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Rina Shah, MD

Thomas Jefferson University

Mitul Kanzaria, MD

Thomas Jefferson University

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A CASE SERIES OF LYME CARDITIS WITH COMPLETE HEART BLOCK

Rina Shah, MD and Mitul Kanzaria, MD

Case 1

A 25-year-old Caucasian male with a history of a benign functional heart murmur presented to the emergency department for evaluation of 10 days of dyspnea with minimal exertion, such as walking a few blocks or climbing a flight of steps. Dyspnea was accompanied by moderate, dull, left-sided chest pain, which had been progressively worsening over the previous 3-5 days. The symptoms initially occurred only with exertion, but progressed to last 30-45 minutes after discontinuation of activity. He also noticed palpitations during these episodes, but did not try any remedies for his symptoms. Of note, two weeks prior to presentation, the patient had been camping in Shenandoah National Park, Virginia for four days. During that trip, he sustained over 30 mosquito bites, but never noticed any tick bites or rashes. He also traveled to Brazil five months ago, during which time he took malaria prophylaxis.

Upon review of systems, the patient complained of generalized fatigue, fevers and chills for a few days after returning from his camping trip. He also noted muscle soreness, most notably around his chest secondary to heavy lifting, as well as post exercise myalgias. He denied syncope or pre-syncope symptoms.

The patient was a social drinker and occasional marijuana user, but denied tobacco or other illicit drug use. He worked as a beer vendor for a baseball stadium and his work often involved heavy lifting. His father and uncle both had myocardial infarctions in their early 50s.

On presentation, the patient had a blood pressure of 156/64 and a heart rate ranging from 43-52 beats per minute (bpm). His cardiovascular exam revealed cannon A waves and a 1/6 systolic murmur over his precordium, but no diastolic murmurs. His physical exam was also remarkable for multiple mosquito bites over both legs. A thorough skin exam revealed no rashes or lesions, and a neurologic exam was unremarkable.

His lab work returned within normal limits and his toxicology screen was only positive for cannabinoids. A resting ECG (Figure 1) revealed complete heart block with an atrial rate of 72 bpm, a narrow QRS escape rhythm of 55 bpm and a normal QT interval. An echocardiogram revealed normal left ventricular systolic function without wall motion abnormalities.

The patient was admitted to the cardiology service and monitored on telemetry. He did not require transcutaneous pacing. Lyme titers were sent and he was started on intravenous (IV) ceftriaxone for presumed Lyme carditis. His titers subsequently returned positive for both IgM and IgG. During his hospital stay, he remained in complete heart block and his escape rate dropped as low as 30 bpm during sleep, but his blood pressure always remained stable. His symptoms began to improve within one day of antibiotics and his complete heart block regressed to Mobitz Type II heart block within less than 24 hours. Over his four-day hospitalization, the patient continued to have gradual improvement in his heart block, as illustrated by several ECG and telemetry strips (Figures 2, 3 and 4). The patient's

