Pacemaker syndrome without a pacemaker

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Abstract

This article presents a case report of pacemaker syndrome without a pacemaker, or pseudopacemaker syndrome. This syndrome refers to a hemodynamic complex similar to the one caused by pacemaker syndrome, that occurs in patients without a cardiac stimulation device. It results from atrial systole during or very close to ventricular systole, which causes atrial contraction against the closed atrioventricular (AV) valves, leading to increased intra-atrial pressure with retrograde transmission to the pulmonary veins and superior vena cava. This causes arterial vasodilation, decreased systolic ventricular volume, a fall in cardiac output, pulmonary congestion and atrial arrhythmias. Its clinical presentation ranges from dyspnea with exertion to full-blown congestive heart failure. Each patient with this syndrome requires personalized treatment, which depends on the triggering cause. Several reports have documented that symptomatic patients with first-degree AV block with a very long PR interval (generally > 300 ms) benefit from stimulation therapy. **(Acta Med Colomb 2023; 48. DOI: https://doi.org/10.36104/amc.2023.2880).**

Keywords: pseudopacemaker syndrome, first-degree atrioventricular block, atrioventricular dyssynchrony, dual chamber pacing.

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Introduction

The occurrence of heart failure signs and symptoms in patients with pacemakers tends to be attributed to different pathophysiological mechanisms. Pacemaker syndrome is a relatively frequent clinical condition today, with a prevalence ranging from 5 to 80% and a total incidence of 18%, 16% during the first year after device implantation, as described in the Mode Selection Trial (MOST) (1, 2).

This condition is infrequently diagnosed and causes impaired quality of life for the patients, especially due to a functional class deterioration, which may progress to frank heart failure (3). This syndrome may occur at any age, predominantly in elderly patients and those with structural heart disease, and it manifests at any time after pacemaker implantation (2).

Two clinical conditions have been reported to produce a symptomatic hemodynamic complex similar to pacemaker syndrome: first-degree atrioventricular (AV) block with an extremely prolonged PR interval (generally > 300 ms) and a node rhythm faster than the atrial rate (4). These conditions may result in delayed atrial contraction, during or after ventricular contraction, leading to hemodynamic consequences similar to those caused by artificial pacing with retrograde AV conduction. Both conditions can be solved with restoration of normal AV node conduction, with dual-chamber pacing, in which the AV interval is established at the most physiological length possible; the node rhythm can be corrected with just atrial stimulation (5).

This pacemaker syndrome-like clinical condition in patients without a stimulation device is known as pacemaker syndrome without a pacemaker or pseudopacemaker syndrome. The incidence of this syndrome is not well known and, ultimately, inaccurate, as this is an underdiagnosed entity (6). The goal in presenting this case report is to define the pacemaker syndrome without a pacemaker, discuss its pathophysiological mechanisms, present a clinical case, and establish treatment options.

Case presentation

This was an 88-year-old man with a history of surgically revascularized coronary disease and hypertension. He had a two-month history of dyspnea on exertion along with fatigue and exercise intolerance, which had worsened over the previous week, leading him to seek care in the emergency room.

On admission, the patient was hemodynamically stable; his physical exam showed grade I jugular vein distension, extinguished vesicular murmur, crackles in both pulmonary bases and symmetrical grade II edema of the lower extremities. Laboratory tests (complete blood count, kidney function, electrolytes) were within normal limits. An electrocardiogram showed first degree AV block with a PR interval of 466 ms, alternating with a Wenckebach phenomenon.

There were no significant findings on the paraclinical tests (Figure 1-A).

During his hospitalization, heart failure treatment was instated with no improvement. Further studies were conducted including an ischemia detection test, which was negative, with preserved ventricular function. A 24-hour Holter test reported sinus rhythm, first-degree AV block with a prolonged PR interval up to 500 ms, and second-degree Mobitz I AV block, without intraventricular conduction disorders (Figure 1-B). A transthoracic echocardiogram showed preserved ventricular function, an ejection fraction of 60%, normal right ventricular function and no significant valve disease.

