Phase 4 block of the right bundle branch suggesting His-Purkinje system involvement in Lyme carditis

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Introduction

Lyme disease due to *Borrelia burgdorferi* infection causes a wide variety of signs and symptoms, including cardiac involvement termed Lyme carditis (LC).[1–3] Though uncommon, LC is a well-established cause of atrioventricular (AV) block, typically at the AV node level, with infrequent involvement of the infranodal conduction system.[1]

We describe an adolescent with symptomatic high-grade AV block and evidence of phase 4 right bundle branch block (RBBB), a previously unreported manifestation of infranodal conduction system involvement in LC.

KEY TEACHING POINTS

- Lyme disease may present with acute symptomatic high-grade atrioventricular (AV) block, even without identified tic exposure or erythema migrans.
- Lyme carditis usually is confined to the AV node with narrow QRS on conducted or junctional beats.
- His-Purkinje system involvement can occur in Lyme carditis.
- Phase 4 bundle branch block suggests significant His-Purkinje system dysfunction.

Case report

A previously healthy 14-year-old boy presented via emergency medical services in midsummer because of syncope. He reported frequent hunting and fishing in a heavily wooded area with tall grass in a Lyme-endemic region, but could not recall any tick bites or erythema migrans. There had been a mild illness over the 2 weeks prior to presentation, with a runny nose, cough, and abdominal pain, followed by progressive headache and, later, nausea. On the day of presentation, he had 2 non-exertional syncopal episodes, each lasting less than 10 seconds, with no seizure-like activity, incontinence, or post-ictal confusion.

Upon arrival to the emergency department, he had bradycardia, altered mental status, headache, and vomiting. Head computed tomography and routine labs were normal. Initial electrocardiogram (ECG) (Figure 1A) showed complete heart block with a regular wide-complex escape of RBBB-like morphology and right axis deviation, suggestive of either junctional escape with bifascicular block (RBBB and left posterior hemi-block) or left anterior fascicular escape. Telemetry (not shown) demonstrated intermittent paroxysmal high-grade block with transient asystole. Isoproterenol infusion improved the heart rate with establishment of mainly 1:1 conduction with first-degree AV block (Figure 1B). However, second-degree AV block continued to occur at times (Figure 2), with evidence of phase 4 RBBB, suggesting Purkinje system involvement. The combination of isoproterenol side effects (nausea and pounding headache) and potentially unstable conduction system disease prompted urgent placement of a temporary pacing system using an active-fixation transvenous lead connected to an external permanent generator.[4] Isoproterenol was discontinued and the patient's clinical status improved immediately.

Ceftriaxone was started (2000 mg every 24 hours) given the presumptive diagnosis of LC. Further workup showed a structurally normal heart, and no alternative etiologies were identified. There was no evidence of erythema migrans or

KEYWORDS Atrioventricular block; His-Purkinje system; Lyme carditis; Phase 4 block; Right bundle branch block
(Heart Rhythm Case Reports 2021;7:112–116)

Conflict of Interest: The authors declare this manuscript was published in the absence of financial or nonfinancial relationships that could be considered a conflict of interest. Funding: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.
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https://doi.org/10.1016/j.hrcr.2020.11.017

