Atrioventricular node ablation and left bundle branch area pacing in a patient with dextrocardia and interruption of the inferior vena cava with azygos continuation

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Introduction

Dextrocardia is a rare condition with an incidence of approximately 1 in 12,000 pregnancies.[1] It is often associated with other congenital disorders such as situs inversus, congenital heart diseases, and heterotaxy syndromes of asplenia and polysplenia. Up to 50% of patients with polysplenia have dextrocardia. Interruption of the inferior vena cava (IVC) with azygos continuation is another rare congenital malformation seen in most patients with polysplenia (58%–100%).[2] Radiofrequency ablation of the atrioventricular node (atrioventricular node ablation, AVNA) with permanent pacemaker implantation is a highly effective rate control strategy for drug-refractory atrial fibrillation (AF). Left bundle branch area pacing (LBBAP) has emerged as a novel "conduction system pacing" technique that overcomes the myriad limitations of His bundle pacing, such as elevated pacing capture thresholds and low R-wave amplitudes, while still achieving fast left ventricular (LV) activation times. We describe the first case of AVNA and LBBAP in a patient with drug-refractory AF and dextrocardia and interruption of the infrahepatic IVC with azygos continuation.

KEY TEACHING POINTS

- This article presents the first case report of an atrioventricular (AV) node ablation and left bundle branch area pacing (LBBAP) using a stylet-driven lead in a patient with dextrocardia and interrupted inferior vena cava (IVC).
- Preprocedural imaging and intraprocedural venograms are important to define anatomic variations in these patients.
- Achieving fast left ventricular activation times by engaging the conduction system is crucial in these patients with 100% ventricular pacing burden.
- Catheter ablation of the AV node is feasible from a superior approach via the internal jugular vein in patients with dextrocardia and interrupted IVC.
- LBBAP pacing is challenging in patients with these anomalies. Manually reshaping the guiding delivery catheter allows correction for the dextrocardia and renders this technique feasible in these patients.

Case report

A 79-year-old female patient diagnosed with hypertension, diabetes mellitus, permanent AF, situs ambiguous with dextrocardia, interrupted infrahepatic IVC with azygos continuation, left bronchial isomerism, and polysplenia was seen in the clinic for highly symptomatic AF with suboptimal rate control. She had failed rhythm control with direct current cardioversion and amiodarone. Even though AF ablation in dextrocardia was shown to be feasible in multiple reports, both percutaneous and thoracoscopic,[3,4] our patient preferred to forgo pulmonary vein isolation. Her wishes were to reach a definitive solution for her condition and avoid the possibility of having to undergo a repeat AF ablation. She presented to our center for AVNA and pacemaker implantation. She had a normal LV ejection fraction and dimensions. To avoid developing right ventricular (RV) pacing–induced cardiomyopathy, we opted to implant an LBBAP permanent pacemaker, in addition to a backup RV septal pacing lead.

Given the dextrocardia, surface electrocardiographic leads were inverted limb leads (i.I, i.II, iIII, iaVR, iaVL) and right-sided precordial leads. These were displayed and recorded by the electrophysiological laboratory recording system (Prucka Cardio-Lab IT system; GE Healthcare, Little Chalfont, UK).

