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(CASE REPORT)

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Pediatric restrictive Cardiomyopathy presenting with congestive heart failure: Rare case report and literature review

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Abstract

Restrictive cardiomyopathy (RCM) is an uncommon form of cardiomyopathy and is characterized by impaired diastolic filling of the ventricles due to increased myocardial stiffness and a normal ventricular systolic functions. In India, RCM is frequently observed in the young and endemic endomyocardial diseases are seen in a significant number of cases. RCM is the least common of all the cardiomyopathies among pediatric patients. RCM has a poor prognosis and commonly requires a cardiac transplant.

We are reporting here a case of seven-year-old female child presenting with recurrent chest infections and congestive heart failure since early childhood.

Keywords: Restrictive cardiomyopathy; Restrictive cardiomyopathy of infiltrative variety; Pediatric restrictive cardiomyopathy; Diastolic heart failure; Dilated pulmonary artery

1. Introduction

Pediatric RCM is very rare, ranging from 2.5% to 3% of all pediatric cardiomyopathies and has a very poor prognosis with a mortality rate of 63% and 75% within 3 and 6 years of diagnosis, respectively [1, 2]. Genetic mutations involving sarcomeric and desmin genes have been described [3].

2. Restrictive Cardiomyopathy

2.1. Definition

RCM is the presence of abnormal compliance without another predominant phenotype of RV or LV dilation, hypertrophy, or systolic dysfunction. In some cases, mild hypertrophy or mild systolic dysfunction coexists with RCM [4].

RCM is the least common type of the cardiomyopathies, accompanied by structurally and functionally abnormal heart musculature in the absence of coronary artery disease, arterial systemic hypertension, valvular disease, or congenital heart disease (Figure 1) [4].

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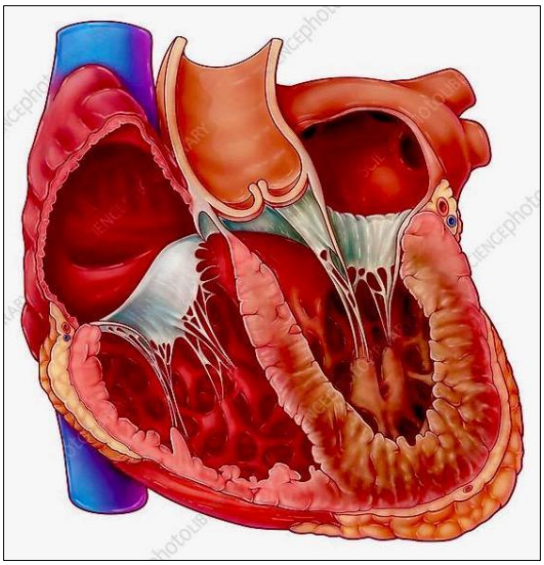


Figure 1 An illustration of restrictive cardiomyopathy, a type of cardiomyopathy in which normal heart muscle is replaced by abnormal tissue, causing the ventricles of the heart become stiff and rigid. When the ventricles are unable to normally relax and fill with blood, the atria become enlarged and blood flow in the heart is restricted.

2.2. Pathophysiology

RCM is characterized by restrictive ventricular physiology in the presence of normal or reduced diastolic volumes, with normal or near-normal left ventricular (LV) systolic function, and normal or near-normal wall thickness [1-5]. There is impaired myocardial compliance, which is caused by abnormalities in the myocyte or the inter-cellular matrix such as interstital infiltration or fibrosis [4]. In RCM, the impaired ventricular compliance is caused by dysfunction of the active relaxation of the ventricle [4].

The abnormal compliance of ventricles in RCM increases end-diastolic filling pressure, which is transmitted to the atria during diastole [4]. Because the atria are thin walled and distensible, the notable result is marked atrial enlargement. Although mitral and tricuspid regurgitation can develop in patients with advanced RCM, the atrial enlargement in RCM is not caused by atrioventricular regurgitation or by inlet obstruction [4] (Figure 2).

