Paroxysmal atrioventricular block

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Paroxysmal atrioventricular block (AVB) is a poorly defined clinical entity characterized by abrupt and unexpected change from 1:1 atrioventricular conduction to complete heart block, leading to syncope and potential sudden cardiac death. Although a dangerous condition because of unreliable escape mechanism, proper diagnosis of paroxysmal AVB is often missed and overlooked because of its unfamiliarity, unpredictability, and in some cases, no clear evidence of atrioventricular conduction disease during normal 1:1 conduction.

KEYWORDS Phase 4 block; Complete heart block; Syncope; Sudden cardiac death; Pacemaker

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Introduction

Atrioventricular block (AVB) as a cause of syncope is important because it is potentially preventable or even treatable. Although tachyarrhythmias are primarily emphasized as etiologies for sudden cardiac death (SCD) and syncope, sinus node dysfunction and cardiac conduction abnormality still account for more than half of arrhythmic syncope,[1] whereas bradyarrhythmias are associated with 16% to 25% of SCD.[2,3] Torsades de pointes (TdP) in the setting of atrioventricular (AV) block is another well-described etiology of SCD in patients with AVB.[4,5] Clinically, significant AV block may present as acquired high-degree or complete heart block, vagally mediated AV block, congenital heart block, and paroxysmal atrioventricular block. In 1933, Sachs and Traynor[6] first described an intriguing phenomenon of paroxysmal AVB, followed by Coumel et al[7] in 1971, who described 2 cases of paroxysmal AVB precipitated by premature atrial beats. The hallmark of this condition is a sudden change from apparently normal 1:1 AV conduction to complete heart block, initiated by a pause, leading to ventricular asystole. Although no official definition is available, we define paroxysmal AVB as a sudden, pause-dependent phase 4 AV block occurring in diseased conduction system. Fundamental understanding and prompt recognition of paroxysmal AVB is essential because asystole with potential SCD may be preventable with permanent pacemaker implantation.

Epidemiology

There is no established prevalence for paroxysmal AVB, but it is likely underreported because of poor recognition, sparse information in medical literature, its unpredictability, and possible lack of an obvious marker for AV conduction disease between culprit episode(s) (Figure 1). In fact, paroxysmal AVB is neither clearly defined nor addressed in the American College of Cardiology/American Heart Association guidelines.[8] Conversely, without clearly defined criteria, any sudden development of AVB has loosely been described in previous literature as paroxysmal AVB, falsely raising its prevalence. In a prospective study of 52 incomplete and complete right bundle branch block (RBBB) patients with syncope and a negative electrophysiology study (EPS), for example, prophylactic implantable loop recorders showed development of sudden complete heart block in 13 of 52 (25%).[9] However, of the 13 patients, 5 (38%) events were triggered by atrial or ventricular premature beats (APB or VPB) and were attributable to paroxysmal AVB, whereas not enough information is available to make such conclusion in the remaining 8 patients.

Although no established predictors for paroxysmal AVB exist, evidence of distal conduction disease at baseline is often present, with RBBB being the most common finding. Combining our unpublished series of 30 patients from Beth Israel Deaconess Medical Center and the Department of Cardiology at the University Hospital of Maastricht, the Netherlands, and 38 cases of paroxysmal AVB reported from previously publishes series,[10,11] 31 of 68 (45%) had underlying RBBB, 10 of 68 (15%) had left bundle branch block (LBBB), 8 of 68 (12%) had intraventricular conduction delay, and 19 of 68 (28%) had normal QRS (Table 1).

Simply by virtue of more prevalent distal conduction disease, paroxysmal AVB is reported more frequently in older adults, although possible paroxysmal AVB in pediatric population has been described.[12] Paroxysmal AVB occurs equally in women and men, and age at presentation ranged from 26 to 99 years (72% age 60 years or older). On our unpublished series of 30 patients with paroxysmal AVB (Table 1), initiation of AVB was by either an atrial prema-