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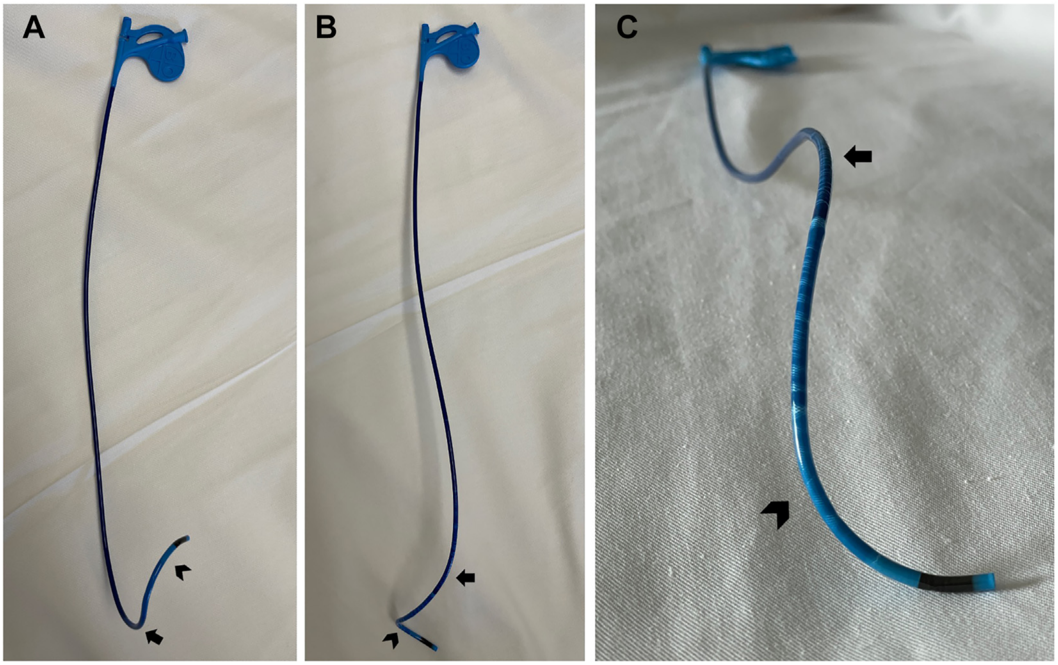

Figure 1 **A:** Original shape of the Selectra 3D delivery sheath (Biotronik, SE & Co, KG, Berlin, Germany). **B, C:** Modified shape of the sheath done by reversing the primary curve (*solid arrow*) and posteriorly directed reshaping of the secondary curve (*arrowhead*).

Contrast venogram via a left arm vein was performed prior to the incision and showed a patent left subclavian vein and a left superior vena cava (SVC) draining into the morphologic right atrium. Vascular access was obtained via the left cephalic vein and a guidewire was advanced into the right ventricle. A Selectra 3D delivery sheath (Biotronik, SE & Co, KG, Berlin, Germany) was manually reshaped with the dilator in place to reverse the primary curve and correct for the dextrocardia. The secondary curve required additional posteriorly directed reshaping to achieve a perpendicular orientation to the septum (Figure 1A–1C). A Solia S60 (Biotronik, SE & Co, KG, Berlin, Germany) pacemaker lead was advanced through the delivery sheath and the lead-sheath system was positioned against the septum as seen in the right anterior oblique projection (true left anterior oblique). When the paced morphology was showing a notched QRS in V_1 ("W" pattern), the pacemaker lead was advanced into the septum until a Purkinje potential was seen on the lead electrogram, fixation beats with a right bundle branch block morphology were seen, the impedances recorded on the lead peaked and fell, the QRS narrowed to 110 ms, and V_6 R-wave peak time (RWPT) shortened to 76 ms (Figure 2A). At this location, capture threshold was satisfactory at 0.9 V @ 0.4 ms and an R-wave sensing of 9.4 mV. Additionally, a backup pacemaker lead (Biotronik Solia S60) was fixated in the low RV septum with adequate threshold, sensing, and impedance. The LBBAP lead was then connected to the LV port of the biventricular pacemaker pulse generator (Edora HF-T cardiac resynchronization therapy [CRT] pacemaker; Biotronik, SE & Co, KG, Berlin, Germany), the RV lead was connected to the RV port, and the atrial port was plugged in to maintain magnetic resonance imaging compatibility.