2.3. Restrictive Cardiomyopathy : Pathophysiology

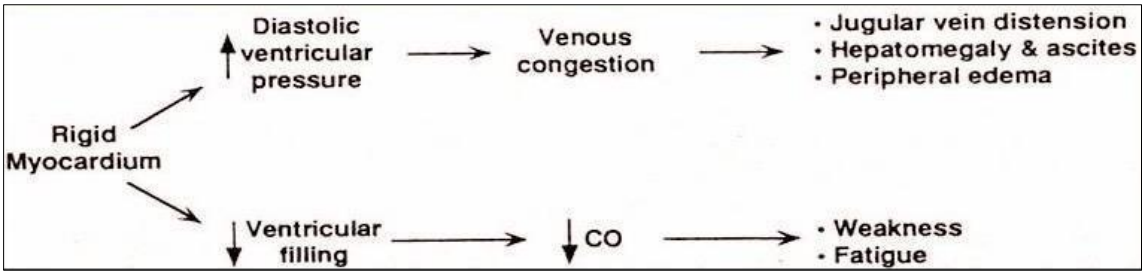


Figure 2 Pathogenesis of restrictive cardiomyopathy**.**

The functional phenotype includes a variety of clinical presentations, from asymptomatic to overt right or left HF, syncope, arrhythmias, thromboembolic complications, and sudden death [4]. Conduction abnormalities and atrial or ventricular arrhythmias can also be the first feature of RCM [4]. Pulmonary hypertension may be mild and reversible or severe and irreversible at presentation.

In the secondary infiltrative variety of RCM, echocardiography may identity thickened or speckled myocardium, endocardial fibrosis and intracardiac thrombi [4].

Therapy is mostly symptomatic; surgical options are limited to heart transplantation. Enzyme replacement therapy can be beneficial if specific enzyme deficiency can be demonstrated in some forms of storage disorders [5].

2.4. Etiology

RCM constitutes a heterogeneous group of heart muscle diseases with various causes (Table 1).

Table 1 Causes of RCM [4]

Primary RCM
Genetic
Sarcomeric
Desmin
Filamin-C
Secondary RCM
Infiltrative
Amyloidosis
Lysosomal storage disorder
Anderson-Fabry disease
Iron overload
Endomyocardial fibrosis
Parasitic infection
Autoimmune disorders
Malignancy with hypereosinophilia
Possible dietary deficiencies or ingested toxins in certain areas of endemic disease
RCM indicates restrictive cardiomyopathy.

2.5. Imaging modalities for restrictive cardiomyopathy

Multiple imaging modalities are currently available for a precise diagnosis of RCM:

- Transthoracic echocardiography
- 2 Dimensional strain echocardiography
- Cardiac magnetic resonance imaging
- Cardiac nuclear imaging
- Cardiac CT

Although non-invasive techniques are sufficient in most cases, final histologic diagnosis may sometimes be necessary, and may be obtained by endomyocardial biopsies (EMB) specimens from the heart and other organs.

2.4.1. Transthoracic Echocardiography [6, 7]

Diastolic dysfunction may be observed with spectral Doppler interrogation of the transmitral filling velocities and tissue Doppler of the mitral annular velocity (Table 2, Figure 3). The presence of restrictive physiology can be supported with the following:

- transmitral flow velocities
- amplitude of mitral E wave >100 cm/s
- deceleration time (DT) <160 ms
- ratio between E/A >2
- tissue Doppler imaging (TDI) of the mitral annulus

Characteristic, and pathognomonic, of restrictive filling physiology is the marked decrease in TDI velocities coupled with brisk reversal of hepatic venous Doppler flow upon inspiration

- an early diastolic (e') septal annular velocity of < 8 cm/s
- lateral annular e' reduced when <10 cm/s
- average E/e' ratio above 13 (using mean of septal and lateral e')
- correlates with elevated left atrial pressure

Pulmonary venous Doppler may be used as an adjunctive measure in equivocal cases, which should demonstrate:

- reversal of normally systolic dominant filling pattern, with an S/D ratio < 1
- elevation in peak atrial reversal (AR) velocities
- above 35 cm/s considered elevated

Table 2 Echocardiographic characteristics of restrictive cardiomyopathy [6]

Normal or mildly reduced EF
Normal or slightly increased LV wall thickness
Normal or slightly decreased LV cavity
Biatrial enlargement
Diastolic dysfunction Increased E/A ratio Short E wave deceleration time Decreased mitral annuluse' velocity Increased E/e' ratio Hepatic vein flow reversal with inspiration

EF: ejection fraction; LV: left ventricular.