He was seen by the electrophysiology group who, considering his clinical signs and symptoms and electrocardiographic results, proposed a diagnosis of pacemaker syndrome without a pacemaker, with an indication for dual-chamber pacemaker implantation. It was decided to implant the device, with the patient and family's informed consent. A dual-chamber pacemaker was implanted with no complications, leaving the ventricular electrode in the septum, which was programmed in DDD mode (Figures 1-C and 2). Once the device was implanted with the stated pacing mode, the patient's symptoms disappeared. After six months of clinical follow up, the patient continued to be in functional class I/IV, asymptomatic.

Discussion

The definition of pacemaker syndrome has evolved over time, enriched by various authors. It was first described in 1969 by Mitsui et al., who defined it as a group of symptoms associated with single-chamber pacing (VVI), including dyspnea, fatigue, syncope and congestive heart failure. It was correlated with an inadequate pacing rate, which ultimately induces AV dyssynchrony (3). In 1991, Shüller et al. defined it as a syndrome occurring in patients with pacemakers, caused by inappropriate AV activation, without excluding dual-chamber pacing modes with suboptimal programming from this definition (5, 7). Although most of the adverse effects reported after pacemaker implantation have been associated with ventricular pacing, they have also been reported during atrial (AAI) and even dual-chamber (DVI, DDI, and much less frequently, DDD) pacing (8). In 1994, Furman redefined this condition as: the result of incomplete restoration of the normal cardiac depolarization pattern, with abnormal atrioventricular contraction and disruption of the physiological AV interval, with retrograde atrial activation (5, 8).

The emergence of a clinical syndrome with similar electrophysiological and hemodynamic characteristics in patients without a heart pacing device has created the need to study its causes and consequences in depth, representing a diagnostic challenge for modern medicine.

