

# Idiopathic Isolated Right Ventricular Apical Hypertrophy

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## INTRODUCTION

Isolated right ventricular hypertrophy is extremely rare and reports in the literature are very sparse. We report a case of focal apical right ventricular hypertrophy without involvement of the left ventricular cavity or the inter-ventricular septum.

## CASE

A 50 year old male, asymptomatic, normotensive patient on routine medical evaluation was found to have inverted T-waves on the electrocardiogram (Figure 1). He had no history of angina or any other symptoms. However, despite complete lack of symptoms, because of the abnormality on the electrocardiography (ECG) an echocardiography was carried out for further evaluation.

Echocardiography revealed focal hypertrophy of the right ventricular (RV) apex almost obliterating the apex (Figure 2A) but sparing the inter-ventricular septum. There was no concomitant left ventricular hypertrophy (wall thickness 1.1 cm) (Figure 2B) and there was no pulmonary valve stenosis (gradient across pulmonary valve: 2 mmHg). Tricuspid annular plane systolic excursion was measured (24 mm) and was normal. Pulsed tissue Doppler applied to the RV free wall at the tricuspid annular level and at the septal and lateral corners at the mitral annular level revealed normal velocities (10 cm/sec and 12 cm/sec respectively). There was no outflow obstruction or de-

creased RV compliance or RV dysfunction.

## DISCUSSION

RV hypertrophy is usually reported in association with left ventricular hypertrophy which in turn could be secondary to hypertrophic cardiomyopathy (HCM), hypertensive left ventricular hypertrophy (LVH) or other infiltrative conditions.<sup>1</sup> However, isolated involvement of right ventricle is extremely rare in these conditions. The only situation where isolated RV hypertrophy is seen is in presence of idiopathic pulmonary artery hypertension. However, the hypertrophy in these cases is rarely focal.

Isolated apical hypertrophic cardiomyopathy is an extremely unusual nonobstructive hypertrophy that is localized to the cardiac apex.<sup>2</sup> Even in these cases sparing of the left ventricular apex with involvement of only the right ventricular apex is even more uncommon. The most common presenting symptom in patients with apical HCM is chest pain, and some patients can present with palpitation, dyspnea and syncope. The most frequent ECG findings are T-wave inversions in the precordial leads with significant T-wave inversion ( $\geq 10$  mm) are characteristically reported in some patients. However, ST-T abnormalities, atrial fibrillation, first-degree atrioventricular (AV) block, sinus bradycardia, short PR interval without delta waves, pathological Q waves consistent with apical infarction have all been reported.<sup>2</sup>

Apical HCM is generally considered to be a benign condition however; these patients may also develop typical HCM symptoms and are at increased risk of apical myocardial infarction and development of apical aneurysm.<sup>3</sup> It is more common in middle aged males and appears to be more common in the Japanese population.<sup>5</sup> However the true incidence of apical hypertrophy involving only the right side is unknown because of its rarity. It is also difficult to ascertain if isolated focal apical hyper-

Received: October 2, 2017 Accepted: January 22, 2018

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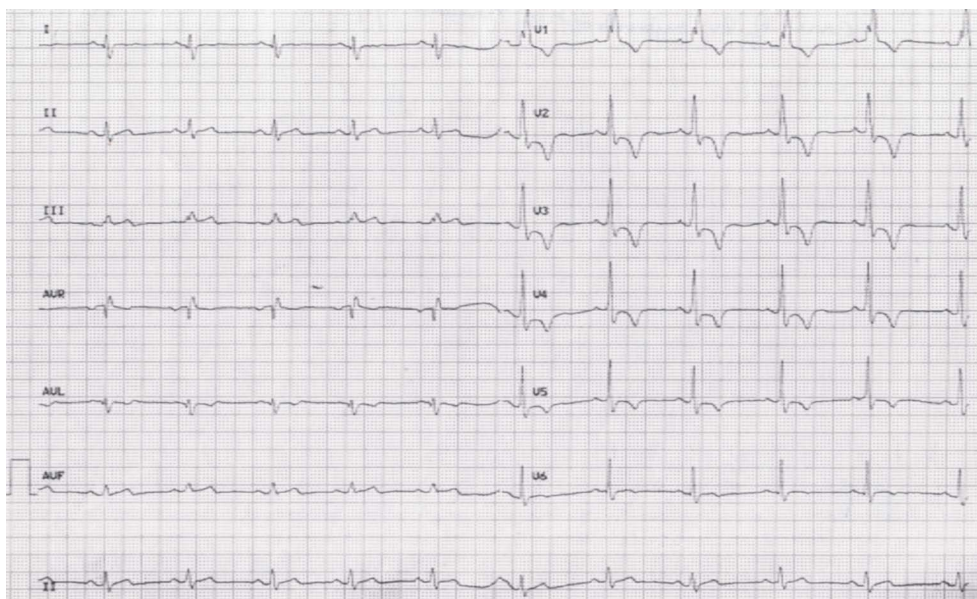


Figure 1. Electrocardiography (ECG) showing T wave inversion in the lateral chest leads.

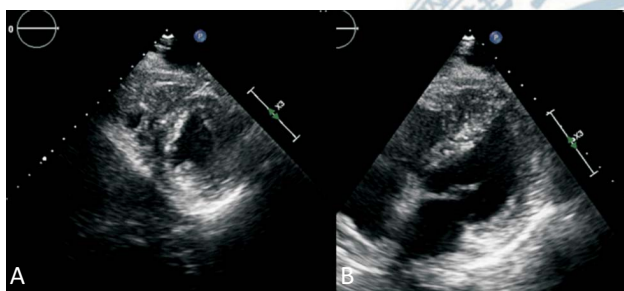


Figure 2. (A) Focal apical hypertrophy almost obliterating the right ventricular (RV) apex. (B) Apical four chamber view showing an uninvolved left ventricular cavity and inter-ventricular septum.

trophy of the right ventricle is merely a stage in the disease which finally involves the left side as well.

Apical hypertrophic cardiomyopathy has been regarded as an atypical phenotype of non-obstructive HCM. The entity is frequently sporadic however, familial distribution with autosomal dominant inheritance has been reported.<sup>2</sup> In these families a sarcomere gene mutation in the alpha-cardiac actin gene has been reported. Cardiac actin mutation has been seen consistently in patients with apical hypertrophy and it appears that there is a definite relationship between this genotype and apical hypertrophy even though the mechanisms by which the actin light chain mutations produce apical HCM are unknown.<sup>2</sup> Recently TNNI3 mutations associated with focal ventricular hypertrophy with marked right-sided

cardiac manifestations has been identified and may explain the presence of isolated focal right ventricular hypertrophy.<sup>4</sup> It has also been suggested that apical hypertrophy can be a result of a single gene defect as well as complex genetics and/or gene-environment interactions.<sup>2</sup>

#### LEARNING POINT

From a clinical perspective these patients usually have a benign course with good long term survival.<sup>5</sup> The findings maybe incidental as most patients maybe asymptomatic and therefore the condition remains under diagnosed and precisely for this reason it is difficult to ascertain what percentage of these patients develop symptoms. However, from a management point of view in the absence of syncopal episodes, arrhythmias, family history of sudden death and absence of high resting RV gradients and significant RV involvement it appears that careful follow up of these patients clinically and with serial echocardiography to monitor the progression of this condition is all that is required at present.

#### CONFLICT OF INTEREST

None.

## FUNDING

None.

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