Case Report

Exertional dyspnea and orthopnea in a patient with a pacemaker: an easily overlooked diagnosis

SE Butler, A Voskoboinik & HC Patel

Abstract

Acute dyspnea represents one of the most common presentations to the emergency department. Identifying the cause of this non-specific symptom among patients with complex medical histories can be challenging.

This case report describes a patient in their 60s with worsening dyspnea on exertion, fatigue and orthopnea on the background of a single chamber pacemaker. Our case demonstrates an often-missed diagnosis of pacemaker syndrome which can be identified on the bedside electrocardiogram (ECG). This is a disease with high morbidity and yet is easily treated once diagnosed.

Keywords

Permanent pacemaker, pacemaker syndrome, electrocardiogram

Key points

- Evidence of new-onset heart failure following PPM insertion should be evaluated with ECG and echocardiogram for possible pacemaker syndrome
- Given the significant morbidity associated with pacemaker syndrome and the highly effective treatment, clinicians should maintain a high suspicion of the condition with a VVI PPM, particularly where additional risk factors are present.
- The ECG remains an important tool in the assessment of any cardiac symptom and is not replaceable by a PPM interrogation.

Case presentation

A patient in their 60s was reviewed in an outpatient clinic with deterioration of dyspnea on exertion, fatigue and orthopnea that had been progressing over the course of a year. Their exercise tolerance had substantially reduced to 50 meters, from a kilometer the previous year. The patient's medical history included an orthotopic heart transplant (20 years prior for a failing Fontan repair for complex congenital heart disease), transposition of the great arteries (previously repaired with a Fontan procedure), persistent atrial fibrillation and chronic kidney disease. They had a single chamber (right ventricle) permanent pacemaker (PPM) inserted two years prior, for bradycardia in the setting of symptomatic sinus node dysfunction. The original intention was to insert a dual-chamber device; however, the atrial lead implant was unsuccessful due to a significantly dilated atria in the context of orthotopic heart transplantation using the bi-atrial anastomosis technique.

At the time of review, the patient's blood pressure was 130/75mmHg and heart rate 60 beats/min. The examination revealed peripheral edema and an elevated jugular venous pressure. The pathology results including full blood count, thyroid function tests and iron studies were all within normal limits. Chest radiograph showed clear lung fields.

A pacemaker check showed a normal functioning device set at VVIR 60. Right ventricular (RV) pacing burden was 62% having increased from 27% in the first year of implant.

A 12-lead electrocardiogram (ECG) is shown in Figure 1.

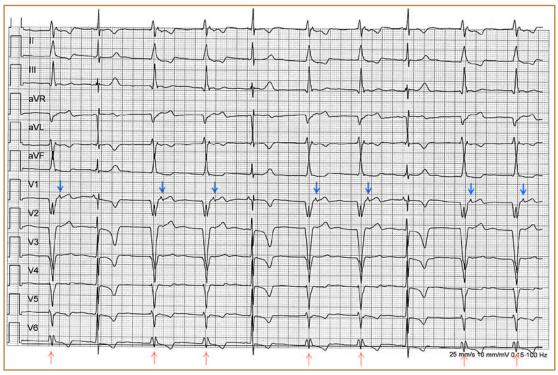

Figure 1. The ECG shows two QRS morphologies, regularly paced broad QRS complexes (red arrow), alternating with narrow QRS complexes. Retrograde P-waves are present in leads V1, II and aVL, but best appreciated in V1 (blue arrow).

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Questions

What is the likely diagnosis?
What is the most appropriate treatment?

Interpretation

The ECG (Figure 1) demonstrates an intermittent ventricular-paced rhythm at 60 beats/min. There are two QRS morphologies. The first (3rd, 4th, 6th, 7th, 9th, and 10th, as well) beat is a wide ventricular paced QRS complex with a left bundle branch block (LBBB) pattern. Immediately following the paced QRS complexes, a P-wave is seen (best identified in lead V1) suggesting retrograde or ventricular-atrial (VA) conduction. The second (5th and 8th as well) beat is a sinus beat with a P-wave, which conducts physiologically through the atrioventricular node resulting in a narrow intrinsic QRS complex.

