



Hypertrophic Obstructive Cardiomyopathy of Right Ventricle without Involvement of Left Ventricle

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Case Report

Volume 5 Issue 1

Received Date: December 09, 2020

Published Date: January 06, 2021

DOI: 10.23880/oajc-16000154

Abstract

Hypertrophic obstructive cardiomyopathy localized to right ventricle is rare. There are no diagnostic clinical or electrocardiographic findings. Echocardiography is diagnostic. A case of isolated right ventricular hypertrophic obstructive cardiomyopathy is reported and literature is reviewed.

Keywords: Hypertrophic Cardiomyopathy; Right Ventricle; Systolic Murmur

Introduction

Hypertrophic obstructive cardiomyopathy localized to right ventricle is rare. There are no diagnostic clinical or electrocardiographic findings. Echocardiography is diagnostic.

Case Report

A 35 years female presented with complaint of breathlessness on effort. Pulse was regular at a rate of 90/minute. Systemic blood pressure was 116/86 mm of Hg. Jugular venous pressure and pulse wave from were normal. Apical impulse was normal. There was no right ventricular impulse. No other pulsations were felt. First and second sound sounds were normal. There was no click. There was a grade 2-3/6 ejection systolic murmur over left second and third intercostal space in parasternal region. It showed no selective radiation. There was no respiratory variation. There was no family history of hypertrophic cardiomyopathy or sudden cardiac death.

Electrocardiogram (Figure 1) showed sinus rhythm. P wave negative in leads II, III, aVR, aVF and was positive in leads V₁, V₅, V₆. P-R interval was 160ms. Ventricular rate was 60/min. QRS duration was 80 ms. Mean frontal plane QRS axis was around +150°. Lead aVR showed QS pattern. Lead

V₁ showed rs pattern (6mm/25mm). Lead V₆ showed Rs configuration (14/3mm). Amplitude of R wave was 11mm and 7.5 mm in leads I and aVL respectively. T wave was inverted in leads V₁, V₂. QTc interval was 432 ms. U wave was positive in leads V₁ to V₅.

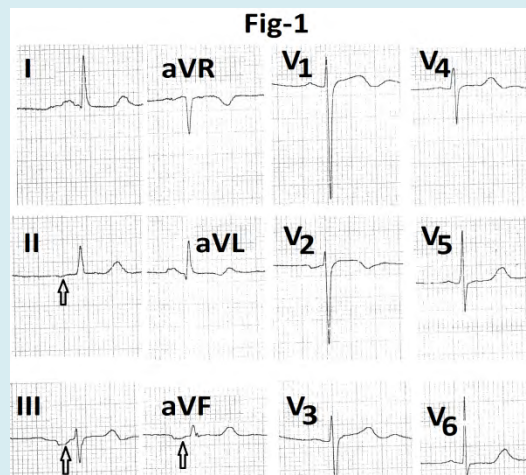


Figure 1: 12 lead electrocardiogram showing P wave inversion in leads II, III, aVF (marked arrow).

On echocardiography, inferior vena cava was normal (1.5 cm) with normal inspiratory collapse (Figure 2). Hepatic

vein flow pattern was normal (Figure 3). Tricuspid flow was normal (E- 0.58m/s, A- 0.49 m/s. Deacceleration time 300ms) (Figure 4). There was minimal TR with peak systolic gradient of 34+5mm Hg) (Figure 5). Tricuspid annulus peak systolic excursion was 3.6 cm (Figure 6). Apical four chamber view revealed increased thickness and systolic obliteration of apex of right ventricle (Figure 7). Colour Doppler evaluation revealed mid cavity turbulence in apical part of right ventricle (Figure 8). Doppler evaluation revealed characteristic late peaking dogger-shaped appearance with right ventricular mid cavity gradient of a 9 mm Hg (Figure 9). Short axis view revealed turbulence in right ventricular outflow (Figure 10) with a peak systolic gradient of 18.2 mm Hg (Figure 11). Pulmonary valve was normal with no additional gradient (Figure 12). Evaluation of left side of heart was within normal limits. M- Mode dimension of left atrium was 2.8cm (Figure 13). End diastolic and end-systolic M-mode dimensions of left ventricle were 4.4 cms and 2-7 cms respectively. There was no left ventricular hypertrophy (Figure 14). Doppler evaluation of mitral valve flow was normal (Figure 15). Tissue Doppler evaluation of lateral mitral annulus (Figure 16) and medial mitral annulus (Figure 17) were normal. Flow across aortic valve was normal (Figure 18).

