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CASE REPORT

BEGINNER

CLINICAL CASE

The Longer the Block, the Harder You Fall

Extrinsic Idiopathic Atrioventricular Block Masquerading as Seizures

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ABSTRACT

History and physical examination are the diagnostic cornerstones of transient loss of consciousness (TLOC). However, details can be scarce and examination unrevealing, thus making the diagnosis elusive. In a case of convulsive TLOC, the initial diagnosis was incorrect, but a fortuitously captured event on telemetry yielded the diagnosis: extrinsic idiopathic atrioventricular block. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2021;3:1086–90) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 62-year-old woman with diabetes and hypertension presented to the emergency department with multiple convulsive episodes corresponding to transient loss of consciousness (TLOC). Witnesses described an event lasting approximately 30 s during which the patient was sitting at a table, developed arm shaking, and then fell to the floor. She then had 2 more episodes of TLOC with jerking of her limbs, the second of which occurred during ambulation. She denied any prodrome, including palpitations, chest pain, lightheadedness, nausea, or flushing, and any preceding aura, incontinence, tongue biting, or up-rolling of her eyes during events, although she did have confusion on awakening. Physical examination showed benign findings, including normal heart sounds and the absence of carotid bruit or recurrent symptoms with carotid massage or arm exercises.

LEARNING OBJECTIVES

- To understand pathophysiology of reflex anoxic seizures, or *convulsive syncope*.
- To learn the types of paroxysmal AVB and their management.
- To diagnose EI-AVB by using an instructive telemetry tracing.

PAST MEDICAL HISTORY

She denied illicit drug use but admitted to heavy alcohol intake the night before symptoms. She had experienced a seizure 15 years earlier but without a diagnosis or treatment. She had no family history of seizures, cardiac disease, or sudden death.

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DIFFERENTIAL DIAGNOSIS

The differential diagnoses included seizure disorder, psychogenic nonepileptic seizures (PNES), and arrhythmogenic syncope.

INVESTIGATIONS

In the emergency department, the results of laboratory tests, an electrocardiogram (ECG), and an electroencephalogram were normal. She was admitted for further work-up; her echocardiogram, Lyme serological examination, and brain magnetic resonance imaging were all unremarkable. Neurology consultants proposed PNES as a likely diagnosis of exclusion because there was no substrate for seizure noted on imaging, the post-episode confusion was brief, and convulsions were bilateral, thereby making a focal seizure unlikely. Later that evening, during telemetry, the patient experienced a witnessed recurrent event. An abrupt atrioventricular (AV) block (AVB) developed, followed by TLOC, and then convulsions. Telemetry reflected the physiology, including motion artifact corresponding to convulsions that occurred after the onset of AVB (**Figure 1**). AV conduction resumed, and the patient awoke in a confused state.

MANAGEMENT

Transcutaneous pacing pads were placed, and an emergent transvenous pacing wire was inserted. On the basis of telemetry findings, through exclusion of other causes, a diagnosis of extrinsic idiopathic AVB (EI-AVB) was made.

The patient underwent dual-chamber pacemaker implantation the next day, programmed in the Managed Ventricular Pacing (MVP) mode (Medtronic, Minneapolis, Minnesota), which avoids unnecessary ventricular pacing during intact AV conduction.

DISCUSSION

Epileptic seizures, syncope, and PNES account for >90% of patients presenting with TLOC (1), with history and physical examination being the crux of the work-up (2). Despite markedly different pathophysiological characteristics, the clinical presentations of syncope and seizure can be difficult to differentiate from each other, particularly when patient-provided history is limited by loss of consciousness and episodes may be unwitnessed. In fact, misdiagnosis in TLOC may be as prevalent as 30% and is associated with increased morbidity (3).

In this case, history and a normal physical examination did not clearly differentiate syncope from seizure. In the work-up for TLOC, normal findings on physical examination, ECG, and echocardiogram preclude further inpatient cardiac testing. Guidelines recommend inpatient telemetry only in the presence of any high-risk feature (2,4). As they pertain to this case, high-risk features include syncope without prodrome or while seated, but only in the presence of structural heart disease or an abnormal ECG (4). However, the negative work-up to this point and recurrent events in such short succession, although not addressed in guidelines, prompted admission to a telemetry unit. Fortuitously, the patient then had an episode captured on telemetry, thus demonstrating that the initial diagnosis of PNES was incorrect. The convulsions were *real*, reflex-anoxic seizures, or *convulsive syncope*, caused by cerebral hypoperfusion from prolonged AVB (5). If the patient had not had a recurrent episode during telemetry, an ambulatory cardiac monitoring device, such as a rhythm monitor patch or implantable loop recorder, would have been appropriate. Device choice depends on the frequency of symptoms (4).

