A case of self-diagnosed painful left bundle branch block

Kunal Tandon, MD,* Jennifer Lakey, MSN, RN,† Alexei Shvilkin, MD*

*From the *Harvard-Thorndike Electrophysiology Institute, Cardiovascular Division, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, Massachusetts, and †University of Maine at Augusta, Augusta, Maine.*

Introduction

Painful left bundle branch block (LBBB) syndrome is a rarely recognized condition characterized by chest pain occurring simultaneously with the onset of a rate-dependent LBBB in the absence of myocardial ischemia. Successful treatment of painful LBBB syndrome cases has been described with different pacing modalities (right ventricular, biventricular, and His bundle, as well as left bundle branch [LBB] area pacing).[1–5] We describe a case of a painful LBBB self-diagnosed by the patient and resolved by LBB area pacing, and we highlight the details of its clinical course and pacemaker programming considerations.

KEY TEACHING POINTS

- Painful left bundle branch block (LBBB) is a poorly recognized entity.
- Left bundle area pacing is a successful treatment modality for painful LBBB.
- Rate-adaptive AV delays can be used to optimize pacemaker function.

Case report

A 42-year-old woman, a registered nurse, developed recurrent episodes of exertional chest pain associated with dyspnea in 2015. At that time, she underwent a stress echocardiogram where "borderline LBBB" with QRS <120 ms was noted at a rate of 130 beats per minute (bpm) coinciding with symptoms, and immediate postexercise images showed mild septal dyssynchrony but no evidence of ischemia. The patient gradually decreased her physical activity to avoid recurring chest pain. This initially helped alleviate her symptoms, but eventually the pain occurred with minimal exertion. She was prescribed metoprolol and nitroglycerin, without significant effect. Between 2017 and 2020, she had 8 emergency room visits with diagnoses of "atypical chest pain," "dyspnea," and "deconditioning." She was seen by cardiology both in the in- and outpatient settings and underwent a nuclear stress test, cardiac catheterization, and 2 computed tomography scans of the chest. The only abnormality discovered was a rate-dependent LBBB appearing at the rate of 130 bpm and disappearing at 114 bpm, coinciding with symptoms during exercise stress test in 2017. Given the lack of a definitive diagnosis and treatment progress, the patient performed a literature search, learned about painful LBBB syndrome, and self-referred to us for electrophysiologic evaluation in 2019. At that time, the patient was definitively diagnosed with painful LBBB syndrome and offered physiologic pacing. Initially, she was reluctant to undergo an invasive procedure, but by the end of 2020 the symptoms became debilitating, and the patient decided to proceed.

A dual-chamber pacemaker (Medtronic Azure; Medtronic, Minneapolis, MN) was implanted via the left axillary vein using a C315 His sheath and Medtronic 3830 lead (Medtronic, Minneapolis, MN). Owing to concerns about long-term lead performance because of the patient's age and difficult anatomy necessitating a distal venous access site, we chose LBB area rather than His bundle pacing (Supplemental Figure 1). A left bundle potential was recorded at the implantation site preceding the QRS by 16 ms and left ventricular activation time during pacing closely matched that in sinus rhythm (Figure 1). In the laboratory, LBBB occurred with atrial pacing at 90 bpm with QRS duration of 120 ms, which did not change when pacing at 140 bpm. The device was programmed initially in DDD mode with baseline paced/sensed AV delays of 160/150 ms, which achieved ventricular capture with near normalization of QRS duration and typical incomplete right bundle branch block pattern during pure ventricular pacing (Figure 2A). Rate-adaptive AV delay was enabled with a start

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Figure 1 From left to right can be seen 2 intrinsic QRS beats with left ventricular activation time (LVAT) of 61 ms and then, on the right, left bundle branch area pacing with LVAT time of 63 ms (almost identical to sinus).

rate of 90 bpm, stop rate of 130 bpm, and minimum paced/sensed AV delays of 120/110 ms. In the following weeks, the patient felt minimal improvement. Carelink transmissions revealed ventricular pacing less than 24% of the time, mostly during atrial pacing at the lower rate limit (Figure 2B). During the pacemaker reprogramming, the sensed AV delay required to achieve full LBB capture during sinus rhythm at resting heart rate (HR) was quite short (70 ms) and the patient felt chest discomfort and pulsations in the neck, possibly owing to pacemaker syndrome. Baseline paced/sensed AV delays were