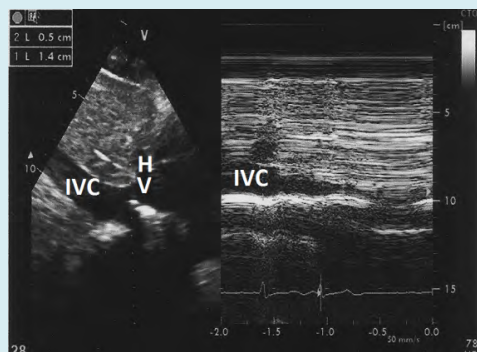


Figure 2: Subcostal long axis view showing normal dimension and normal inspiratory collapse of inferior venacava (IVC).

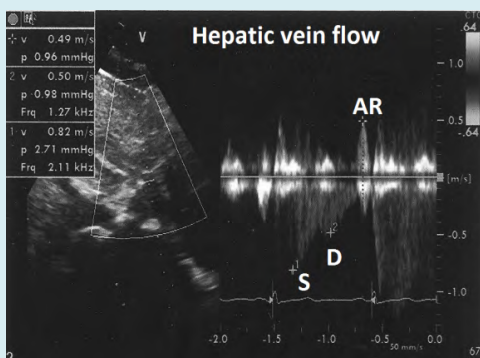


Figure 3: Showing normal hepatic vein flow pattern. S- Systolic, D- Diastolic, AR-atrial reversal.

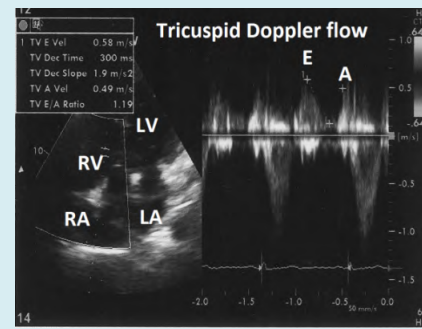


Figure 4: showing normal tricuspid Doppler flow. E- early diastolic flow, A-flow during atrial systolic.

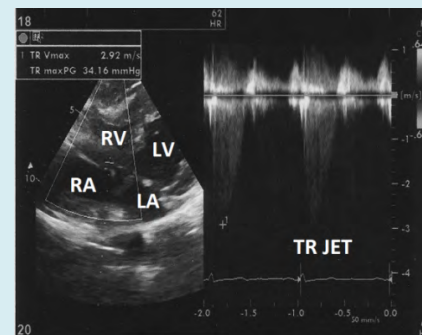


Figure 5: Showing Doppler evaluation tricuspid regurgitation Jet.

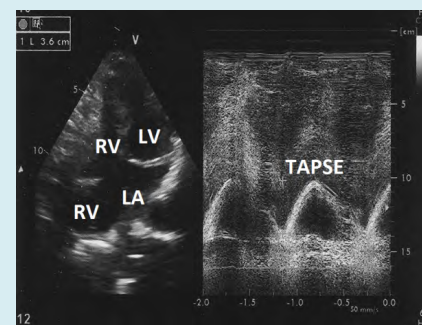


Figure 6: M- mode echocardiogram showing tricuspid annulus peak systolic excursion of 3.6 cm.

