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Dual Chamber Pacing Relieves Obstruction in Japanese-Variant Hypertrophic Cardiomyopathy

Omar Wever-Pinzon, MD*, Jorge Romero, MD, and Juan P. Cordova, MD

Division of Cardiology, University of Utah, Salt Lake City, UT.

Abstract

Japanese-variant or apical hypertrophic cardiomyopathy (HCM) is a specific type of HCM, first described in Japan and initially thought to carry a benign prognosis. However, current evidence suggests that these patients experience severe symptoms and are at increased risk of ventricular arrhythmias and death, especially in the presence of an apical akinetic chamber. The management of patients who do not respond to medical therapy is challenging. We describe a patient with Japanese-variant HCM, with an apical akinetic chamber and severe symptoms who failed medical therapy. The use of dual chamber pacing relieved obstruction and significantly improved the patient's symptoms.

Keywords

hypertrophic cardiomyopathy; HOCM; dual chamber pacing

INTRODUCTION

Obstructive hypertrophic cardiomyopathy (OHCM) is a complex cardiac disease with several pathophysiologic effects leading to substantial morbidity. Symptoms include dyspnea, exercise intolerance, chest pain, syncope, and general fatigue. The most common presentation involves septal hypertrophy with left ventricular (LV) outflow tract obstruction due to systolic anterior motion of the mitral valve.¹ Less common variants of HCM are midventricular and apical HCM. These have been associated with distal LV apical aneurysm and an apical akinetic chamber.² Previous reports on mid and apical HCM found that patients with midventricular thickening and subsequent encroachment of the LV cavity with small LV volumes were more likely to have severe symptoms. Patients with an apical akinetic chamber are more symptomatic and experience monomorphic ventricular tachycardia more often.²

Several treatment options are available to patients with mid and apical HCM. These include pharmacologic, surgical, and pacing therapies. Pharmacologic therapy in HCM involves beta-blockade or verapamil.^{1,2} Beta-blockade is the initial first-line treatment. It causes negative inotropy thereby improving dynamic LV outflow obstruction and decreasing ejection acceleration. Verapamil, a potent calcium channel blocker, also functions as a negative inotropic agent. It decreases the LV outflow tract gradient and improves symptoms and exercise tolerance. Disopyramide has also been used as a therapeutic option in patients

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*Address for correspondence: Division of Cardiology, University of Utah, 30 North 1900 East, Room 4A100, Salt Lake City, UT 84132. weveromar@hotmail.com.

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with OHCM. Disopyramide is a type 1 antiarrhythmic drug, with potent negative inotropic effects.¹ In normal patients, it decreases fractional shortening by 28%. Disopyramide works as a sodium channel blocker and may have calcium channel blocking properties that may benefit patients who are refractory to beta-blockade and would otherwise require surgical septal myectomy or alcohol septal ablation.

Septal myectomy and alcohol septal ablation have also been included as therapeutic options; however, their use has been debated due to risks of further damage to the myocardium.² Pacing is a less invasive option that has shown to be promising in patients with OHCM. Dual chamber (DDD) pacing with a short atrioventricular (AV) delay has shown to “improve exercise capacity and clinical symptoms in patients with severe exertional dyspnea and fatigue due to hypertensive hypertrophy and distal cavitory obliteration.”³ The AV delay function has been shown to allow for maximum ventricular preexcitation but is sufficiently long not to interfere with left atrial emptying.⁴

We describe the case of 66-year-old man with midapical OHCM with an akinetic apical chamber who was referred for DDD pacing with short AV delay, after medical therapy failed to relieve the midcavitory obstruction and symptoms. DDD pacing improved his cardiac symptoms, functional class, and reduced the apical akinetic chamber size.

CASE REPORT

A 66-year-old Asian man with a history of hypertension and recurrent syncope was diagnosed with Japanese-variant Hypertrophic Cardiomyopathy (HCM) after collapsing on the treadmill during an exercise stress test, which was performed secondary to the patient's complaints of exertional angina.

Physical examination

On examination, the patient was found to have a blood pressure of 120/70 mm Hg with a pulse of 70 b·min⁻¹. His chest was clear to auscultation. Cardiac examination found regular rhythm and no murmur with standing or Valsalva maneuver. Abdominal examination showed no organomegaly. Extremities were warm, without edema and with good capillary refill. Distal pulses were 2+ and symmetrical.

Laboratory findings

The electrocardiogram showed a normal sinus rhythm, LV hypertrophy with diffuse T-wave inversions throughout the precordial leads.

On echocardiographic examination, the patient was found to have mid and apical LV hypertrophy consistent with subbasal HCM. There was thickening of the midsegments up to 22 mm in the anterior wall with no thickening of the base. Also, thickening of the apex and an apical severely hypokinetic chamber of 1.5 × 1 cm was seen. The use of contrast was necessary to better assess the apical hypokinetic chamber. Doppler imaging revealed a paradoxical diastolic jet flow and a peak systolic mid-LV gradient of 50 mm Hg.

The patient's history of multiple syncopal episodes and failure to respond to beta-blockade led the treating physicians to suggest and implant a dual chamber pacemaker. Six months after the implantation of the DDD pacemaker, an echocardiogram showed an apical akinetic chamber that was significantly smaller compared with the patient's previous echocardiogram. There was increased systolic flow through the midcavitory obstruction with nearly complete emptying of the apical chamber. The midcavitory gradient was abolished (Fig. 1). His functional class improved from New York Heart Association class III to II.

DISCUSSION

Medical therapy is generally the first-line therapy used in patients with OHCM, with success in more than two-thirds of patients.⁵ Beta-blockade, verapamil, and disopyramide are generally used in patients with OHCM. However, our patient was unusual because his case involved mid and apical OHCM with an apical akinetic chamber that was refractory to conventional medical therapy. Previous research on patients with mid and apical HCM found that DDD pacing with a short AV delay helped to reduce the mid-LV gradients and improve symptoms.¹ The AV delay allows for maximum ventricular preexcitation but does not interfere with left atrial emptying. The authors of this study were unclear as to the exact mechanism by which DDD pacing reduced the intracavitary pressure gradient but found that correct lead placement led to success in DDD pacemaker therapy.

This case report provides further insight in the pathophysiology and cause of severe symptoms in this unusual type of HCM. The response of our patient to DDD pacing provides additional evidence to support its use in patients with midventricular and apical HCM who do not respond to medical therapy.

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FIGURE 1.

Left panel shows diastolic and systolic frames pre-dual chamber pacing, revealing an apical akinetic chamber. Post- dual chamber pacing, on the right panel, shows complete emptying of the apical akinetic chamber.