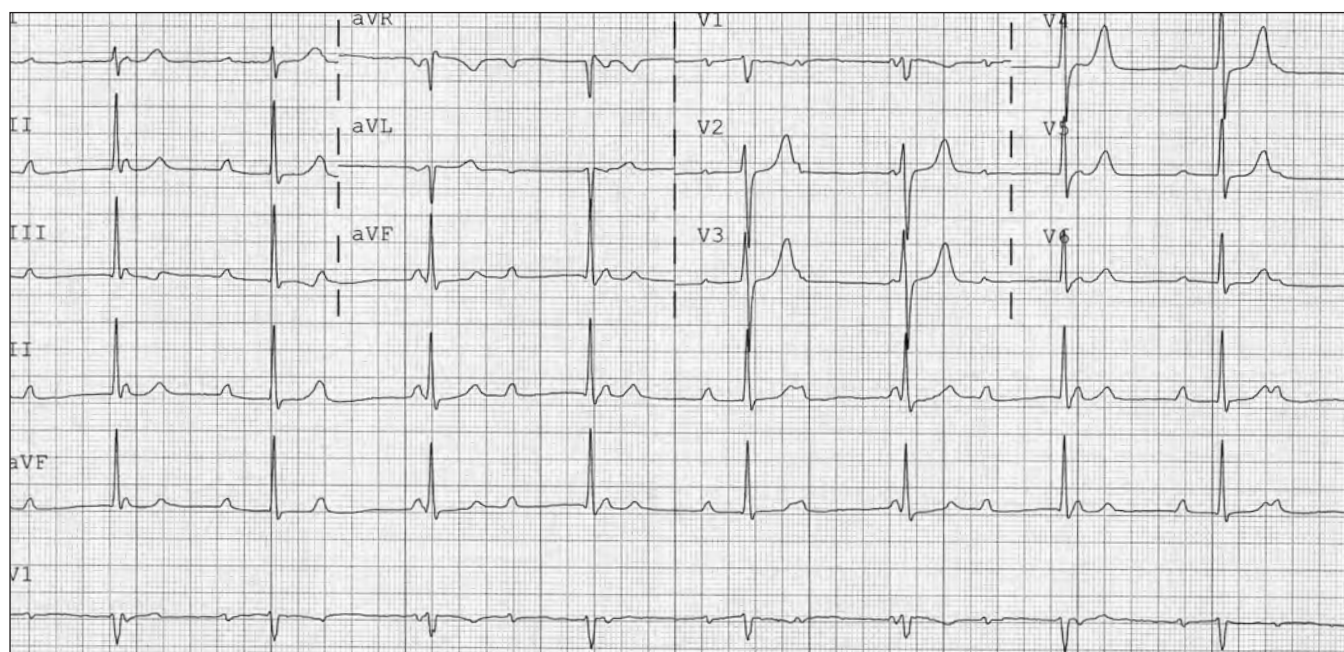


Figure 1. Patient's ECG revealing high degree of atrioventricular (AV) block with narrow QRS escape rhythm