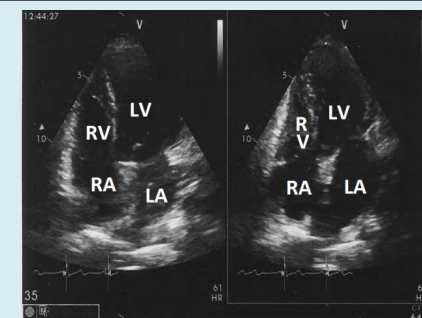


Figure 7: Apical four chamber view showing increased thickness and systolic obliteration of apex of right ventricle.

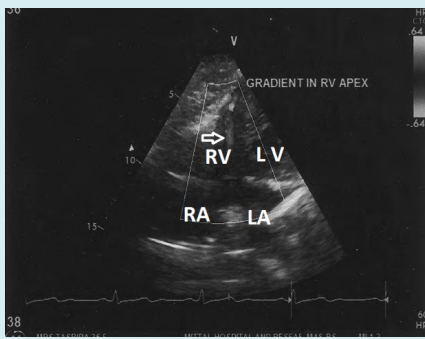


Figure 8: Colour Doppler evaluation showing turbulence in right ventricular apex (marked arrow).

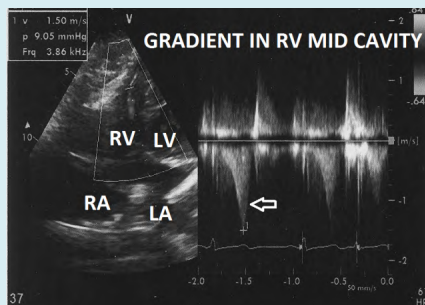


Figure 9: Doppler evaluation showing right ventricular mid cavity systolic gradient of 9mm Hg (marked arrow).

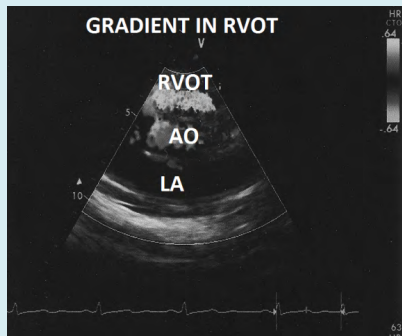


Figure 10: Short axis view showing turbulence in right ventricular outflow tract.

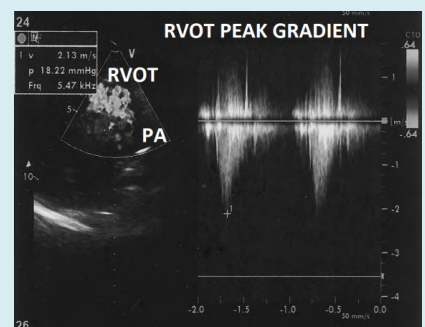


Figure 11: Doppler evaluation showing right ventricular outflow tract peak systolic gradient of 18.2 mm Hg.

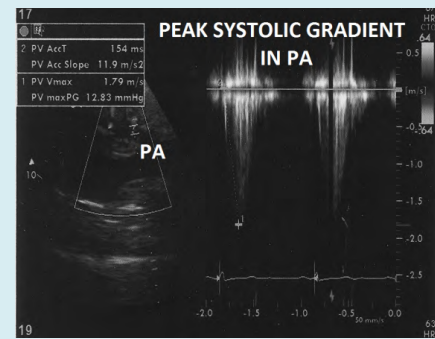


Figure 12: Doppler evaluation of pulmonary artery showing peak systolic gradient of 12.8 mm Hg.

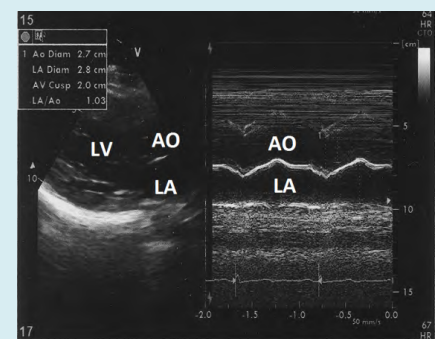


Figure 13: M-mode echocardiogram showing left atrial dimension of 2.8 cm.