Paroxysmal AVB can be divided into 3 subtypes, differentiation among which is critical because treatments differ: extrinsic vagal AVB (EV-AVB, also known as *vasovagal syncope* or *neurocardiogenic syncope*), which is typically treated with lifestyle modification, isotonic maneuvers, and rarely medication or pacemaker insertion (6); intrinsic AVB (I-AVB, also known as *Stokes-Adams syndrome*), treated with pacemaker implantation (7); and, EI-AVB (8), also treated with pacemaker implantation. The patient's telemetry tracings excluded the more common EV-AVB and I-AVB. Although there was subtle PR interval prolongation on the last conducted beat, there was no PP interval prolongation before the block, as would be found in EV-AVB. In fact, the PP interval *shortened* (i.e., sinus rate *increased*) during block, a sympathetically mediated compensatory response and the opposite of what would be found in EV-AVB. Contrary to what is often observed in I-AVB, there was no evidence of the following: native conduction

ABBREVIATIONS AND ACRONYMS

AAI = single-chamber atrial pacing mode

APL = plasma adenosine level

AV = atrioventricular

AVB = atrioventricular block

DDD = dual-chamber pacing mode

ECG = electrocardiogram

EI-AVB = extrinsic idiopathic atrioventricular block

ENT1 = equilibrative nucleoside transporter 1

EV-AVB = extrinsic vagal atrioventricular block

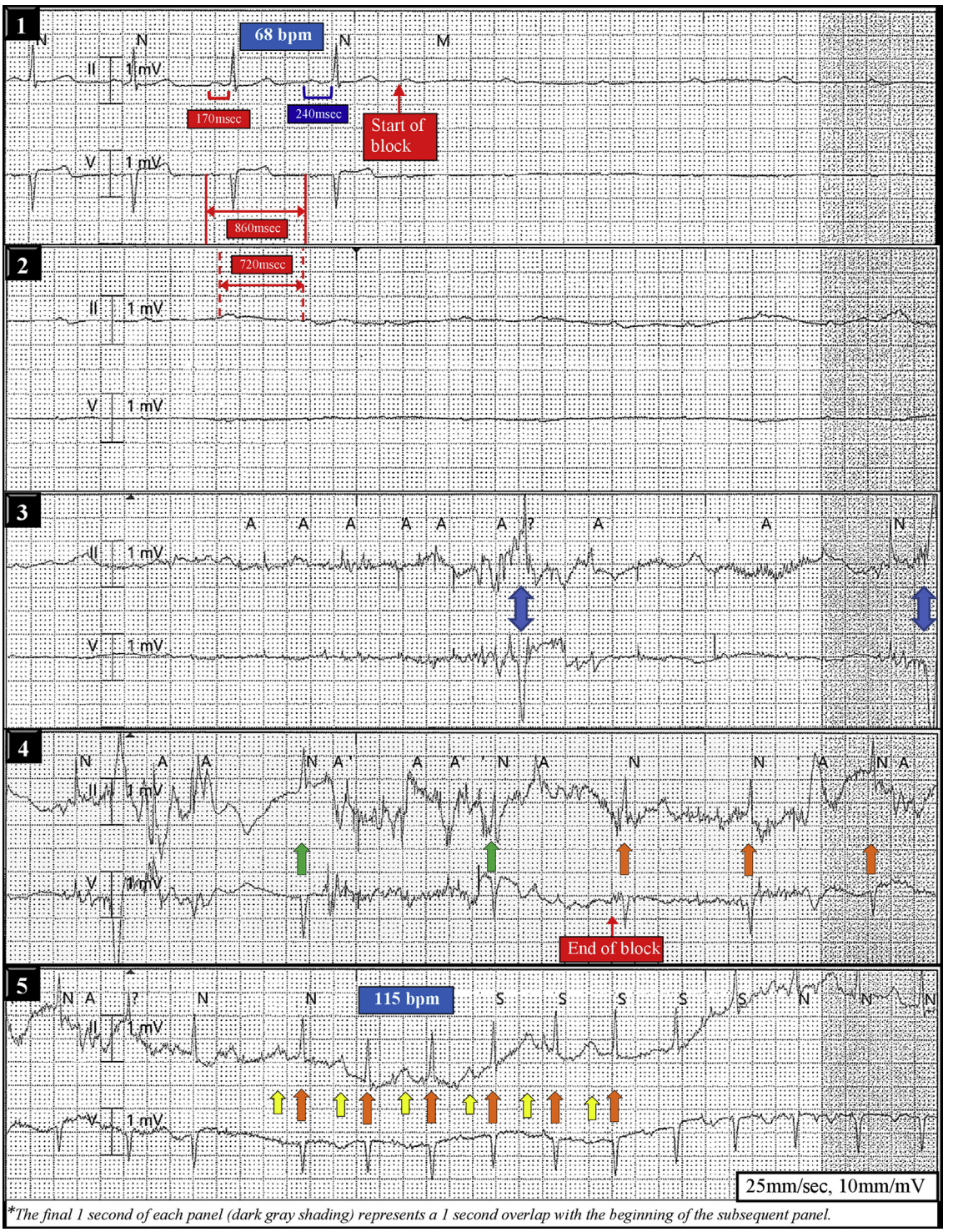
I-AVB = intrinsic atrioventricular block