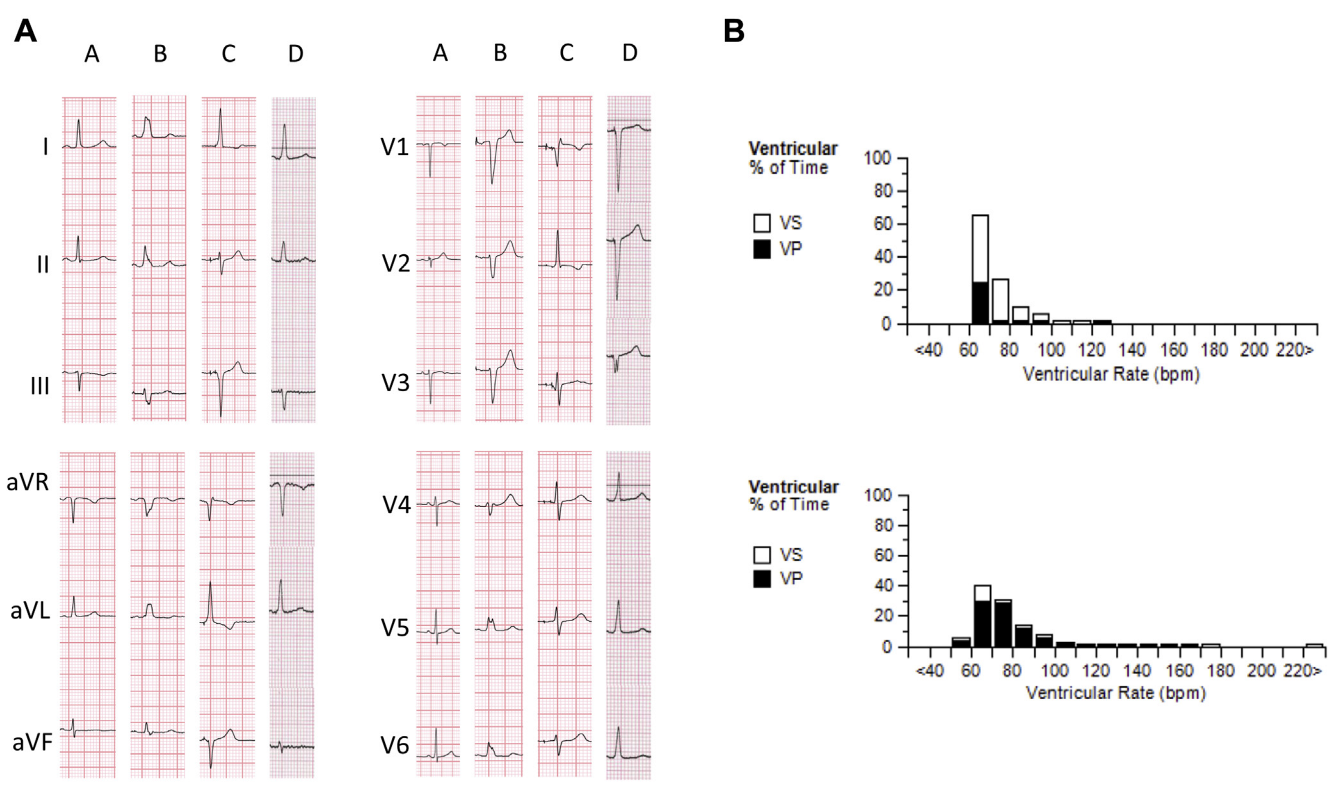

Figure 2 **A:** Column A - narrow intrinsic QRS with sinus rate ∼60 beats per minute (bpm); column B - typical left bundle branch block with atrial pacing at 85 bpm; column C – DDD pacing with a short AV delay allows selective left bundle capture with typical qR morphology in V_1; column D – DDD pacing with a longer AV delay shows fusion of intrinsic His-Purkinje conduction and left bundle pacing resulting in narrow QRS. **B:** Pacemaker interrogation with initial settings (top) showing high percentage of ventricular sensing (VS) and final settings after adjustment of rate-adaptive AV with now predominantly ventricular pacing (VP) after adjustment (bottom).