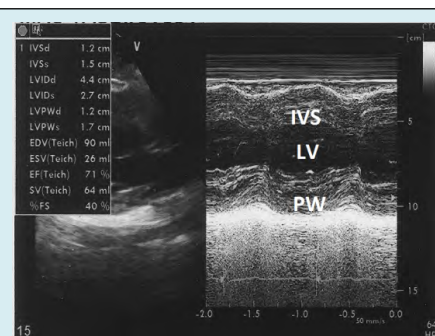


Figure 14: M-mode echocardiogram showing normal dimension and wall thickness of left ventricle.

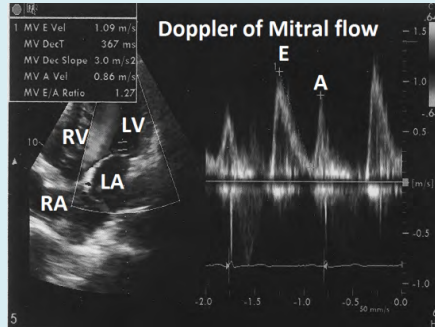


Figure 15: Showing Doppler evaluation of mitral flow showing normal pattern.

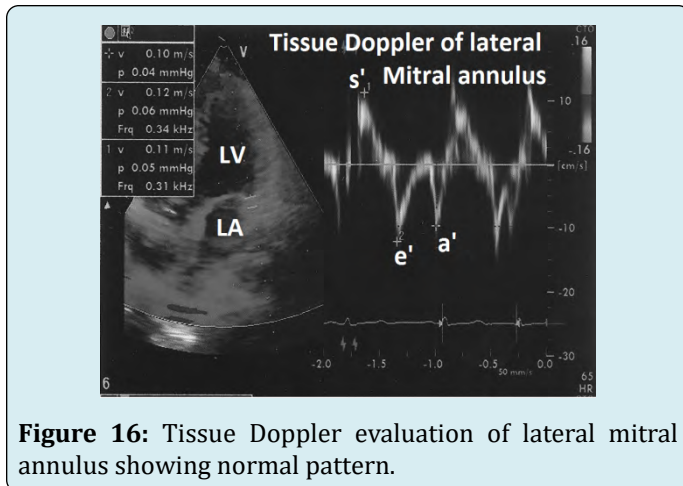


Figure 16: Tissue Doppler evaluation of lateral mitral annulus showing normal pattern.

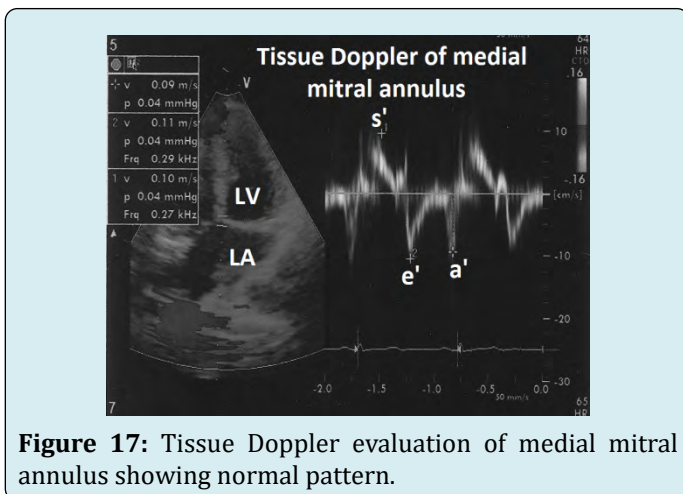


Figure 17: Tissue Doppler evaluation of medial mitral annulus showing normal pattern.