Subsequently, a left internal jugular (IJ) venous access was obtained under ultrasound guidance and an 11F sheath (11 cm) was inserted. A contact force irrigated 3.5 mm STSF ablation catheter (Biosense Webster, Inc, Irvine, CA) was used to create a 3D electroanatomic map (EAM) of the morphologic right ventricle, morphologic right atrium, and SVC. In addition, a computed tomography reconstruction was merged with the EAM to complete the morphologic right ventricle, RV outflow tract, pulmonary artery, and morphologic left atrium. A His potential was recorded on the distal tip of the ablation catheter and marked on the EAM (Figure 3A). At this location, radiofrequency energy was applied for 2 minutes at an average of 37 W, resulting in complete atrioventricular (AV) block and initiation of backup VVI pacing at 40 beats per minute (bpm). AV block was confirmed after a 30-minute waiting period and the device was set at VVI 80 bpm. Importantly, the LBBAP lead was set to pace 80 ms earlier than the RV septal lead. A chest radiograph showed stable lead positions and no complications. The patient returned 1 month later to the clinic with a significant improvement in her symptoms. The LBBAP lead threshold was 1.3 V @ 0.4 ms; she remained in complete AV block and V-pacing 100% of the time. Her electrocardiogram at follow-up showed a narrow QRS and a short V_6 RWPT, identical to her native QRS preablation (Figure 2B and 2C). At that time, the device was programmed to VVIR 60 bpm.

Discussion

AF is the most common cardiac arrhythmia, affecting more than 3 million people in the United States alone and projected

