



## Case Report

# Lev's Syndrome: A rare case of progressive cardiac conduction disorder presenting to the emergency department☆☆☆



Brandon M. Carius, MPAS, PA-C\*, , Dr. Brit Long, MD, , Dr. Steve Schauer, DO

Emergency Medicine Department, Brooke Army Medical Center, Fort Sam Houston, TX, USA

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## ABSTRACT

Lev's Syndrome is a rare, progressive cardiac conduction defect (PCCD) due to myocardial fibrosis first described by Maurice Lev in 1964. This condition, proposed to start in the fourth decade of life, involves a sclerotic fibrofatty degeneration of the Bundle of His and Purkinje fibers, which Lev proposed caused increasing AV delay with age. With the prevalence of electrocardiogram (ECG) use in the emergency department (ED) for cardiac and non-cardiac complaints, dysrhythmias can be incidentally found and confuse diagnosis and disposition. We highlight the case of an 84-year-old male who presented to the ED for acute onset of diffuse facial paresthesias with elevated blood pressure at home and was found to be significantly bradycardic on initial evaluation. On serial ECGs, the conduction rhythm changed from an initial new first-degree atrioventricular (AV) block with left bundle branch block (LBBB), to a later first-degree AV block without LBBB. Cardiology was consulted. Serial ECGs demonstrated an evolving conduction block arrhythmia consistent with Lev's Syndrome. Here we describe a case of symptomatic bradycardia found to be consistent with Lev's Syndrome.

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## 1. Case

An 84-year-old male presented to the emergency department (ED) complaining of acute, diffuse facial numbness and tingling approximately 3 h prior to presentation. He stated that he had been sitting at home at the time of symptom onset, and a home blood pressure device measurement returned "unreadable". His symptoms resolved within 5–10 min of applying a 0.2 mg/h transdermal nitroglycerin patch. He said he took the nitroglycerin sporadically for various non-specific symptoms. The patient also noted mild, diffuse weakness leading up to the event that persisted during evaluation in the ED. He denied any other symptoms, including neurologic symptoms. Medical history was significant for coronary artery disease with a cardiac catheterization and stent placement 5 years prior to presentation, renal artery stenosis, hypertension, iron deficiency anemia, and acid reflux. He stated compliance on his medications of aspirin, telmisartan, carvedilol, amlodipine, atorvastatin, pantoprazole, and ranitidine. In the ED, systolic and diastolic blood pressures reached peaks of 165 mm Hg and 72 mm Hg,

respectively. He consistently demonstrated fluctuating bradycardia that dropped as low as 32 beats per minute (bpm). Laboratory testing, including thyroid stimulating hormone, electrolyte levels, and cardiac markers, was unremarkable. An electrocardiogram (ECG) from 5 weeks prior was referenced, which showed a normal sinus bradycardia (Fig. 1). Initial ECG on triage showed a sinus bradycardia with a new first-degree atrioventricular (AV) conduction block and left bundle branch block (LBBB), without evidence of acute ischemia (Fig. 2). After initial evaluation, a repeat ECG approximately 90 min later showed only the first-degree AV block, narrow QRS, and normal axis, but no LBBB (Fig. 3). With his persistent fluctuating bradycardia, significant cardiovascular history, and significantly altered ECG patterns, the cardiology service was consulted.

Following cardiology evaluation, a third ECG was obtained (Fig. 4). Taken approximately 2 h after the second ECG (3.5 h after the first ECG), this ECG showed a significantly different pattern. As demonstrated with the rhythm strip along the bottom, the first beat appeared as a third-degree AV block, first with a widened QRS indicating a ventricular escape beat, then 3 junctional escape beats. After this fourth ventricular beat, the final 2 beats on the rhythm strip transformed to a 2:1 second-degree AV block with ventricular capture. The ventricular rate shown (34 bpm) appeared consistent with the patient's initial heart rate on triage.

Given the patient's alternating levels of bradycardia, ECG changes, and continued risk for syncope and SCD, he was admitted to the cardiac

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\* Corresponding author at: 3551 Roger Brooke Drive, Fort Sam Houston, TX 78234, USA.

E-mail address: [brandon.m.carius.mil@mail.mil](mailto:brandon.m.carius.mil@mail.mil) (B.M. Carius).