MVP = Managed Ventricular Pacing

PNES = psychogenic nonepileptic seizures

TLOC = transient loss of consciousness

FIGURE 1 **Telemetry Tracing of Witnessed Event of TLOC With Convulsions**



(1) Before atrioventricular block (**red arrow,** "start of block"), there is slight PR interval prolongation (**red bracket:** PR interval, 170 ms; **blue bracket:** PR interval, 240 ms). There is no PP interval prolongation before block **(solid red line),** and there is PP interval shortening during block **(dotted red line).** The duration of atrioventricular block measures 22.8 s (**vertical arrows,** "start of block "to "end of block"). **(2)** Period of complete heart block without any ventricular rhythm. **(3)** Complete heart block with 2 ventricular escape beats, which are slightly wider than the patient's conducted QRS complexes **(blue arrows). (4)** Junctional escape beats, which match the patient's conducted QRS complexes **(green arrows),** followed by sinus rhythm with 2:1 conduction (P waves, not labeled because of motion artifact; conducted QRS complexes, **orange arrows**). **(5)** Sinus rhythm with 1:1 conduction (P waves, **yellow arrows**; conducted QRS complexes, **orange arrows**). Note that sinus rate has increased from 68 to 115 beats/min by the end **(blue box),** denoting a physiological sympathetic response to pathological atrioventricular block.

disease (e.g., bundle branch block); the *condiciones sine quibus non* of an initiating premature atrial or ventricular complex, causing conduction tissue to become unexcitable secondary to slow partial depolarization and sodium channel inactivation (i.e., phase 4 block); or a subsequent ventricular complex resetting the membrane potential, thus prompting resolution of AVB (7). Finally, the pattern observed was not consistent with phase 3 block, where a premature beat or faster atrial rate leads to AVB. Thus, the diagnosis of EI-AVB was made.

EI-AVB is an underrecognized cause of TLOC that occurs in patients with low baseline circulating plasma adenosine levels (APLs). The low APL renders patients hypersensitive to surges in endogenous adenosine, which induces AVB. Given that results of standard testing in patients with EI-AVB are normal, and because APL testing is not routinely available, the diagnosis of EI-AVB remains a diagnosis of exclusion and hinges on documentation of the specific pattern of AVB, usually with ambulatory rhythm monitoring (9).

Extracellular and intracellular adenosine levels in the heart are mediated by the equilibrative nucleoside transporter (ENT)-1, and ethanol has been shown to inhibit ENT1-dependent 2-chloroadenosine uptake in cardiac cells, an action that may promote adenosinergic pathways (10). This effect diminishes in patients with long-term alcohol use as a result of heterologous desensitization. In this patient, who was not a regular consumer of alcohol, the previous night's uncharacteristic consumption of alcohol may have mediated an increase in extracellular adenosine, thereby inducing recurrent paroxysmal AVB.

FOLLOW-UP

At 3 months of follow-up, the patient had no syncope. However, she had 7 ventricular pacing events, with intrinsically conducted ventricular rates of 77 to 98 beats/min and AV delays of 130 to 160 ms immediately before ventricular pacing, consistent with episodes of EI-AVB treated with ventricular pacing. The MVP feature in this patient elucidates the necessity of pacemaker therapy in a patient with a very low ventricular pacing burden. The MVP mode prevents unnecessary ventricular pacing in the setting of a physiological AV delay (PR interval prolongation). With this mode, the device switches from single-chamber atrial pacing mode (AAI) to the dual-chamber pacing mode (DDD) only when ventricular pacing become necessary. The device counters revealed 7 switches in the MVP mode from AAI to DDD yet accounted for only $<0.1\%$ of ventricular pacing. This low ventricular pacing burden belies the necessity of ventricular pacing in this patient and speaks to the imprecise meaning of the term "pacemaker dependence," often used to describe patients with persistent, obligate ventricular pacing.

CONCLUSIONS

A higher level of suspicion of uncommon causes, long-term ambulatory monitoring, and improved predictive tools to help guide work-up for TLOC may help reduce misdiagnosis and underdiagnosis. EI-AVB is an underappreciated cause of recurrent syncope, which, when manifesting with convulsions, as in other cerebral hypoperfusing arrhythmias, may obscure an arrhythmic origin of TLOC by masquerading as a primary neurological disorder. In patients with paroxysmal AVB, although ventricular pacing is uncommon, it may nonetheless be lifesaving.

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