Figure 2. Patient's telemetry strip revealing a Mobitz I AV block

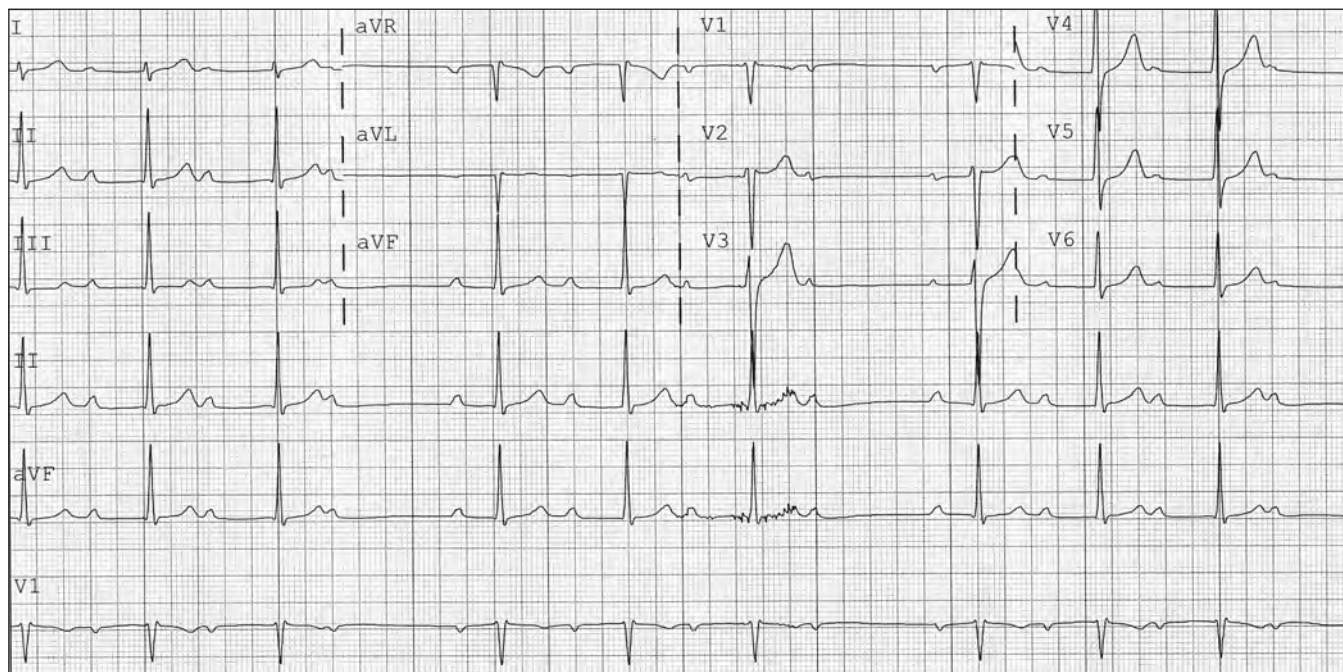


Figure 3. Patient's ECG revealing Mobitz I AV block



Figure 4. Patient's telemetry strip revealing a 1st degree AV block

symptoms also improved and after four days of IV ceftriaxone, his conduction became 1:1 with marked PR prolongation. Once AV conduction resumed, the patient was switched from IV ceftriaxone to oral doxycycline and was stable for discharge.

The patient returned for a follow up visit at the Arrhythmia clinic and reported complete recovery of his exercise capacity with no further episodes of chest pain or dyspnea. His heart rate normalized to a sinus rhythm at a rate of 59 bpm (Figure 5).

Case 2

A 49-year-old male presented to his primary care provider with a two-week history of decreased exercise tolerance after noticing he was unable to complete his normal 5 mile bike ride to work. He also reported measuring his pulse as 40 beats per minute and had an episode of pre-syncope with exertion. On review of symptoms, the patient denied any recollection of recent tick bites or fevers, chills or myalgias. He did experience mild hip and knee stiffness at the onset of his symptoms. The patient had recently

traveled through rural England for three weeks, and noted that his symptoms started three days after his return home. He also spent a few days in the Poconos about six weeks ago. Aside from a band-like vesicular rash on his thorax that was diagnosed and treated as shingles five months ago, he recalled no other rashes. He was on no chronic medications, did not have any other significant past medical history, and denied any tobacco or alcohol use. Upon review of systems, he denied any syncopal events.

In the outpatient office, an ECG showed the patient to be in 3rd degree AV block. After being referred to a cardiologist, the patient was admitted to the hospital with symptomatic bradycardia. The ECG at that time showed 2nd degree Mobitz 2:1 AV heart block. On admission, his blood pressure was 123/54, heart rate was 41 bpm, and temperature was 98.1° F. His cardiovascular exam demonstrated bradycardia but no murmurs. No rash was noted on a thorough skin examination and his musculoskeletal and neurologic exams were unremarkable. After admission, an exercise-ECG stress test was non-diagnostic for ischemia due to