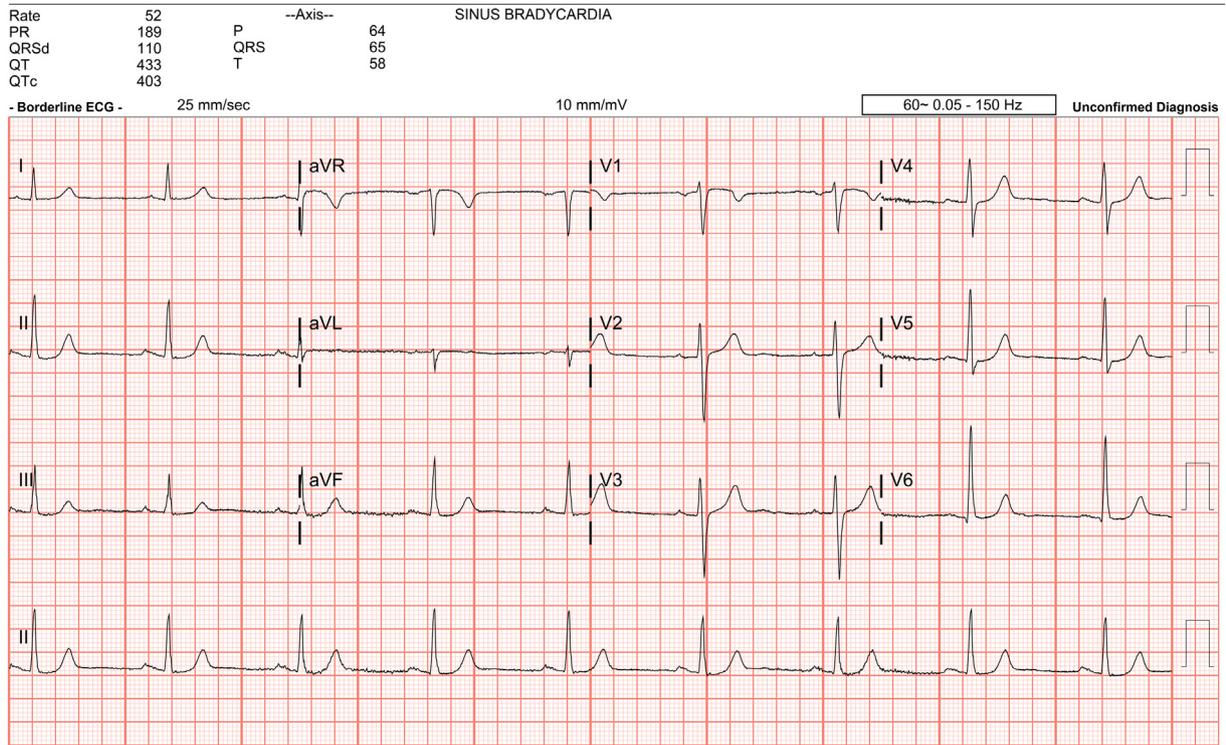


Fig. 1. The patient's reference electrocardiogram (ECG) from 5 weeks prior to presentation demonstrated a sinus bradycardia.

care unit. The next day, a dual-chamber pacemaker was placed without complication. An ECG taken after pacemaker implantation showed a regular rhythm and widened QRS complex consistent with ventricular pacing (Fig. 5). Subsequent follow-up showed symptomatic resolution and no adverse events.

2. Discussion

Lev's Syndrome is a rare progressive cardiac conduction defect (PCCD) due to myocardial fibrosis first described by Maurice Lev in 1964 [1]. Lev described PCCD in 1964 and developed a theory of

