

Syncope and Idiopathic (Paroxysmal) AV Block



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KEYWORDS

• Atrioventricular block • Implantable loop recorder • ECG • Syncope

KEY POINTS

- Syncope due to idiopathic AV block is characterized by: 1) ECG documentation (usually by means of prolonged ECG monitoring) of paroxysmal complete AV block with one or multiple consecutive pauses, without P-P cycle lengthening or PR interval prolongation, not triggered by atrial or ventricular premature beats nor by rate variations; 2) long history of recurrent syncope without prodromes; 3) absence of cardiac and ECG abnormalities; 4) absence of progression to persistent forms of AV block; 5) efficacy of cardiac pacing therapy.
- The patients affected by idiopathic AV block have low baseline adenosine plasma level values and show an increased susceptibility to exogenous adenosine. The APL value of the patients with idiopathic AV block is much lower than patients affected by vasovagal syncope who have high adenosine values.

DIAGNOSIS OF IDIOPATHIC AV BLOCK

Syncope due to idiopathic atrioventricular (AV) block (**Box 1**) is a distinct clinical form of syncope characterized by common clinical and electrophysiologic features¹:

- Electrocardiographic (ECG) documentation (usually by means of prolonged ECG monitoring) of idiopathic paroxysmal complete AV block with one or multiple consecutive pauses; AV block occurs without P-P cycle lengthening or PR interval prolongation and is not triggered by atrial or ventricular premature beats nor by rate variations (**Fig. 1**)
- Long history of recurrent syncope without prodromes

- Absence of cardiac and ECG abnormalities
- Absence of progression to persistent forms of AV block
- Efficacy of cardiac pacing therapy

Patients affected by idiopathic AV block have low baseline adenosine plasma level (APL) values and show an increased susceptibility to exogenous adenosine. In one study,¹ the median baseline APL of these patients was significantly lower than that found in the age-matched and sex-matched population of 81 healthy subjects: 0.33 μM (interquartile range 0.20–0.56) versus 0.49 μM (0.38–0.68) ($P = .017$). APL values of patients with idiopathic AV block are much lower than those of patients affected by typical vasovagal syncope and patients with positive tilt table

This article originally appeared in Cardiac Electrophysiology Clinics, Volume 5, Issue 4, December 2013. The authors have nothing to disclose.

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Cardiol Clin 33 (2015) 441–447

<http://dx.doi.org/10.1016/j.ccl.2015.04.012>

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Box 1 Definitions

Idiopathic (paroxysmal) AV block is defined as a paroxysmal third-degree AV block that displays an abrupt onset and no other rhythm disturbances before or during the block and that occurs in patients with a normal heart and normal ECG

Syncope due to idiopathic paroxysmal AV block is defined as syncope caused by long asystolic pauses due to complete paroxysmal AV block

testing who have higher APL values than normal subjects (Fig. 2).

Other relevant laboratory findings are as follows:

- Most patients with idiopathic AV block show a high susceptibility to the rapid IV injection of 18 mg adenosine or 20 mg adenosine triphosphate (ATP) test (Fig. 3). The adenosine/ATP test fairly reproduces spontaneous AV block. The adenosine response is abolished by theophylline, an adenosine antagonist, but not by atropine, a vagal antagonist.²
- Tilt table test may be positive, but positivity rate is lower than in patients with vasovagal syncope and is never able to reproduce an AV block. Thus tilt table test response seems to be nonspecific.
- Carotid sinus massage is almost invariably negative.

DIFFERENTIAL DIAGNOSIS FROM OTHER TYPES OF AV BLOCK

Idiopathic paroxysmal AV block has different clinical and electrophysiologic features from those of the 2 other known types of paroxysmal AV block: intrinsic AV block due to AV conduction disease and extrinsic vagal AV block. Well-defined clinical and electrophysiologic features differentiate them.

Intrinsic paroxysmal AV block, which usually occurs in patients with underlying heart disease and/or abnormal standard ECG, is regarded as a manifestation of an intrinsic disease of the AV conduction system (Stokes-Adams attack), which is confirmed by abnormal electrophysiologic findings.^{3,4} The AV block is usually initiated by atrial, His, or ventricular premature extrasystole, increased heart rate (tachy-dependent AV block) or decreased heart rate (brady-dependent AV block), all features that support a diagnosis of intrinsic AV block (Fig. 4). The outcome is characterized by a rapid progression toward permanent AV block.^{3,4}

Extrinsic (vagal) AV block is localized within the AV node and is associated with slowing of the sinus rate. A classic vagal effect on the conduction system includes gradual slowing of the sinus rate (P-P interval) and AV conduction (prolonging PR), which are occasionally followed by sinus arrest or complete AV block. The 2 conditions frequently coexist, indicating a simultaneous vagal action on sinus node and AV node. Even when a more

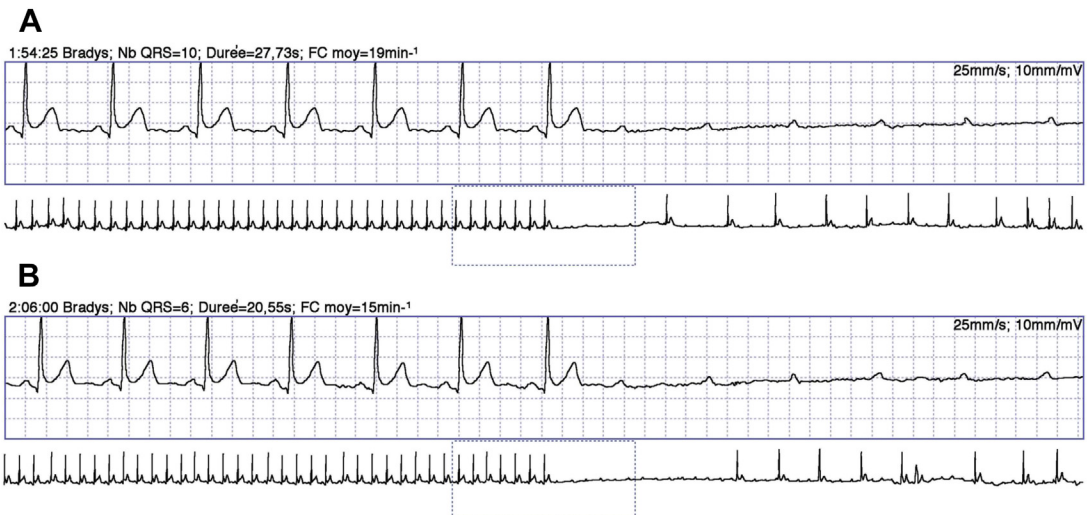


Fig. 1. Holter recording of 2 episodes of spontaneous syncope (A, B) that occurred a few minutes apart. The 2 episodes were very similar and were characterized by sudden-onset complete AV block without changes in P-P cycle length, which constantly remained 720 ms (top, trace), and long ventricular asystole of 7 and 11 seconds, respectively (bottom, compressed trace). (From Brignole M, Deharo JC, De Roy L, et al. Syncope due to idiopathic paroxysmal atrioventricular block. Long-term follow-up of a distinct form of atrioventricular block. *J Am Coll Cardiol* 2011;58:170; with permission.)

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