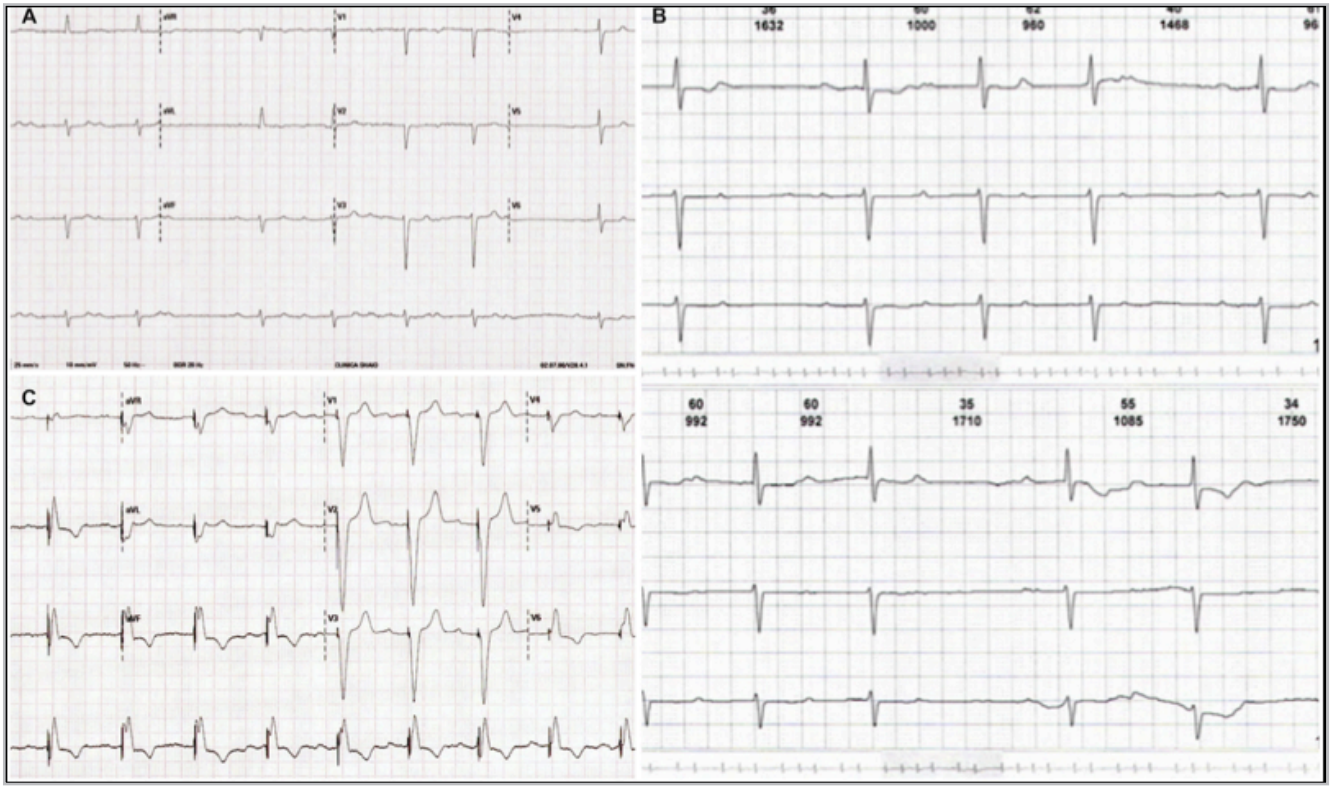

Figure 1. A: *EKG on admission: shows first-degree AV block with a prolonged PR interval of up to 466 ms which alternates with second-degree Mobitz I AV block.* **B:** *24-hour Holter monitoring: sinus rhythm, first-degree AV block with a PR interval of up to 500 ms, alternating with second-degree Mobitz I AV block.* **C:** *EKG after implanting a dual-chamber pacemaker programmed in DDD mode, with adequate pacing and sensing..*

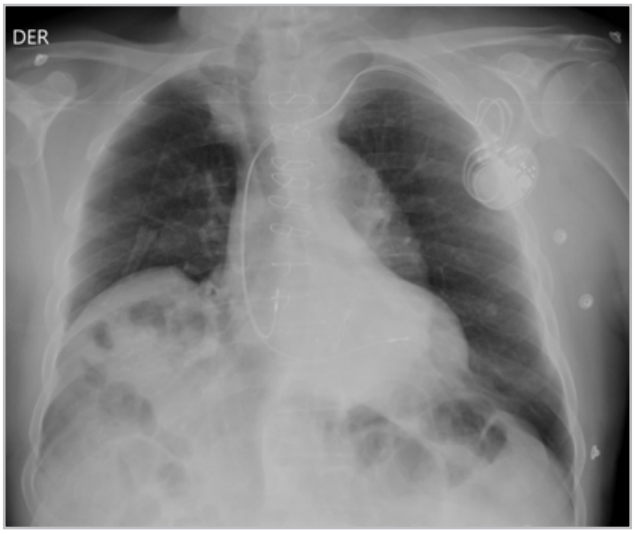

Figure 2. *Chest x-ray after implanting a dual-chamber pacemaker, with the ventricular electrode in the septum.*

The overall concept of pseudopacemaker syndrome includes three fundamental elements: loss of AV synchrony, retrograde ventriculoatrial conduction, and an inappropriate heart rate for responding to the body's metabolic needs, all in patients without a pacemaker (7, 9).

Some conduction disorders that affect the timing of atrial and ventricular conduction may cause this syndrome, which has been reported to be a rare but well recognized complication of first-degree AV block with a very long PR interval (generally > 300 ms), when the P wave is very close to the previous QRS complex, resulting in atrial systole during or very close to ventricular systole, producing atrial contraction against the closed AV valves with increased intra-atrial pressure. This pressure is transmitted in a retrograde fashion to the pulmonary veins (left atrium) and superior vena cava (right atrium), producing atrial natriuretic peptide release, causing arterial vasodilation, decreased systolic volume, reduced cardiac output, pulmonary congestion and atrial arrhythmias, with a concomitant fall in systolic right and left ventricular function, as well as arterial pressure. All these hemodynamic, neurohumoral and electrophysiological mechanisms are responsible for the signs and symptoms of this syndrome (3, 6, 10, 11).