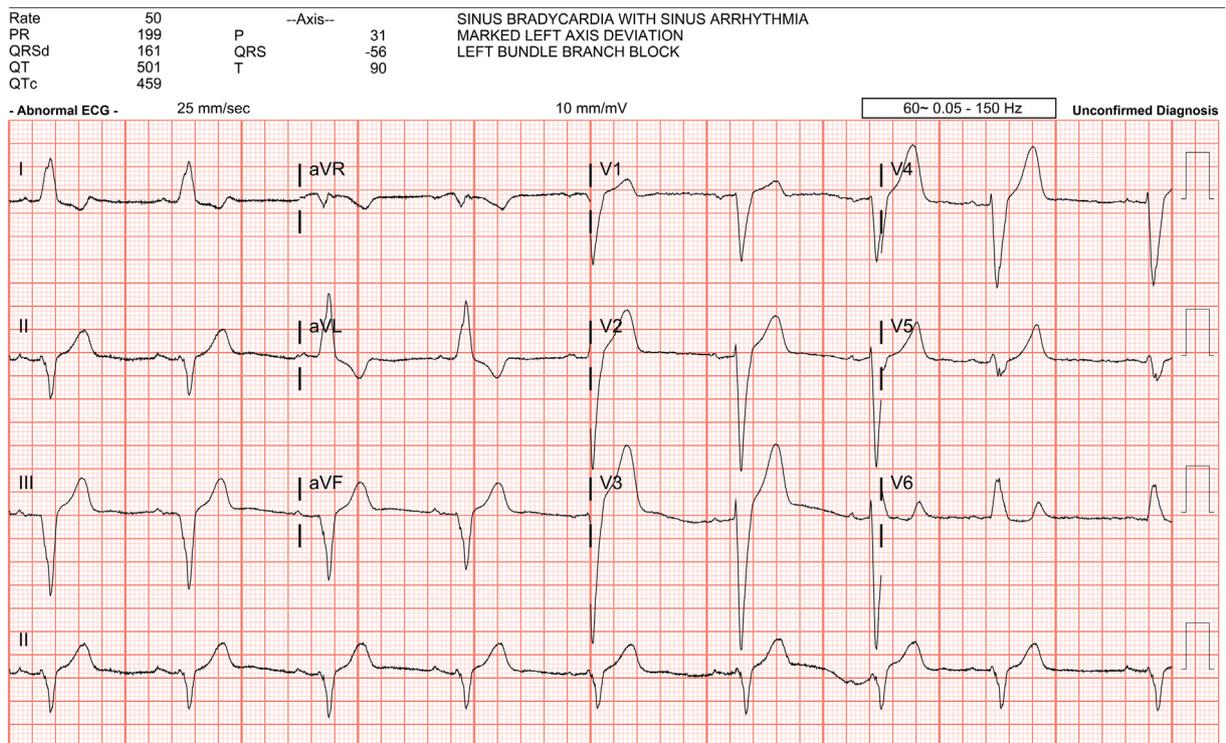


Fig. 2. ECG upon presentation showed a new first-degree atrioventricular (AV) block with left bundle branch block (LBBB).

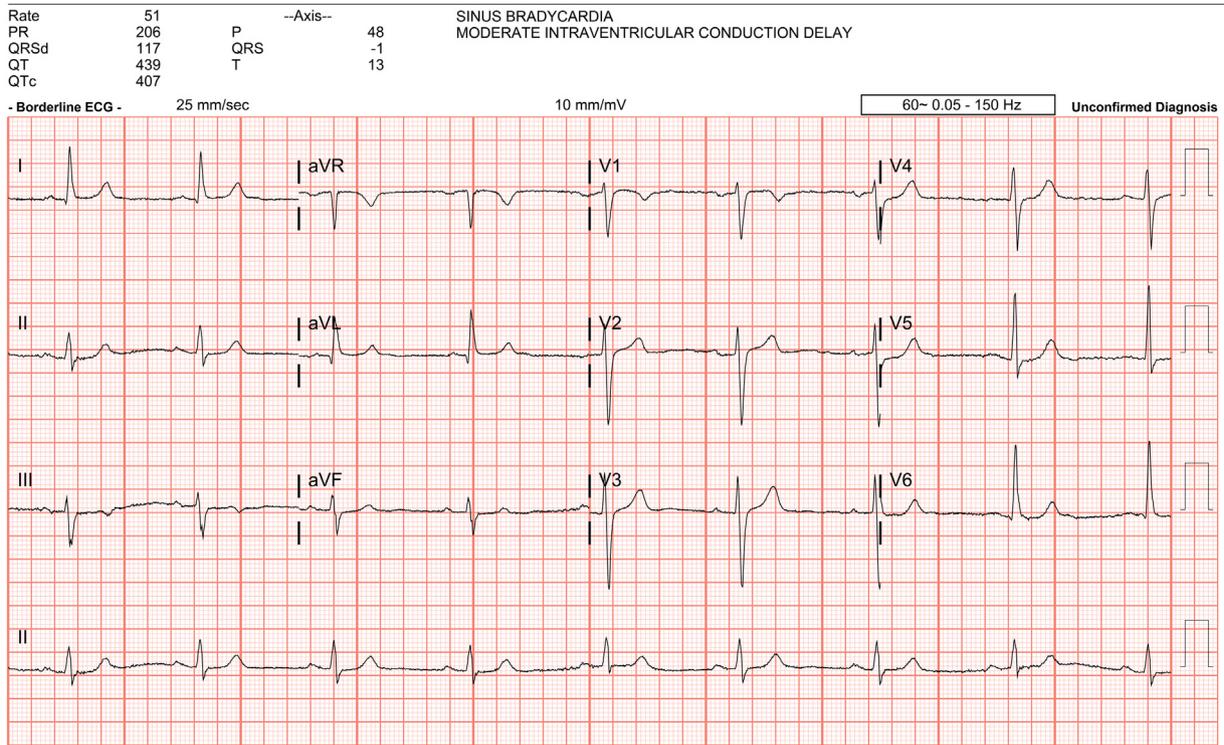


Fig. 3. Repeat ECG approximately 90 min after initial presentation found only a first-degree AV block with narrow QRS and no LBBB.