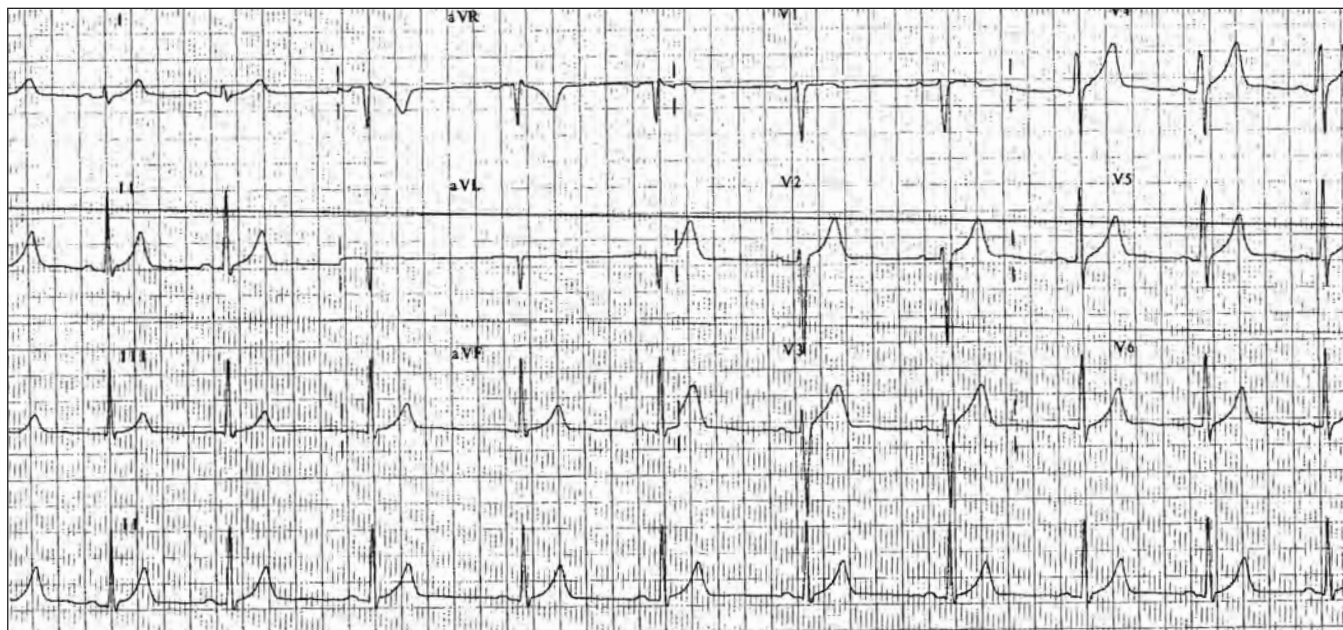


Figure 5. Patient's outpatient ECG revealing a return to sinus rhythm

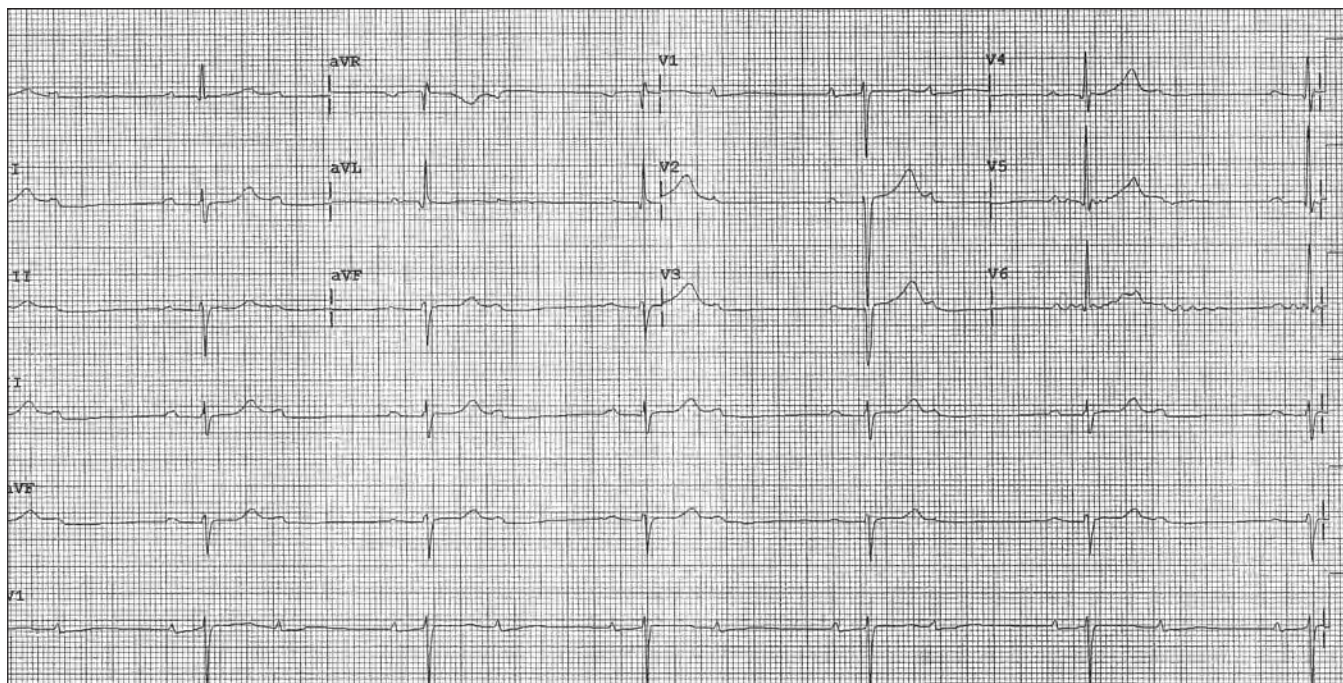


Figure 6. Patient's ECG revealing Mobitz I 2:1 AV block

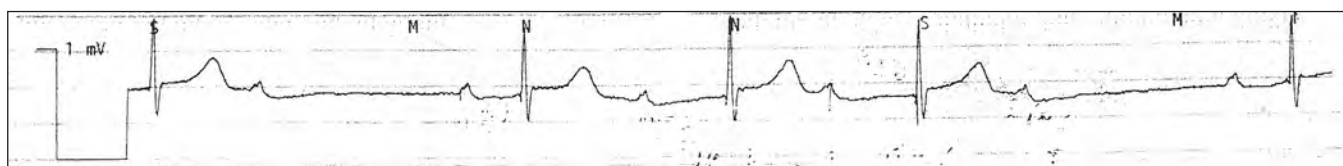


Figure 7. Patient's telemetry strip revealing Mobitz II AV block

the maximum heart rate of 83 bpm with 2nd degree AV block, but the patient achieved 13 metabolic equivalents (METs). A cardiac MRI did not demonstrate an infiltrative disease. ECGs and rhythm strips during the hospital stay showed periods of 2nd degree 2:1 AV block (Figure 6), and 2nd degree Mobitz type II AV block (Figure 7).

The patient was discharged with a plan to get a pacemaker as an outpatient. After discharge, his Lyme IgG and IgM titers were positive by ELISA and western blot, and the patient was readmitted to initiate parenteral antibiotic treatment. He was started on IV ceftriaxone and his heart rhythm resolved to 1st degree AV block within two days at the time of discharge. He was sent home with treatment with IV ceftriaxone 2 grams daily for two weeks and was to follow up with cardiology and infectious disease as an outpatient to determine if he could then convert to an oral regimen to complete his antibiotic course.

Discussion

Lyme disease was first recognized as a clinical entity in the mid-1970s, after the discovery of unexplained arthritis in a group of children in Lyme, Connecticut.^{1,5} There are about 15,000 cases of Lyme disease per year, making it one of the most common tick-borne illnesses in the United States (US).^{1,4,7} This multi-organ disease is caused by the spirochete pathogen *Borrelia burgdorferi* *sensu lato* group and is spread via the vector *Ixodes scapularis*. There are three distinct species: *B. burgdorferi* (*sensu stricto* and *sensu lato*), *B. garinii*, and *B. afzelii*.^{1,7} Although nearly all cases of Lyme reported in the US have been attributed to *B. burgdorferi*, most of the illnesses in Europe have been caused by *B. garinii* and *B. afzelii*. These latter two species are respectively responsible for lymphocytoma and acrodermatitis chronica atrophicans.¹