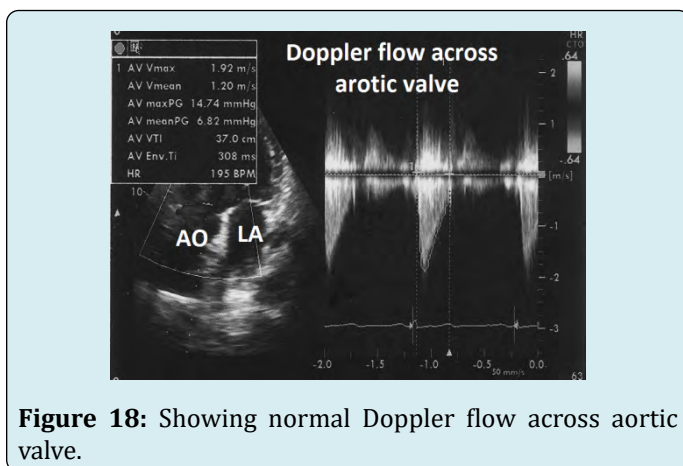


Figure 18: Showing normal Doppler flow across aortic valve.

Discussion

Right ventricle can be involved in some cases of left ventricular hypertrophic cardiomyopathy [1-5]. Isolated right ventricular hypertrophic obstructive cardiomyopathy is, however, rare. There are isolated case reports in literature [6-10]. Most of the reported cases are those who are

symptomatic because of severe hypertrophy. Knowledge about this entity is, therefore, incomplete.

Exact prevalence is not known as there is no prospective population based or autopsy study focusing on this field. There is no information about genetic basis because of report of small number of cases and back of genetic studies in these reported cases. There is only one case report of history of hypertrophic obstructive cardiomyopathy in two brothers of a case of hypertrophic cardiomyopathy of the right ventricle [3]. One brother of another patient had none Compaction of left ventricle [9].

Patients with isolated mild or moderate hypertrophy with or without mild to moderate obstruction may not have any specific symptoms. Those with severe obstruction are likely to have angina or breathlessness on effort similar to any other significant right ventricular outflow tract obstruction.⁶⁻⁹ Arterial pulse is likely to be normal. There are no reports of techy arrhythmias. One patient had junctional rhythm. Jugular venous pulse pressure is likely to be normal except in cases with severe hypertrophy of right ventricle. Such cases may show prominent 'a' wave in jugular venous pulse. Clinically detectable systemic venous congestion is not seen.

Apical impulse is normal. Left parasternal heave is uncommon unless outflow obstruction is severe [6,7,9]. There are no other abnormal pulsations. Commonest finding is grade 2-3/6 ejection systolic murmur over left 3rd intercostal space [6-9]. Murmur may or may not show clear inspiratory increase in intensity. Systolic thrill is unusual [3,4]. First and second sounds are normal [3,7,8,]. There is no click. Right sided fourth heart sound is rare.⁶ its right ventricular origin may be difficult to appreciate.

Skiagram of chest is unremarkable. Electrocardiogram may be unremarkable with mild to moderate hypertrophy. Right axis deviation and right ventricular hypertrophy suggest severe right ventricular hypertrophy. Unexplained turbulence in right ventricular outflow tract is the commonest echocardiographic finding that raises suspicion to the presence of this entity. Right ventricular outflow tract lies perpendicular to cursor in short axis view. Turbulent flow is, therefore, likely to be recorded on either side of base line. Therefore, calculation of exact gradient, by echocardiography, may be difficult. Exclusion of any additional gradient across pulmonary valve or in main or branch pulmonary arteries is necessary to exclude any secondary cause for right ventricular hypertrophy. Cardiac catheterization is usually not indicated.

Most of the echocardiographers focus on left ventricle. Isolated right ventricular hypertrophic cardiomyopathy is,

therefore, likely to be missed unless specifically looked for. Isolated right ventricular hypertrophic cardiomyopathy without obstruction is more likely to be missed as it does not produce any turbulent flow that may attract attentions of echocardiographer exact incidence, clinical profile, electrocardiographic findings and natural history will be clear only on study of larger number of cases. Like classical hypertrophic obstructive cardiomyopathy, patients may have partial relief of symptoms with betablockers. Non responders may need surgical intervention [4,5].

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