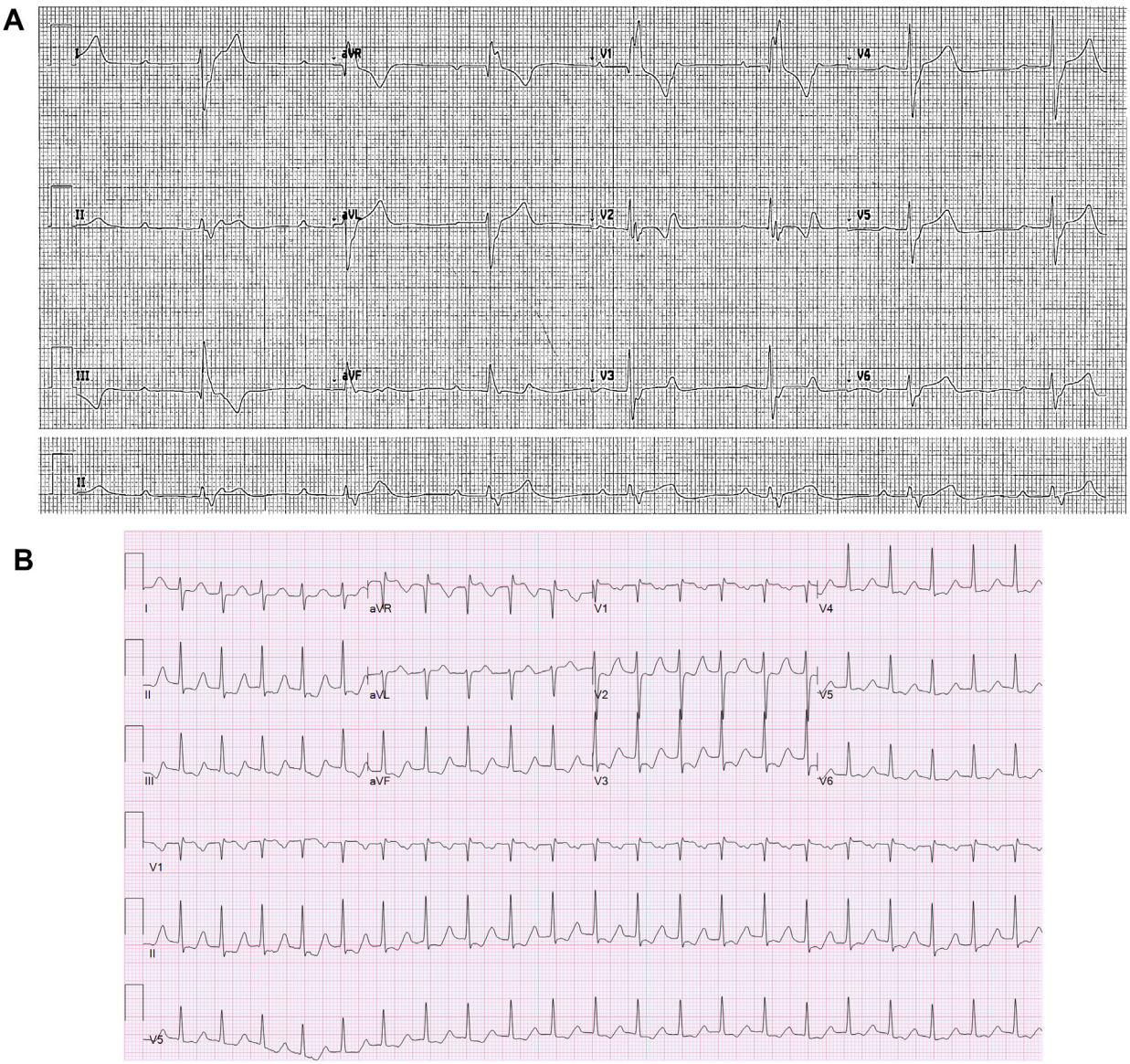

Figure 1 Electrocardiograms. **A:** At presentation, with third-degree block and wide-complex escape at 43 beats/min. **B:** On day 0 after starting isoproterenol, with first-degree block and narrow QRS.

myopericarditis. By the Suspicious Index in Lyme Carditis scoring system, he received an intermediate score of 5 points (owing to constitutional symptoms, outdoor activities in endemic region, male, and young age).[5] On day 4 of admission, Lyme disease serology results supported the clinical suspicion, with positive antibody screen and confirmatory Western blot.

Heart rate support was provided with ventricular demand pacing at 60 beats per minute (bpm). Conduction improved over the subsequent week (Figure 3A), with rapid establishment of mainly 1:1 AV conduction with first-degree block. The pacemaker was reprogrammed to demand pacing at 45 bpm with hysteresis rate of 30 bpm. Following this, he received only sporadic pacing following an occasional nonconducted P wave. The PR interval remained significantly prolonged, sometimes exceeding 400 ms, and right axis deviation persisted. After 5 days of intravenous ceftriaxone, AV conduction was consistently 1:1, with PR intervals ranging from 220 to 240 ms. The temporary pacemaker was removed, and ceftriaxone was transitioned to oral doxycycline with plan to complete a total of 21 days of antibiotics. The PR interval continued to shorten (Figure 3B).