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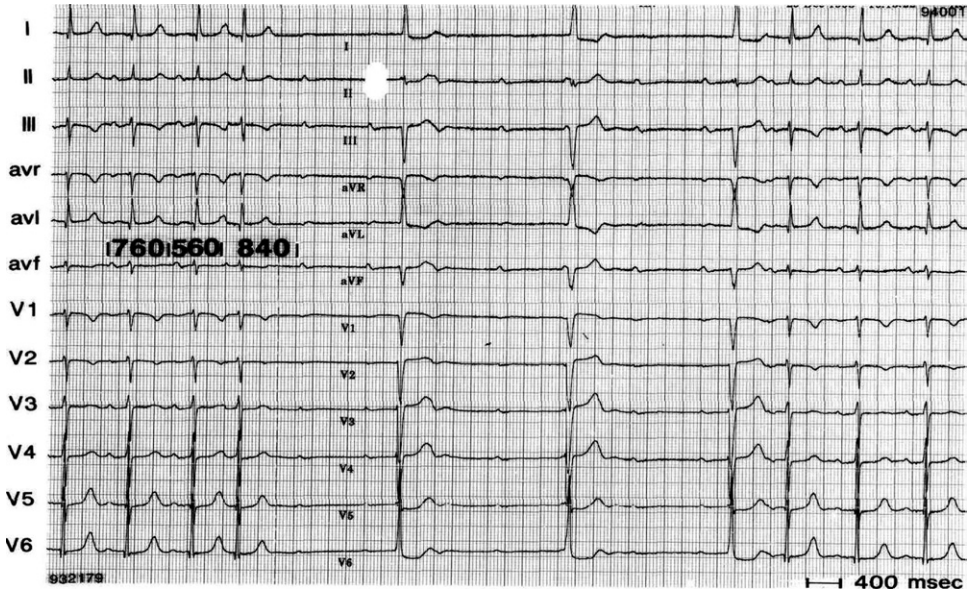

Figure 1 Paroxysmal AVB in a 49-year-old man with dizziness and a completely normal baseline ECG. Heart block is initiated by a conducted atrial premature beat with likely His bundle escape rhythm. Note prolonged P-P interval after an atrial premature beat compared with preceding sinus P-P interval. 1:1 AV conduction resumes near the end of the recording. AVB = atrioventricular block; ECG = electrocardiogram.

ture beat (9 of 30; 30%), ventricular premature beat (7 of 30; 23%), or His extrasystole (3 of 30; 10%). Initiation in the remaining 11 of 30 (37%) patients were variable including supraventricular tachycardia, carotid sinus massage, Valsalva maneuver, and spontaneous sinus rate slowing.

Mechanism

Phase 4 depolarization is a normal property of the sinoatrial (SA) node and a specialized conduction system that is responsible for automaticity. Phase 4 bradycardia-dependent block or aberrancy results when a supraventricular or ventricular impulse reaches a diseased His-Purkinje system (HPS) during phase 4 of the action potential at a time when sodium channels are inactive. Consequently, subsequent impulses can no longer depolarize the diseased tissue, leading to asystole. Paroxysmal AVB is a unique disorder of HPS, and is secondary to local phase 4 block in the His bundle[13] or in the bundle branches after a critical change in the H-H interval. During a long pause (prolonged diastolic period), the fibers of the often-diseased HPS spontaneously

Table 1 Characteristics of the 30 patients with paroxysmal AVB at Beth Israel Deaconess Medical Center, University of Maastricht, and combined data from previous publications[10,11]

Baseline characteristics	BIDMC and UoM (n = 30)	Total (n = 68)
Sex, no. (%)		
Male	19 (63%)	58%*
Female	11 (37%)	42%
Age, years (range)	69 (30–99)	67 (26–99)*
Normal ECG, no. (%)	8 (27%)	19 (28%)
Mean QRS duration, x (msec)	123.5 ± 32	N/A
≤120 ms	12 (40%)	19 (28%)
>120 ms	18 (60%)	49 (72%)
RBBB	16 (53%)	31 (45%)
RBBB alone	5	14
Bifascicular (RBBB + Lt ant hemiblock)	9	19
Bifascicular + long PR	2	N/A
Left bundle branch block	1 (3%)	10 (15%)
Intraventricular conduction delay	4 (13%)	8 (12%)
PR interval, x (msec)	183.7 ± 48	N/A
Left ventricular ejection fraction, x (%)	55.3 ± 11	N/A
<35%	3 (10%)	
Asystole duration, sec	9.8 ± 4.9 (4–20)	8.4 ± 6.2* (2.1–36)

BIDMC = Beth Israel Deaconess Medical Center, Boston, Massachusetts; N/A = not applicable; RBBB = right bundle branch block; UoM = University of Maastricht, the Netherlands.

*Calculation based on available data.