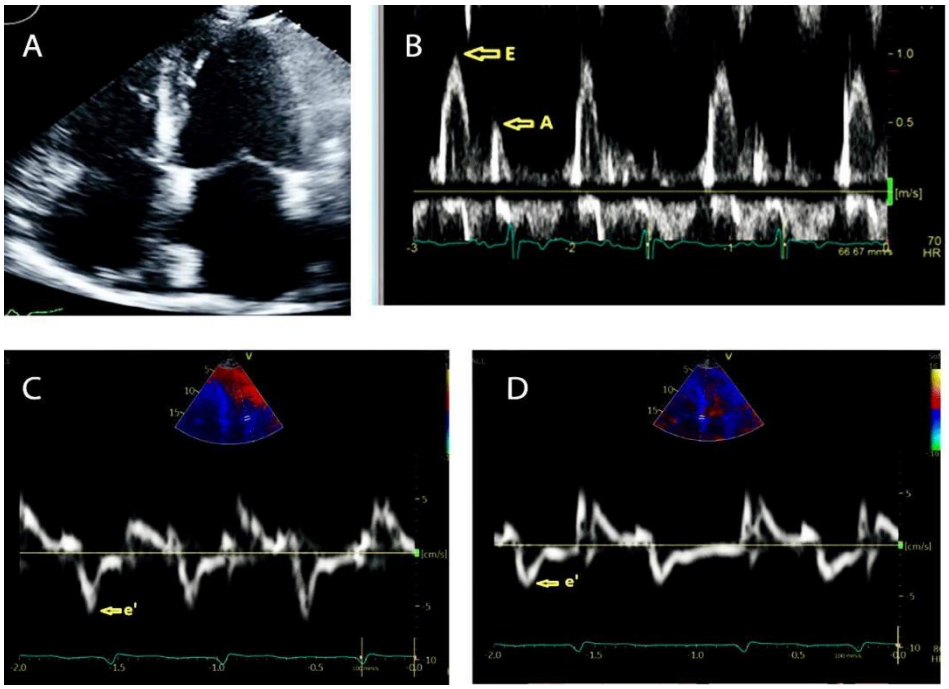


Figure 3 Transthoracic echocardiography. (A) Apical 4-chamber view showing biatrial enlargement. (B) Mitral inflow doppler showing increased E/A ratio (feature of diastolic dysfunction). (C, D) Lateral and septal mitral annular tissue doppler showing decreased e' velocity.

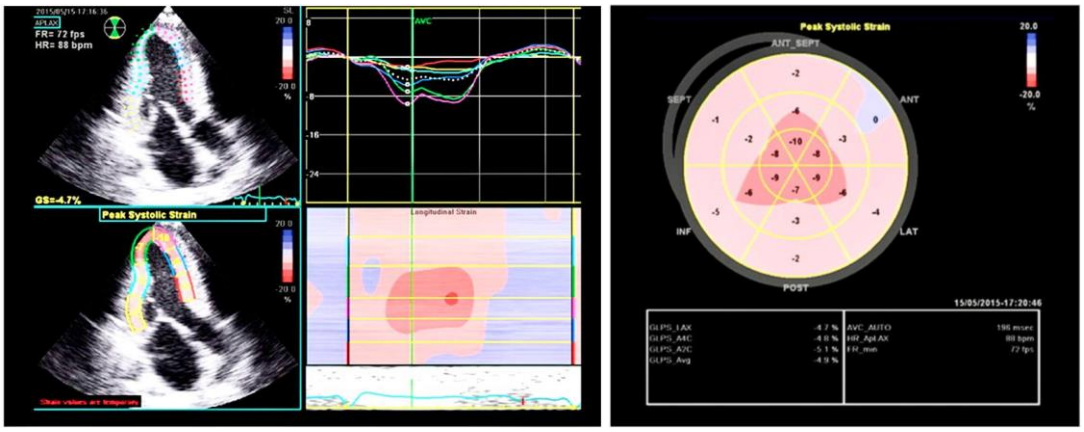


Figure 4 2-Dimensional strain echocardiography in secondary restrictive cardiomyopathy. (A) Two-dimensional-STE apical longitudinal view in systemic amyloidosis: severely abnormal longitudinal strain, particularly in the basal and medial LV segments. (B) Systemic AL amyloidosis, multiple myeloma: 2D-STE, relative apical sparing, typical of cardiac amyloidosis. Note the abnormal GLS (-4.9%).