progressive conduction delay with aging, approximately around the fourth decade, involving multiple cardiac structures: the aortic sinuses, ventricular septum, central fibrous body, and mitral annulus [2,3]. Most significantly, the bundle of His and Purkinje fibers displayed a sclerotic “fibro-fatty degeneration” with age that Lev proposed caused increasing

AV delay [3-5]. Jean Lenegre developed similar theories on PCCD at the same time as Lev. Comparison of the two syndrome theories show Lev’s Syndrome with diffuse fibrosis of the fibrous cardiac skeleton and aforementioned structures, as well as the conduction system, primarily involving the Bundle of His and the proximal bundle branch fibers [3,5-

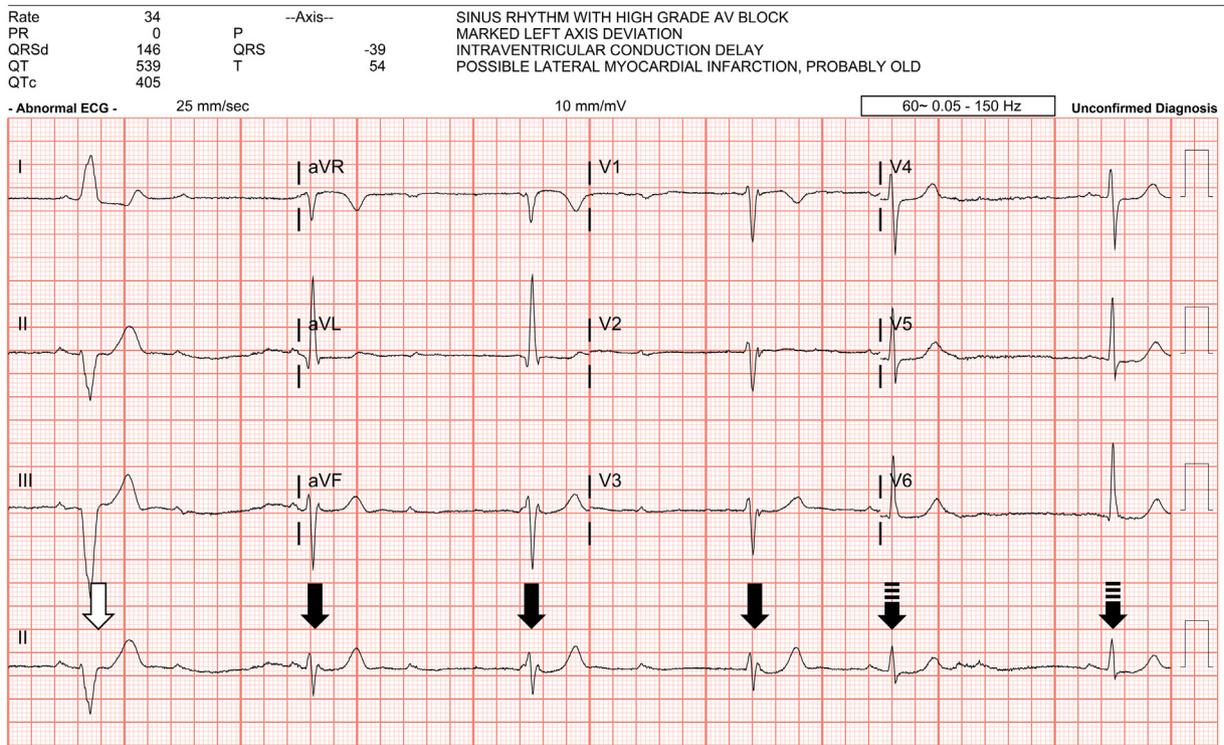


Fig. 4. Third ECG approximately 3.5 h into emergency department presentation demonstrated significant variation in conduction on the rhythm strip (lead II). This rhythm first showed a third-degree AV block, first with a widened QRS indicating a ventricular escape beat (white arrow), then three junctional escape beats (black solid arrows). After this fourth ventricular beat, the final 2 beats on the rhythm strip are a 2:1 second-degree AV block with ventricular capture (black dotted arrow).