The findings of increased RV pacing asynchronous to atrial activity and new onset of heart failure symptoms support a diagnosis of pacemaker syndrome.

Clinical course

The differential diagnoses for the patient's non-specific symptoms are wide and include transplant rejection, cardiac allograft vasculopathy, pacing-induced cardiomyopathy and constrictive pericarditis. These were effectively excluded by the additional findings of stable ventricular function on serial echocardiography, no rejection on endomyocardial biopsies and on cardiac CT coronary angiogram unobstructed coronary arteries with no pericardial thickening Echocardiography showed low-normal left ventricular function with severe bi-atrial dilatation in keeping with a cardiac transplant. There was mild-moderate aortic regurgitation, moderate-severe tricuspid regurgitation due to annular dilatation and mild-moderate pulmonary hypertension (peak RVSP 57 mmHg).

The patient underwent successful insertion of an atrial pacing lead. Following this, the patient reported complete resolution of their heart failure symptoms. They were 99% atrial pacing and <1% ventricular paced. A repeat ECG confirmed regular atrial pacing with intrinsically conducted narrow QRS complexes. Routine surveillance echocardiography showed unchanged ventricular function and normalized estimated RV systolic pressure.

Discussion

Pacemaker syndrome does not have a universal definition. Commonly it can be diagnosed when symptoms (fatigue, dyspnea, dizziness) occur secondary to loss of atrio-ventricular (AV) synchrony. The incidence among patients with sinus node dysfunction and a single-chamber ventricular PPM is at least 20%,[1] but the overall incidence of pacemaker syndrome has likely decreased with guidelines promoting physiological dual chamber pacing.[2] A high index of suspicion is often required to successfully make the diagnosis.

AV dyssynchrony results in non-physiologically timed atrial contractions with loss of atrial contribution to ventricular filling and decreased cardiac output. Atrial systole contributes 20-30% of normal cardiac output in healthy individuals and up to 50% in the setting of decreased ventricular compliance.[3] Furthermore, sudden pressure changes due to the atrium contracting against closed AV valves can cause venous cannon A-waves and activate autonomic reflexes leading to palpitations and symptomatic hypotension. The severity of these symptoms is influenced by the patient's underlying heart rate, the presence of comorbidities including structural heart disease and the persistence of dyssynchrony.[4] Mild AV dyssynchrony, may be asymptomatic.

A major contributor to AV dyssynchrony is retrograde (VA) conduction, whereby electrical conduction from the ventricles or AV node is transmitted through to the atria and therefore leads to non-physiologically timed atrial contraction. It is thought that VA conduction co-exists in 35-55% of patients with normal AV conduction at baseline[5,6] and in over 60% of patients with sinus node dysfunction.[7] The presence of complete AV block does not exclude the possibility of retrograde conduction even in the absence of a concealed accessory pathway.[8]

Pacemaker syndrome is a diagnosis of exclusion, however, identifying the cause of non-specific symptoms is challenging in the setting of a complex medical history including heart transplant. After excluding other plausible causes, management for patients with a single-chamber PPM involves upgrading to a dual chamber system. The original PPM implant in this case was complicated by a failed atrial lead insertion due to anatomical difficulties, thus adding to the complexity of possible revision. Whilst large trials have demonstrated no mortality benefit of the dual chamber PPM over VVI pacing in sinus node disease,[1,9,10] international guidelines advocate for dual chamber PPM insertion first-line given the possibility of significant morbidity.[2] This includes poorer exercise capacity, pacemaker syndrome and higher rates of atrial fibrillation and stroke.

The present case illustrates a reversible syndrome that can be easily rectified if diagnosed. The identification of retrograde P-waves on ECG provided an important clue to help identify the underlying disease process. The ECG is an important non-invasive tool in the assessment of cardiac symptoms and can yield crucial diagnostic clues in conjunction with PPM interrogation.

Declaration of competing interests

Nothing to declare.

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