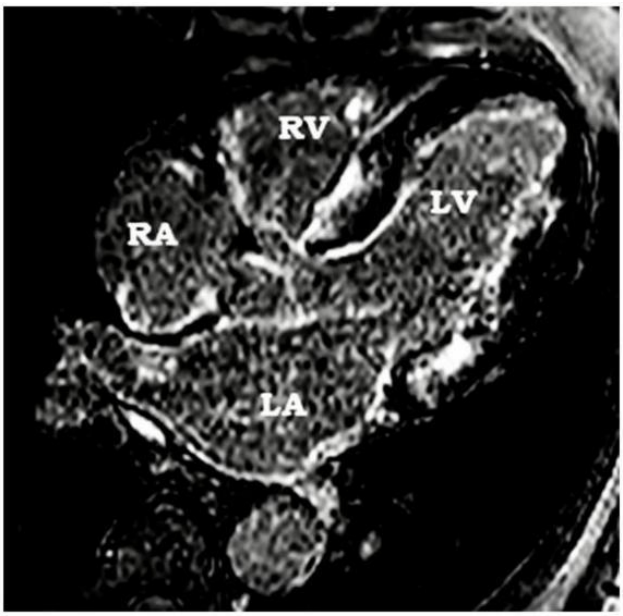


Figure 5 Cardiac magnetic resonance imaging. Cine CMR showed global LV hypertrophy, impaired longitudinal LV shortening, and dilated atria. Late gadolinium enhanced CMR in the figure showed diffuse endocardial enhancement consistent with infiltrative disease. LV, left ventricle; RV, right ventricle; LA, left atrium; RA, right atrium.