The clinical manifestation of Lyme disease can be categorized into three phases: *early local*, *early disseminated*, and *late*. The *early local phase* is defined by a 7-14 day incubation period and patients typically present with erythema migrans and systemic symptoms. During the *early disseminated* phase, usually 3-10 weeks after the initial infection, patients may develop multiple erythema migrans lesions, facial nerve palsies, aseptic meningitis or carditis. The *late* phase is typically defined by arthritis involving large joints. Some of the most common findings of Lyme borreliosis are limited to the skin, as noted in Table 1. The extra-cutaneous manifestations, including myocarditis, neuroborreliosis, and arthritis, are further outlined in Table 2.

Lyme disease has a similar prevalence in males and females and affects people of all ages. While some studies have shown Lyme carditis to have a 3:1 female predominance, others have demonstrated a 3:1 male predominance.^{1,6} Cardiac manifestations of Lyme disease are extremely rare. The prevalence of Lyme carditis varies from study to study. While some case series have illustrated an incidence of less than 1% in the US and less than 4% in Europe, other studies have quoted incidences as

Table 1. Cutaneous Findings¹

Erythema migrans	Expanding erythematous patch (>5cm)
Lymphocytoma cutis	Painless bluish-red tumor-like nodule or plaque near ear lobe, nipple or scrotum
Acrodermatitis chronica atrophicans	Bluish-red atrophic skin discoloration on extensor surfaces of extremities

Table 2. Extracutaneous Findings¹

Carditis	Acute onset of AV block and subtle myocarditis (days to weeks)
Neuroborreliosis	Lymphocytic meningitis, cranial neuritis (gen facial nerve palsy), radiculoneuropathy, and rarely encephalomyelitis
Arthritis	Recurrent brief attacks of swelling in one or more joints, usually one or both knees, treatment resistant (weeks to months)