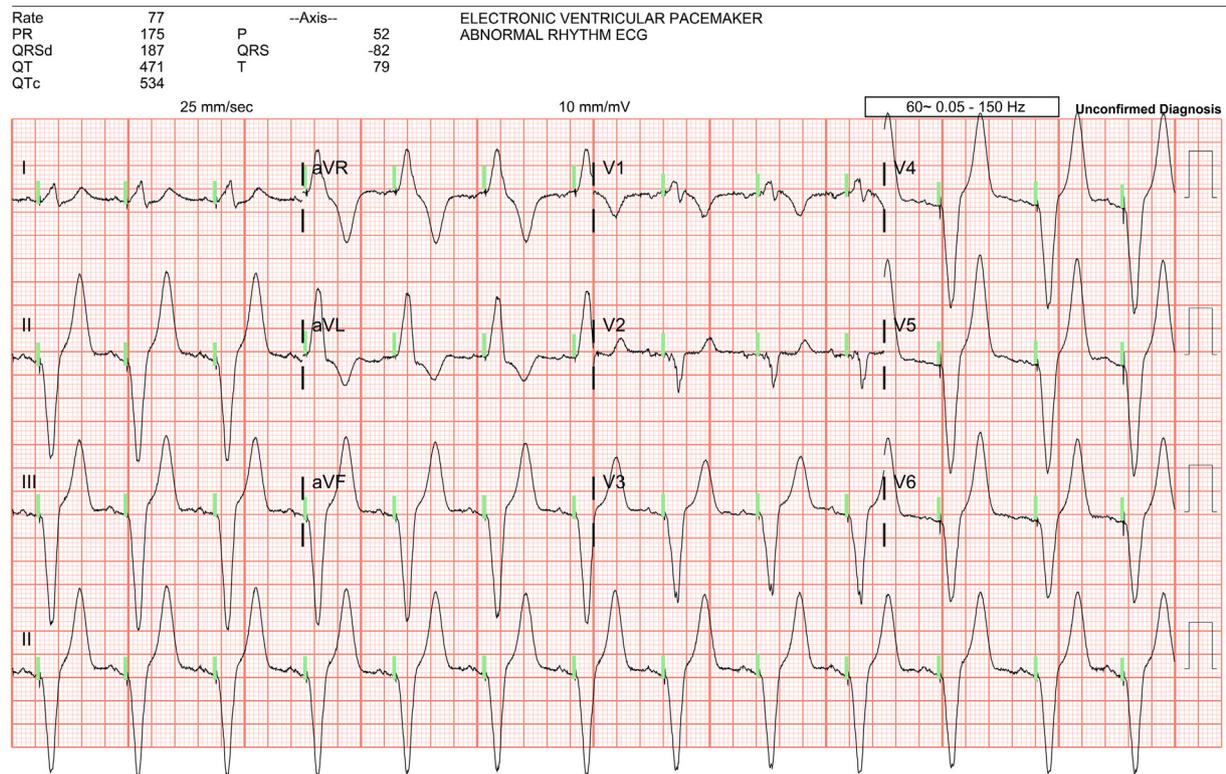


Fig. 5. ECG following dual-chamber cardiac pacemaker placement showed a regular rhythm and widened QRS complex consistent with ventricular pacing.

7]. In contrast, Lenegre's Syndrome centered exclusively on the conduction system and the fibrotic degeneration of the distal Purkinje fibers [5,6]. Despite their differences, limited literature has grouped the two models together as 'Lev-Lenegre Syndrome' [7,8]. Although Lev proposed that PCCD may result as part of the normal aging process, limited literature supports genetic predisposition [1,2,5,6]. ECG findings may be incidental on screening, or may be part of an evaluation for unexplained bradycardia, palpitations, or syncope [1,7]. Lev's Syndrome is listed as a possible etiology of sudden cardiac death (SCD) [1,9].

We describe a case of Lev's Syndrome presenting as intermittent bradycardia in the ED. Initial ECG findings of first-degree AV conduction block with LBBB evolved over subsequent ECGs to include second- and third-degree AV blocks. All of these conduction findings differed from his most recent pre-visit ECG showing a sinus bradycardia just five weeks earlier. These ECG findings are consistent with a rarely-found PCCD caused by diffuse fibrosis of the cardiac skeleton and conduction structures.

A search of the literature revealed no specific treatment options, recommendations, or consensus regarding Lev's Syndrome. Rather, recently described treatment strategies show Lev's Syndrome grouped with similar conduction abnormalities that have concern for increased risk of cardiovascular-induced syncope. This treatment chiefly consists of pacemaker implantation and strategies to minimize high-risk activities, such as driving [9].

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images for submission. The view(s) expressed herein are those of the author(s) and do not reflect the official policy or position of Brooke Army Medical Center, the U.S. Army Medical Department, the U.S. Army Office of the Surgeon General, the Department of the Army, the Department of the Air Force and Department of Defense or the U.S. Government.

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