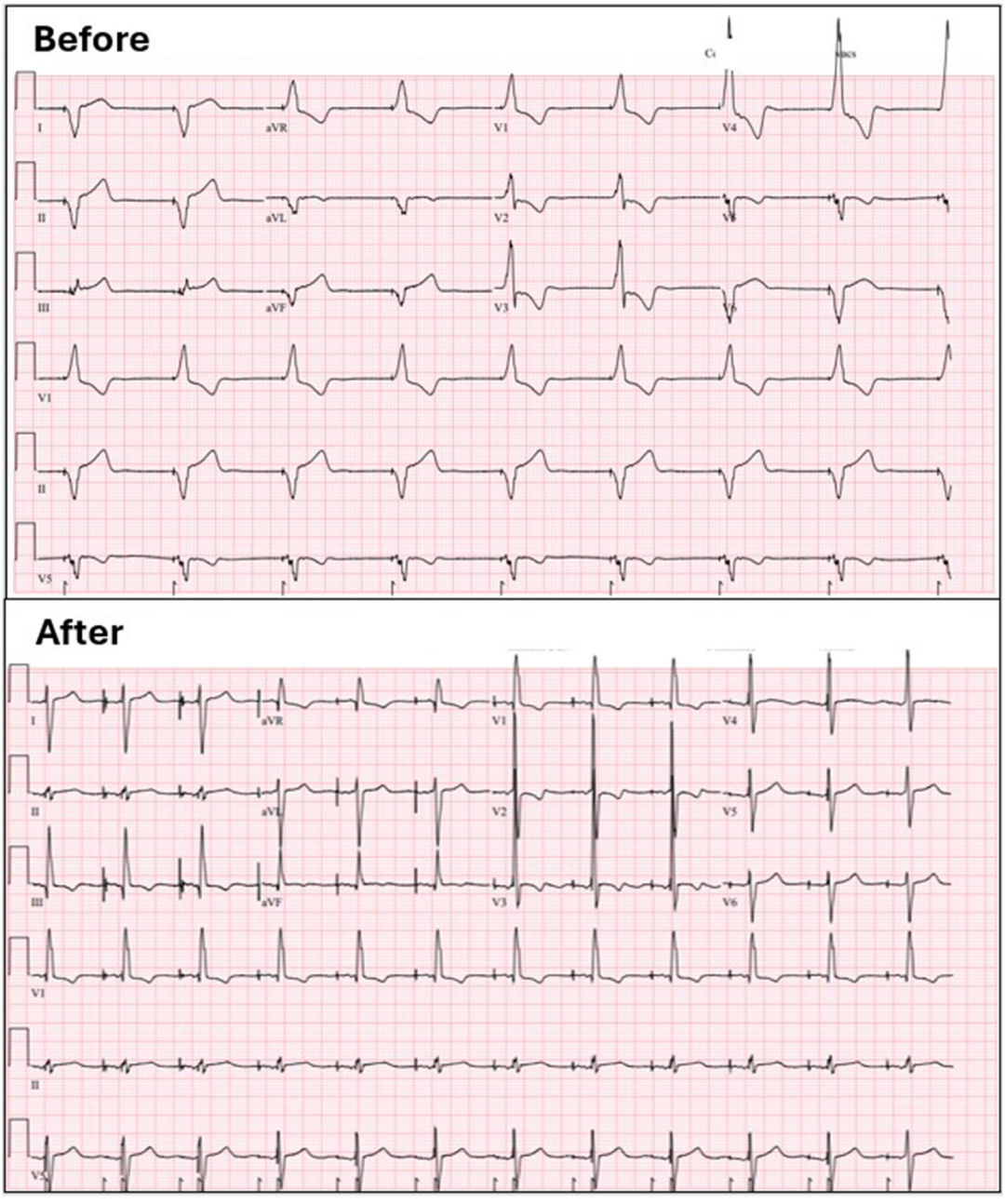

Figure 3 Electrocardiogram before and after upgrade to dual-chamber leadless pacemaker system.

be carefully considered when selecting a pacing device for these patients.

Conclusion

This is the first case report demonstrating the feasibility of a dual-chamber pacemaker in patients with d-TGA with an atrial switch using the Senning procedure, despite anatomic differences and the need to navigate the delivery sheath via a baffle. Special considerations include the patency of the baffle to advance the leadless sheath, the potential for phrenic nerve stimulation from atrial leadless pacemaker placement, and the distance between the 2 devices in the context of a dilated and hypertrophied systemic right ventricle and a small subpulmonic left ventricle. Testing for phrenic nerve capture before deploying the atrial leadless pacemaker is essential. Preprocedural and periprocedural imaging can be beneficial in anatomically challenging cases.

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