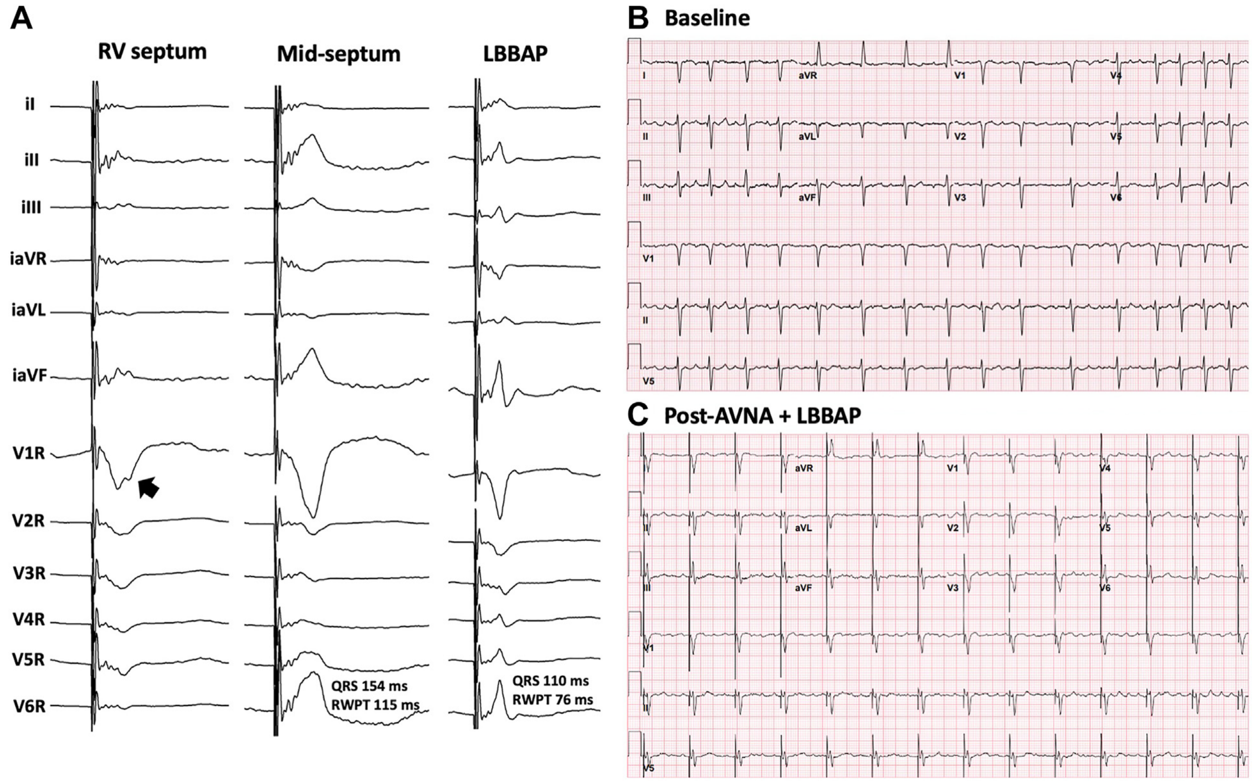

Figure 2 **A:** Twelve-lead electrocardiograms (ECG) with inverted limb leads and right-sided precordial leads of the paced QRS. At the level of right ventricular (RV) septum, note the "W" morphology in lead V_1 (*solid arrow*). When the lead is screwed in the septum, note the loss of the notch in V_1 while V_6 R-wave peak time (RWPT) remains prolonged (115 ms) and QRS is still wide (154 ms). Nonselective left bundle branch pacing, with shortening of V_6 RWPT to 76 ms and QRS width to 110 ms. **B:** Baseline ECG in atrial fibrillation. **C:** ECG following atrioventricular node ablation (AVNA) and left bundle branch area pacing (LBBAP). Note the similarity of the QRS morphology, width, and axis between the native conduction in panel A and paced rhythm in panel B.

to reach 16 million by 2050.[5] Although pharmacotherapy remains the first-line treatment for rate control of AF, ablation of the AV node with a pacemaker implantation, the so-called "ablate and pace" strategy, is a viable option for patients with drug-refractory symptoms.

Dextrocardia with interrupted IVC poses a significant anatomical challenge for cardiac ablations. Accessing the heart in these patients can be achieved either through a femoral then azygos vein or through a subclavian or internal jugular vein access, also known as the superior approach. Additionally, for AVNA a left-sided retrograde transaortic approach through a femoral artery access is feasible, but it is usually reserved for challenging cases when AV block is not achieved through the conventional right-sided approach.[6]

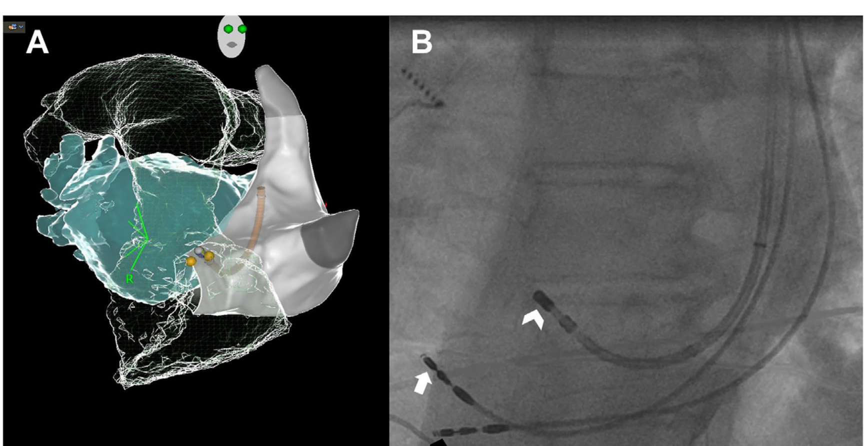

Figure 3 **A:** Left anterior oblique (LAO) view equivalent to a "true" right anterior oblique (RAO) 3-dimensional electroanatomic map merged with a reconstructed computed tomography scan of the heart. The ablation catheter is positioned at the His potential and radiofrequency energy is applied, resulting in complete atrioventricular block. **B:** LAO 17 degrees ("true" RAO) fluoroscopic projection showing the relative positions of the ablation catheter (*white arrowhead*) to the left bundle branch pacing lead (*white solid arrow*) and right ventricular septal lead (*black arrowhead*).

Kelesidis and colleagues[7] reported a case of successful AVNA via the azygos vein in a patient with dextrocardia and interrupted IVC.