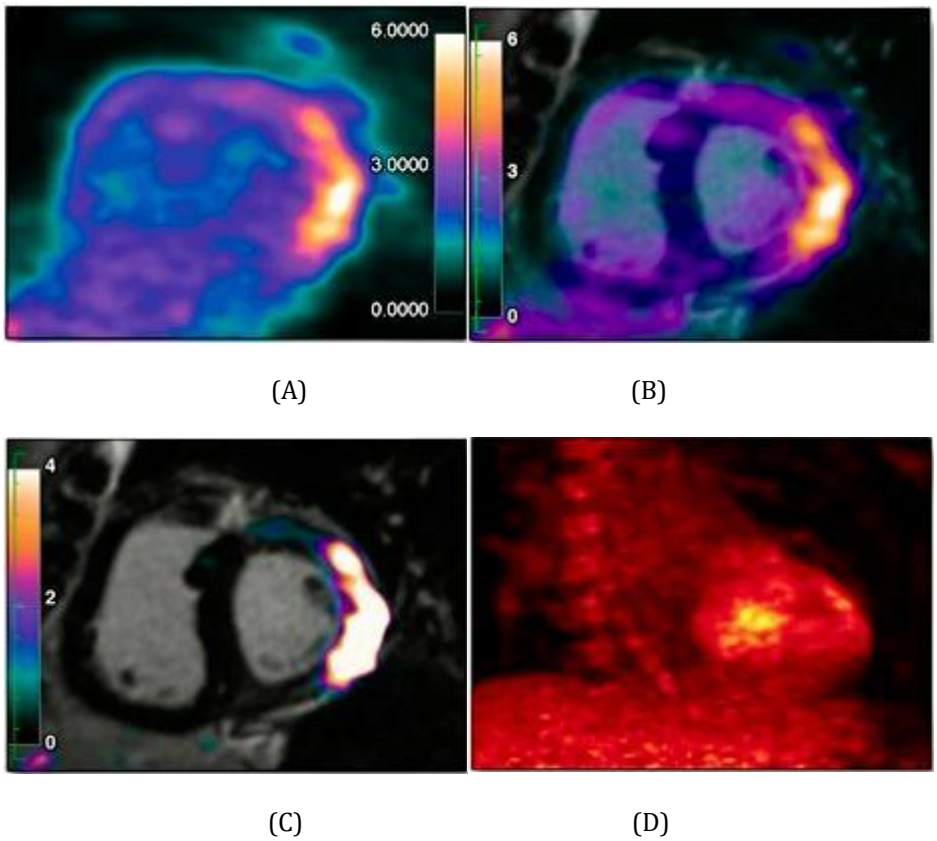


Figure 6 Nuclear Imaging by FDG-PET. (A) Matched FDG-PET and (B and C) fused FDG-PET/MR images obtained in short axis view showed intense uptake in exactly the same territory as the pattern of injury on cardiac MR, (D) Maximum intensity projection FDG-PET cone view confirmed abnormal myocardial uptake without evidence of increased activity outside of the heart.

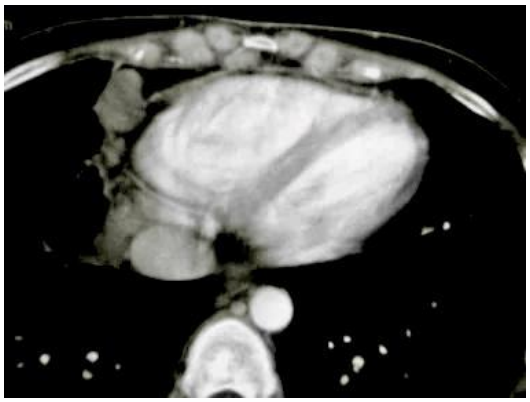


Figure 7 Cardiac CT for evaluation of restrictive cardiomyopathy. Axial contrast-enhanced CT image through the heart (same patient as in previous image) shows a thin pericardium without calcification. Note the cardiophrenic and internal mammary lymph nodes. The patient had extensive mediastinal and hilar adenopathy, as well as interstitial lung changes.

RCM is a challenging diagnostic problem for clinicians because it can be difficult to differentiate from constrictive pericarditis (CP). Cardiac catheterization can help identify the typical hemodynamic responses of RCM and CP [9, 10]. Here, we report a rare case of pediatric RCM that was diagnosed by comprehensive transthoracic echocardiography.

3. Case Report

A seven year old female child was referred to us for clinical cardiac evaluation and transthoracic echocardiography (TTE). As per the history narrated by the parents the child was full term normal delivery born out of non-consanguineous marriage. There was no history of maternal risk factors of CHD (obesity, diabetes, febrile illness, smoking, alcohol intake, teratogenic drug use, or radiation exposure). They informed that the child was suffering from recurrent chest infections accompanied with breathlessness and failure to thrive, since early childhood. However, they denied any history of loss of consciousness or cyanosis.

On clinical examination, the patient was thin built, sick looking and breathless (Figure 8).

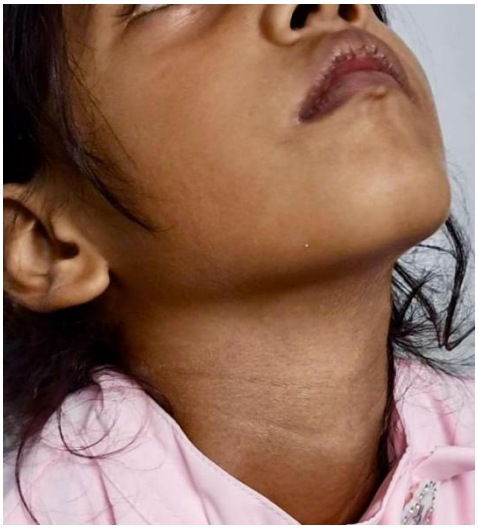


Figure 8 Facial appearance of the child : thin facies, sick looking and presence of jugular venous engorgement at > 60 degree chest inclination.

The infant's weight was 22 kg, height was 100 cm, pulse rate was 105/min, blood pressure was 90/60 mmHg, respiratory rate was 25/min and SPO2 was 98 % at room air. All the peripheral pulses were normally palpable without any radio-femoral delay.