high as 10% of untreated patients infected with *B. burgdorferi* as having cardiac complications.^{1-3,5,7} Cardiac complications include fluctuating degrees of AV block, myocarditis, mild left ventricular dysfunction, and rarely, cardiomegaly or fatal pericarditis.⁷ Chronic cases of Lyme have led to dilated cardiomyopathy in Europe, but there have been no reported cases of cardiomyopathies secondary to Lyme disease in the US.^{1,7} The most common cardiac manifestation of Lyme disease is fluctuating AV block.^{1-3,5} Almost all (98%) adults with Lyme carditis will manifest 1st degree AV block, and of those patients, up to 50% may then progress to complete heart block.³ These symptoms typically present during the early disseminated phase of the disease but can occur anywhere from several weeks and up to three months after the initial infection.^{1,5} During this time frame, patients may also have concomitant neurologic or arthritic involvement. In a study by Bartnek et al that involved 221 cases over a course of several years, early antibiotic therapy was found to prevent late cardiac involvement in nearly every case.⁴

The mechanism by which Lyme diseases affects the conduction system is thought to be related to either the dissemination of spirochetes into the cardiac tissue or an inflammatory response associated with the infection.^{4,6} Animal studies have illustrated macrophage and lymphocyte predominant transmural inflammation of the connective tissue, most notably at the base of the heart, basal interventricular septum, and perivascular regions. The site of the conduction block occurs at or above the AV node, although sinoatrial and intra-atrial conduction disturbances have also been reported.⁶

Diagnosing Lyme depends on the stage of Lyme disease. The early local phase is a diagnosis best made clinically based on a history of a tick bite or classic erythema migrans rash, because antibodies against *B. burgdorferi* are insensitive during the first few weeks of infection.^{3,7}

Meanwhile, patients suspected of having *early disseminated* or *late* disease should be diagnosed clinically and serologically.³ Enzyme-linked immunosorbent assay (ELISA) against serum IgG and IgM antibodies to *B. burgdorferi*, as well as Western blot assays should be ordered to confirm the diagnosis.^{1,4} Another useful tool in the evaluation of Lyme myocarditis is gadolinium-enhanced cardiac magnetic resonance imaging (cardiac MRI). Cardiac MRI will illustrate a subtle increased signal on T2-weighted echo images in the mid-wall region of the basal anteroseptal wall, which is in close proximity to the location of the AV node. A few cases have reported significantly diminished late gadolinium enhancement at 6 weeks of follow-up, which correlates with significant improvement in the AV block after appropriate antibiotic treatment.²

The Infectious Diseases Society of America (IDSA) recommends treatment for Lyme carditis based upon the degree of AV block and PR interval length. Patients with 1st degree AV block can be treated with a 14-21 day course of one of the following oral regimens: doxycycline 100mg twice daily, amoxicillin 500 mg three times a day, or in case of a Doxycycline or amoxicillin allergy, cefuroxime 500 mg twice daily.^{1,7} Patients with higher degrees of AV block or those with PR interval > 0.3 seconds should be treated with one of the following intravenous regimens for 14-28 days and have cardiac monitoring to evaluate the need for transcutaneous pacing.^{1,7} Different regimens for IV antibiotics include ceftriaxone 2g daily, cefotaxime 2g every eight hours, or penicillin G 3.3 million units every 4 hours.¹ Although a pacemaker is generally not necessary, up to one-third of Lyme carditis cases may require temporary cardiac pacing.⁶ Once the patient's condition has stabilized, they may complete the duration of therapy with oral antibiotics.⁷ Over 90% of cases of Lyme carditis complete recover.⁶

Conclusion

Lyme disease is a multisystem disease caused by *Borrelia burgdorferi*. Cardiac involvement with Lyme disease is extremely rare and occurs during the early disseminated phase of the disease, typically within 3-10 days of initial exposure. The most common expression of Lyme carditis is fluctuating degrees of transient AV block, the most common being 1st degree AV block, with up to 50% of these patients progressing to complete heart block. Diagnostic testing with Lyme antibodies and Western Blot is critical for patients presenting with complete heart block. Early appropriate antibiotic therapy depends on the degree of AV block and length of PR interval. For those with 1st degree AV block and PR interval < 0.3 sec, oral antibiotic regimens are appropriate. Patients with higher levels of AV block or PR interval > 0.3 sec require intravenous antibiotics and cardiac monitoring to assess the need for transcutaneous pacing. AV conduction disturbances in Lyme carditis have been illustrated to be transient and even patients with high levels of heart block generally have a good prognosis.

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