Moreover, there have been multiple previous reports of successful slow pathway ablation using the superior approach in patients with interrupted IVC or IVC filters.[8,9]

In recent years, the LBBAP technique has gained remarkable enthusiasm in the world of physiologic pacing. Although the physiological advantages from the recruitment of the intrinsic conduction system are compelling, the long-term outcomes of this novel technique and the rates of lead failure remain unknown. For this reason, we also elected to implant a conventional pacing lead in the low septum. The CRT-pacemaker was programmed with an LV lead offset of 80 ms, causing the conventional lead to pace in the ventricular refractory period to provide backup pacing in case of LBBAP lead failure.

LBBAP in patients with dextrocardia using the lumenless Medtronic SelectSecure 3830 lead has been recently reported.[10,11] Our case is the first to report a successful implant of a standard stylet-driven lead with an extendable helix design, the Biotronik Solia S60, in a patient with dextrocardia. LBBAP using a stylet-driven lead was shown to be comparable to the lumenless lead in terms of procedural and fluoroscopic times, success rates, and lead parameters.[12] The delivery sheath necessitated manual reshaping to correct for the dextrocardia. Our workflow consisted of initially pace mapping the right-sided septum while the helix is withdrawn, followed by extending the helix at the desired location on the septum, then advancing the green stylet guide and finally rotating the body of the lead about 10–15 times to penetrate deep in the septum.[13]

LBBAP implantation is guided by pace mapping and radiological anatomy. The insertion site of an LBBAP lead is typically 15–20 mm apical from the His bundle potential, which is approximated radiographically as the tricuspid valve summit. At this site, the paced QRS shows a "W" pattern with a notch at the nadir of lead V_1, and discordant QRS between II and III., although concordant positive or negative QRS polarity in II and III can result in successful engagement of the left anterior fascicle and left posterior fascicle, respectively. The lead should then be screwed and advanced into the septum until the paced QRS becomes narrower and loses notches/slurs, a Qr/qR/rsR′/R appears in V_1, and V_6 RWPT shortens.

LBBAP electrocardiogram criteria have been recently described in a consensus statement.[14] In our case, a left bundle potential was observed on the device analyzer during LBBAP placement. Unfortunately, the strip showing the left bundle branch (LBB) discrete potential was not saved. Later, it was unfeasible to record an LBB discrete potential on the electrogram of the LBBAP lead given the therapeutic AV block. The gold standard to assess LBB capture is the transition in the QRS morphology and a sudden increase in V_6 RWPT ($\geq$15 ms) during threshold testing. This criterion exploits the differences in excitability and/or refractoriness between the conduction system and adjacent myocardium. With decremental voltage output one should expect a transition from simultaneous capture of the LBB and septal myocardium (nonselective LBB pacing) to selective capture of either the LBB only (selective LBB pacing) or septal myocardium only (left ventricular septal pacing).[14] Also, Jastrzębski and colleagues[15] showed that in patients with a narrow QRS/right bundle branch block, a V_6 RWPT $<$74 ms was 100% specific for LBB capture. Hence, our case probably demonstrates a nonselective LBB capture given a short V_6 RWPT of 76 ms and QRS duration of 110 ms.

To our knowledge, this is the first case report of a successful AV node ablation in a dextrocardia patient performed from a superior approach via the left IJ vein with a successful LBBAP implantation. The only other case of AVNA was performed via the azygos continuation of the IVC in a patient with preexisting dual-chamber pacemaker.[7] Our preprocedural venogram revealed a patent left SVC draining into the morphologic right atrium. We considered this approach more straightforward for AV node ablation when compared to access from below, which would require manipulation through the azygos vein to reach the AV node.

Conclusion

AVNA remains an important therapeutic option for refractory AF. This case highlights the feasibility of AVNA via the left IJ vein and an LBBAP implantation in patients with dextrocardia and interruption of the IVC with azygos continuation. CRT has shown mortality benefits over RV apical pacing in the "ablate and pace" patients and should be considered in this population. Finally, LBBAP might be a reasonable alternative to biventricular pacing and is probably associated with similar physiologic advantages, and it is the subject of several ongoing studies.

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