On cardiovascular examination, there was presence of grade 3/4 left parasternal heave, palpable IInd heart sound, grade 3/6 mid systolic murmur in the pulmonary area, closely split loud IInd heart sound and right ventricular S3 gallop was heard in the tricuspid area (these clinical finding are consistent with significant pulmonary hypertension). First heart sound was normal. No clicks were heard.

On systemic examination there was florid signs of congestive heart failure:

- JVP was raised
- Liver was 5 cm enlarged in the right hypochondrium, firm and tender
- Bilateral interscapular and basal crepitations.

Xray chest (PA) view (Figure 9) demonstrated marked cardiomegaly with pulmonary venous congestion. Pulmonary blood flow was normal.

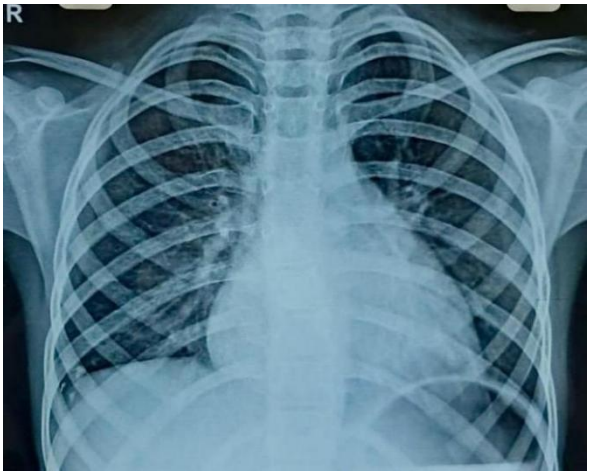


Figure 9 Xray chest (PA) displaying marked cardiomegaly with pulmonary venous congestion**.**

Resting ECG (Figure 10) exhibited :

- sinus tachycardia (ventricular rate 105/min)
- peaked P wave in L_2 L_3 AVF, V_2 - V_6, suggestive of right atrial enlargement
- P mitrale in L, AVL alongwith increased P terminal force in V_1, indicating left atrial enlargement
- left ventricular hypertrophy with strain
- QRS axis +100°

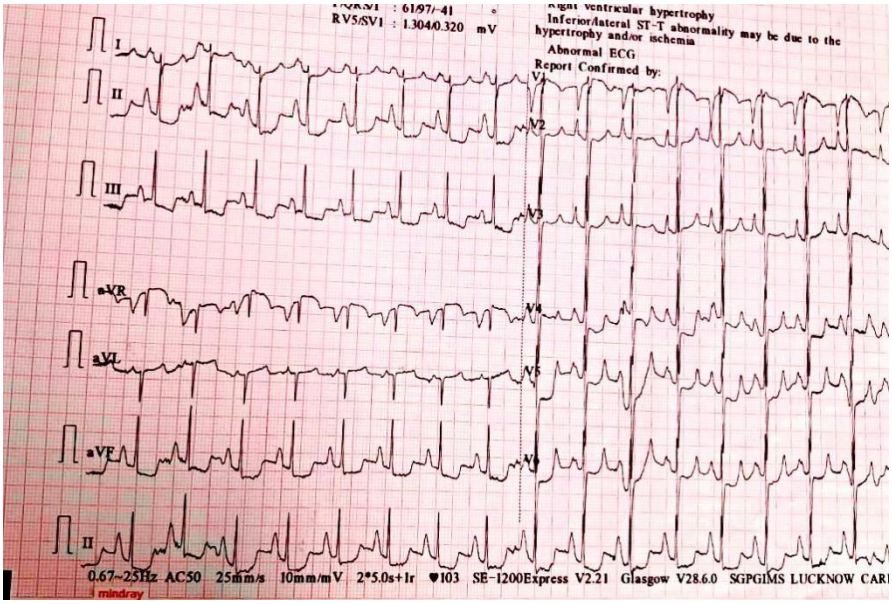


Figure 10 Resting ECG revealed sinus tachycardia biatrial enlargement, left ventricular hypertrophy with strain and QRS axis of +100°.

Transthoracic Echocardiography

All echocardiography evaluations were performed by the author, using My Lab X7 4D XStrain echocardiography machine, Esaote, Italy. The images were acquired using an adult probe equipped with harmonic variable frequency electronic single crystal array transducer while the subject was lying in supine and left lateral decubitus positions.