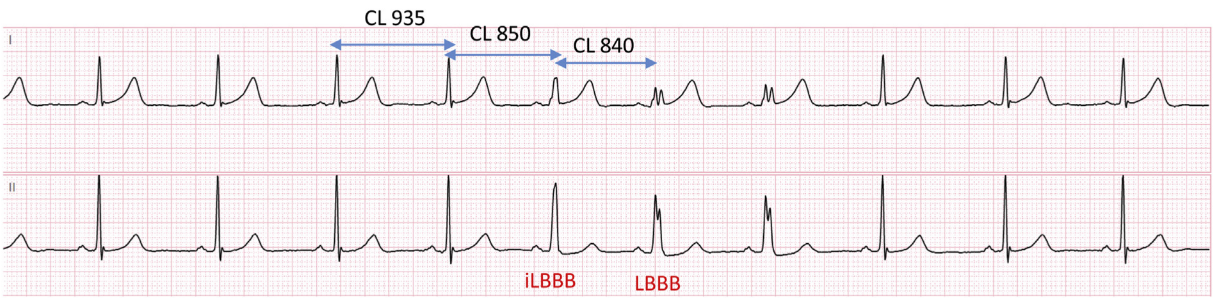

Figure 3 Mobile cardiac telemetry showing the development of initially incomplete left bundle branch block (iLBBB) and then complete left bundle branch block (LBBB) with progressive sinus rate acceleration at cycle length (CL) of 850 and 840 ms, respectively.

decreased to 130/100 ms with minimum rate-adaptive AV delays of 100/80 ms (start/stop HR 85/140 bpm). With this programming the patient felt much better during moderate physical activity but "hit the wall" during strenuous exercise as well as continued to have intermittent vague symptoms of chest discomfort at rest and the beginning of exercise. A KardiaMobile (AliveCor, Mountain View, CA) recording showed intermittently wide QRS at 72 bpm. Device interrogation showed the presence of ventricular sensed rhythm between 60 and 100 bpm and over 160 bpm, which was the upper tracking rate. Mobile cardiac telemetry (MCT) monitoring was performed and demonstrated multiple short periods of incomplete and complete LBBB development at 70 bpm (Figure 3) and below, as well as loss of LBB pacing at upper tracking rate corresponding to onset of exertional chest pain.

The AV delay adjustment was performed while observing electrogram configuration on the portable iPad Medtronic programmer with the patient performing physical activity (walking/brisk walking) outside the office while reporting symptoms in real time. The final pacemaker parameters were DDD mode 50–175 bpm with a baseline paced/sensed AV delay of 170/100 ms and a rate-adaptive paced/sensed AV delay of 120/70 ms with a start/stop rate 60/70 bpm. Since the final adjustment, the patient has reported no further symptoms and has resumed normal levels of activity, resulting in an improved quality of life.

Discussion

Despite its description at least 45 years ago,[6] painful LBBB is rarely diagnosed owing to the lack of awareness of this condition. Patients typically undergo extensive diagnostic evaluation and often treatment for incident coronary artery disease.[1] In our case, a well-educated patient self-diagnosed and self-referred for electrophysiologic evaluation after she could not find help from her cardiology providers for several years. The following features of painful LBBB are highlighted by our case:

(1) Painful LBBB can occur at relatively narrow QRS (120 ms in our case).
(2) The symptom intensity can vary depending on the level of exertion/heart rate. When it occurred at rest and a lower HR the patient described symptoms as "discomfort," but during exertion described them as "pain," despite identical QRS width and morphology.
(3) The LBBB onset rate decreases over time (in our case from 130 bpm in 2017 to 70 bpm and below) and can vary depending on the physiologic conditions. While it was 90 bpm during atrial pacing, MCT monitoring demonstrated intermittent LBBB at rates as slow as 63 bpm, which can create fleeting and difficult-to-characterize symptoms.
(4) AV delays required to relieve LBBB can be quite short and may result in pacemaker syndrome symptoms, particularly at slow heart rates.
(5) In cases of LBB area pacing, rate-adaptive AV delays with a sharp, step-wise decrement around the anticipated LBBB onset rate are useful to allow native conduction during narrow QRS morphology as well as complete LBBB relief at faster rates. The "rate-adaptive" AV delay feature on the Medtronic pacemakers has 4 programming settings: start rate, stop rate, and minimum sensed and paced AV delays. The AV delay will linearly decrease from the baseline at the start rate to the minimum AV delay at the stop heart rate and remain constant for all faster heart rates. Use of the HR histogram feature can help determine at what heart rates ventricular sensing or pacing are occurring so as to determine at what heart rates a change in AV delay is required.
(6) The use of an MCT monitor, a wearable consumer electrocardiogram monitor, and a Bluetooth-connected portable programmer were helpful to capture transient symptoms as well as to appropriately program the pacemaker during real-life activities.

Conclusion

Despite an increase in publications in the past few years, painful LBBB syndrome remains a rarely diagnosed condition. In contrast to medical management that is largely ineffective, ventricular resynchronization provides consistent symptomatic relief in all published reports. While LBB area pacing provides superior lead pacing characteristics as compared to His bundle pacing, careful programming of the AV delay is required to optimize its effect.

Appendix

Supplementary data

Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2021.09.005.

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