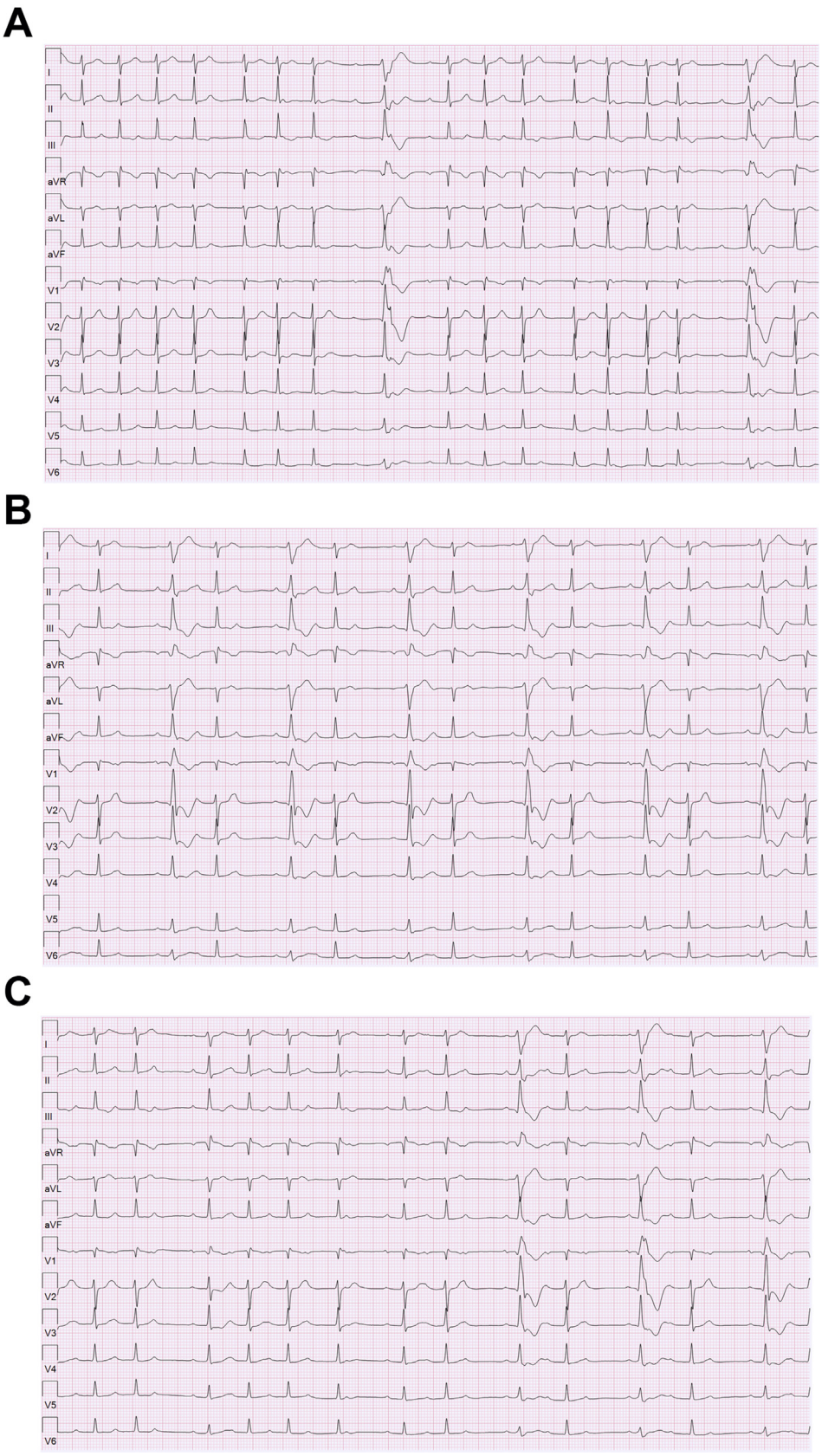

Figure 2 Three additional electrocardiograms on day 0, showing fluctuating conduction abnormalities despite isoproterenol. **A:** Variable atrioventricular (AV) block with either fascicular escape beats or post-pause right bundle branch block (RBBB). **B:** Variable AV block with intermittent wide-complex beats of several morphologies, either fascicular escape beats with variable fusion or variable post-pause RBBB. **C:** Second-degree 3:2 AV block with repetitive post-pause RBBB.