Conventional M-mode, two-dimensional and pulse wave doppler (PWD) and continuous wave doppler (CWD) echocardiography was performed in the classical subcostal, parasternal long axis (LX), parasternal short axis (SX), 4-

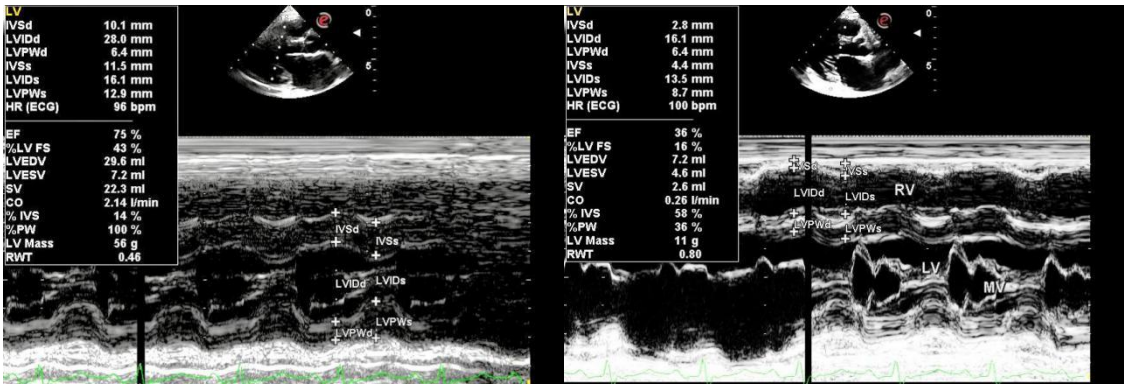
Chamber (4CH), 5-Chamber (5CH) and suprasternal views. Contemporary sequential segmental approach for echocardiographic analysis of our index patient was accomplished and the characteristic features were outlined (Figures 11-23).

3.1. M-mode Echocardiography

M-mode echocardiography of right and left ventricle was performed and the estimated measurements are outlined (Table 3, Figure 11).

Table 3 Calculations of M-mode echocardiography

Variables	LV	RV
IVS d	10.1 mm	2.8 mm
LVID d	28.0 mm	16.1 mm
IVS s	11.5 mm	4.4 mm
LVID s	16.1 mm	13.5 mm
LVPW d	6.4 mm	6.4 mm
LVPW s	12.9 mm	8.7 mm
EF	75 %	36 %
%LVFS	43 %	16 %
LVEDV	29.6 ml	7.2 ml
LVESV	7.2 ml	4.6 ml
SV	22.3 ml	2.6 ml
LV Mass	56 g	11 g



(A) (B)

Figure 11 M-mode echocardiography. Estimation of LV and RV volumetric data; IVS, interventricular septum; LVID, left ventricular internal dimension; LVPW, left ventricular posterior wall; d, diastole; s, systole; (substitute RVFW and RVPW for IVS and LVPW, respectively, in RV M-mode estimation of RV volumetric data); RVFW, right ventricular free wall.

3.2. Summary of M-mode echocardiography

- The ventricular cavity dimensions were small of both LV and RV.
- There was presence of concentric hypertrophy of LV and RV. LV mass was 56g and RV mass was 11g.
- LVEF was normal - 75 %; conversely RVEF was severely reduced - 36 %.

2Dimensional color echocardiography

Transthoracic color echocardiography exhibited multiple features as outlined below:

- Levocardia
- Situs solitus
- A-V concordance
- V-A concordance
- Concordant D-bulboventricular loop
- Normally related great arteries (NRGA)
- Left aortic arch
- Normal pulmonary and systolic venous drainage

In the apical 4CH and parasternal views, conspicuous hypertrophy of LV and RV was elucidated. The cavity sizes were small. On the biplane simpson's protocol LVEF was 58 %.

In the subcostal view, hepatic congestion alongwith dilated IVC was present indicating the presence of congestive heart failure.