It is important to highlight that the diagnosis of this condition is eminently clinical, based on a detailed history and thorough physical exam. Electrocardiographically, it may manifest with the onset of a retrograde P wave or retroconduction wave (9). Despite not having established diagnostic criteria, the essential pillars, from a clinical perspective, are listed in Table 1 (9).

The clinical signs and symptoms of pacemaker syndrome without a pacemaker in our patient with first-degree AV block with an extremely long PR (up to 500 ms), were similar to the classic characteristics of pacemaker syndrome in patients with inappropriate pacing, retrograde conduction and AV dyssynchrony. A P wave very close to the previous QRS causes the same hemodynamic disruption as VVI pacing with retroconduction. The main clinical manifestation reported is dyspnea, which may or not be coupled with the complex of fatigue, palpitations, diaphoresis, presyncope or syncope, hypotension or high pulmonary pressure. In our case, the patient presented with functional class deterioration, fatigue, exercise intolerance and signs of fluid overload. In these patients, the most prominent symptoms result from decreased cardiac output and severe arterial hypotension, which may end up in frank congestive heart failure, as the retrograde VA conduction causes a "negative atrial kick," with a hemodynamic effect due to the loss of AV synchrony (12). However, some patients have more subtle signs and symptoms associated with the electrocardiographic findings described.

Table 1. *Pseudopacemaker syndrome: signs and symptoms.*

Dyspnea on exertion	Syncope or presyncope
Cannon A waves	Diaphoresis
Arterial hypotension	Fatigue, tiredness
Hepatic congestion	Lower extremity edema
Palpitations	Pulmonary congestion

Each patient with this syndrome requires personalized treatment which depends on the underlying cause, and its management is based on restoring physiological AV conduction. Several reports have stated that patients with symptomatic first-degree AV block with a very long PR interval and preserved left ventricular function benefit from dual-chamber pacing, with which adequate AV synchrony can be achieved. In some case reports and uncontrolled clinical trials including patients with a similar clinical picture and electrocardiographic findings, this intervention has been associated with improved signs, symptoms and quality of life, as occurred in the clinical case we presented (3).

The 2012 ACC/AHA/HRS guidelines do not recommend pacemaker implantation in patients with asymptomatic first-degree AV block, except in patients with neuromuscular diseases like myotonic dystrophy. However, the same guidelines recommend pacemaker implantation in patients with symptomatic first-degree AV block (12). Moreover, the European Society of Cardiology (ESC) indicates, as a class IIa and level of evidence C recommendation, permanent pacemaker implantation in patients with persistent symptoms attributable to first-degree AV block (PR > 300 ms) (13,14).

His bundle pacing has recently gained importance in the field of electrophysiology. This strategy has been proposed as the only cardiac pacing mode able to precisely reproduce the synchronous activation of the left ventricle, with its trifascicular nature: 1. high anterior paraseptal wall, 2. central left upper interventricular septum, and 3. distal posterior paraseptal wall. This is believed to be a potential treatment alternative appropriate for patients like the one presented in this case. However, there are some limitations in its implementation (15).

Conclusions

In this article, we present the case of a patient with pacemaker syndrome without a pacemaker, which is an underdiagnosed clinical condition. However, it is found relatively frequently in clinical practice, especially in electrophysiology services. The main cause is first-degree AV block with a PR interval greater than 300 milliseconds, and it is characterized by variable clinical symptoms like dyspnea, functional class deterioration, tiredness, fatigue, atrial arrhythmias, and even heart failure. According to the international guidelines, dual-chamber pacing is recommended for patients with this group of symptoms associated with this syndrome.

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