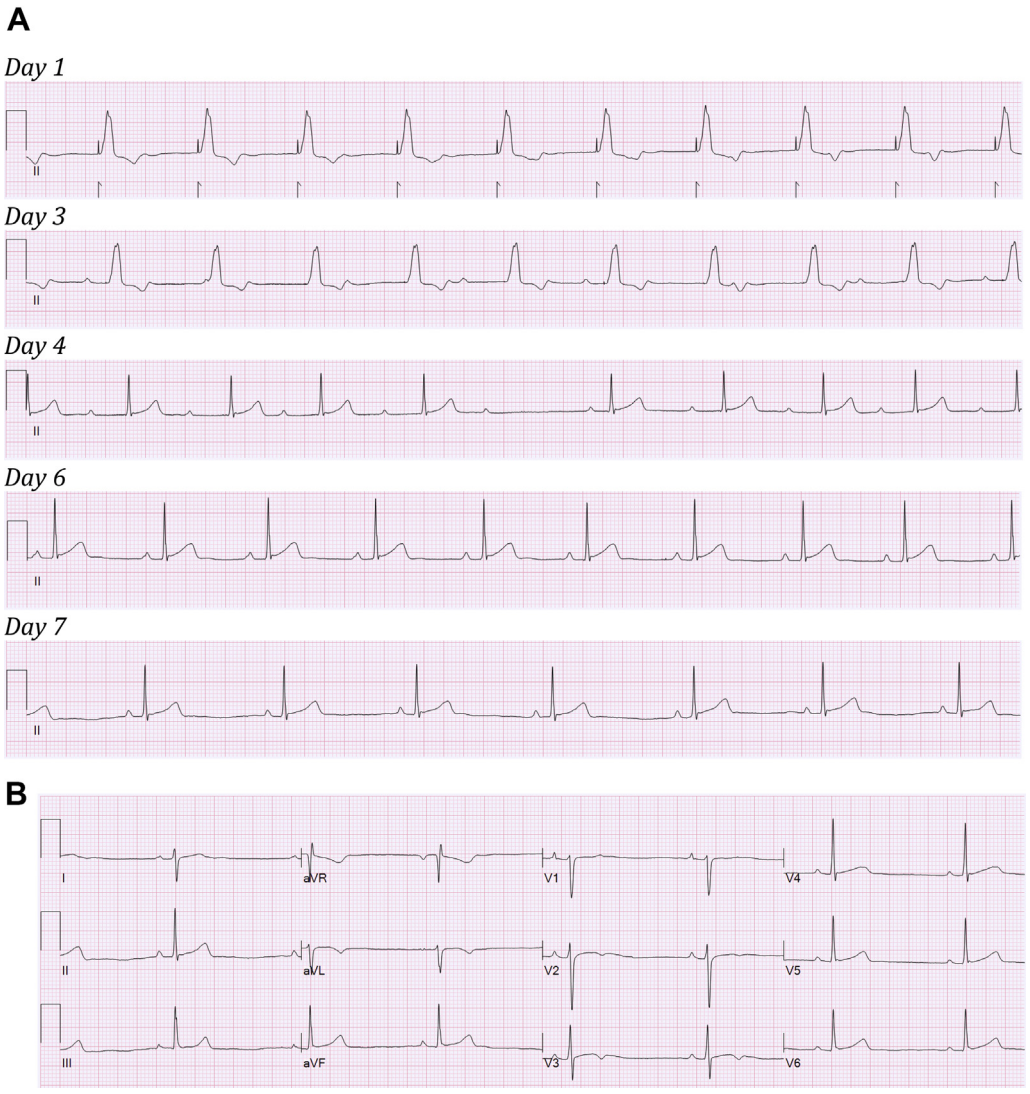

Figure 3 **A:** Sequential rhythm strips during hospitalization, with improving conduction. **B:** Electrocardiogram at discharge, with sinus bradycardia and right axis deviation.

He was monitored in the hospital for a total of 7 days and then discharged with an auto-detecting wireless ambulatory monitor for continuous surveillance until outpatient follow-up appointment and stress test. The monitor showed no pauses, and in fact demonstrated normal AV conduction up to heart rates in the 190s (while playing soccer, despite recommendation for exercise restriction while recuperating). In outpatient follow-up 2 weeks later, the patient was well, with completely normal ECG (not shown); the prior right axis deviation had resolved.

Discussion

Significant AV block due to LC occurs in about 1% of Lyme disease cases.[1,2] In LC, the AV node is most commonly affected, with bundle branch block/intraventricular conduction delay reported in only 13% of cases. Some evidence from invasive electrophysiology studies shows that block most often occurs within the AV node, above the level of the His bundle.[1,2]

We report an adolescent with acute symptomatic high-grade AV block and intermittent RBBB. ECGs demonstrate narrow QRS at faster rates, with RBBB during bradycardia and after longer R-R intervals, consistent with phase 4 block within the right bundle branch.

Phase 4 block is attributed to hypopolarization or spontaneous depolarization in phase 4 of the action potential in response to a preceding pause, typically occurring in diseased His-Purkinje fibers.[6–11] Either mechanism precludes myocardial cells from being at the resting membrane potential when the next action potential arrives and therefore will result in a failure to depolarize when stimulated.

Phase 4 block is most often identified when it produces high-grade AV block. Phase 4 bundle branch block is uncommon.[6,7,9,12] We believe this is the first reported case of phase 4 bundle branch block in LC.

The phase 4 bundle branch block strongly suggests His-Purkinje system involvement by direct spirochete invasion or inflammation, which has been reported very rarely.[1] However, we performed neither invasive electrophysiology testing nor cardiac magnetic resonance imaging, either of which might have been able to identify or localize inflamed tissue.[1,13]

Conclusion

Conduction system involvement from Lyme carditis is usually confined to the AV node. We present a case with high-grade AV block and phase 4 right bundle branch block, suggesting His-Purkinje system involvement. Our patient was successfully treated with temporary transvenous pacing and antibiotics, with dramatic improvement within days and complete normalization within weeks.

Acknowledgments

We appreciate the assistance of Dr Megan McGreevy with this case.

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