Diastolic dysfunction assessment

Mitral inflow velocity pattern

- Tall E small A
- Mitral E wave velocity = 0.52 m/sec
- Mitral A wave velocity = 0.23 m/sec
- E/A ratio = 2.2:1
- E wave deceleration time = 52 ms
- E wave pressure half time = 30 ms

Tissue doppler imaging of mitral annulus

- Lateral E' velocity = 17 cm/sec
- E/E' ratio = 3.0:1

LV strain echocardiography

- GLS was significantly reduced (-8.6%)

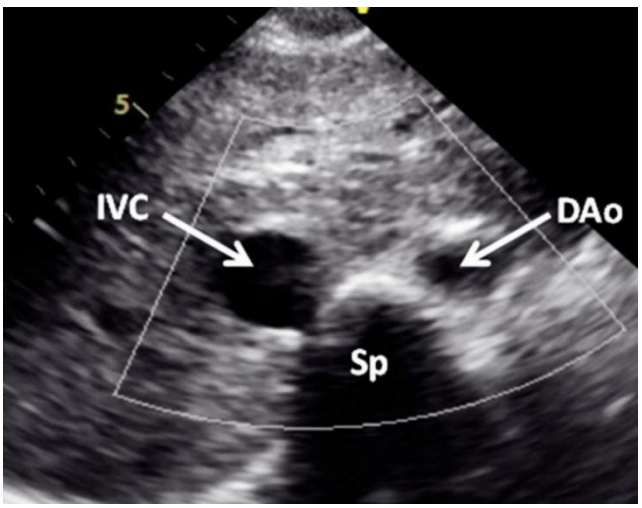


Figure 12 Visceral situs solitus. In the subcostal view the spine (Sp)was seen in the centre, descending aorta (DAo) on the left of spine and inferior vena cava (IVC) on the right of spine.

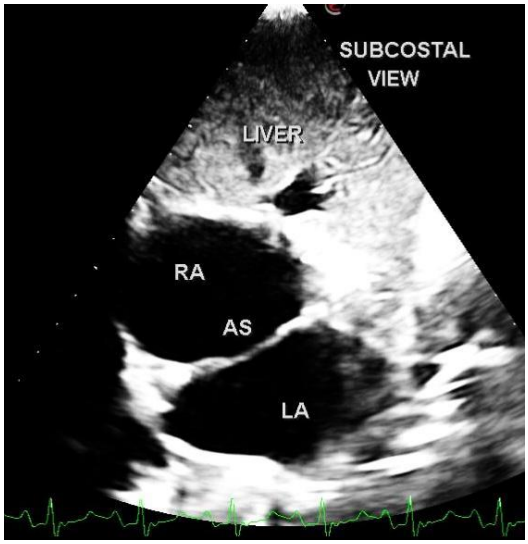


Figure 13 Atrial situs solitus. In the subcostal view the RA is lying to the right of LA.

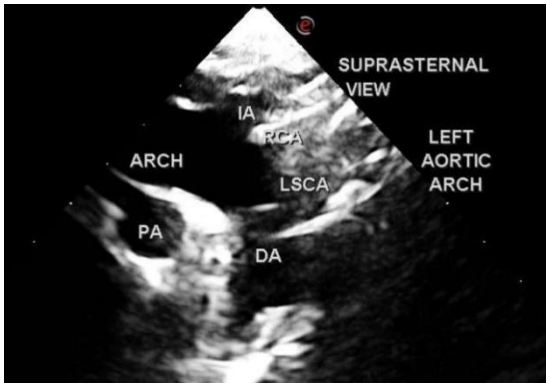


Figure 14 Suprasternal view - Left aortic arch.

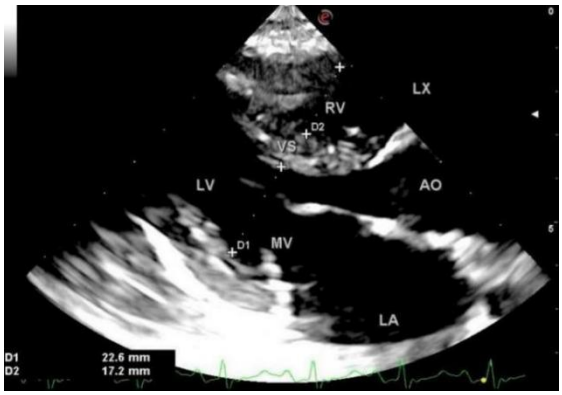
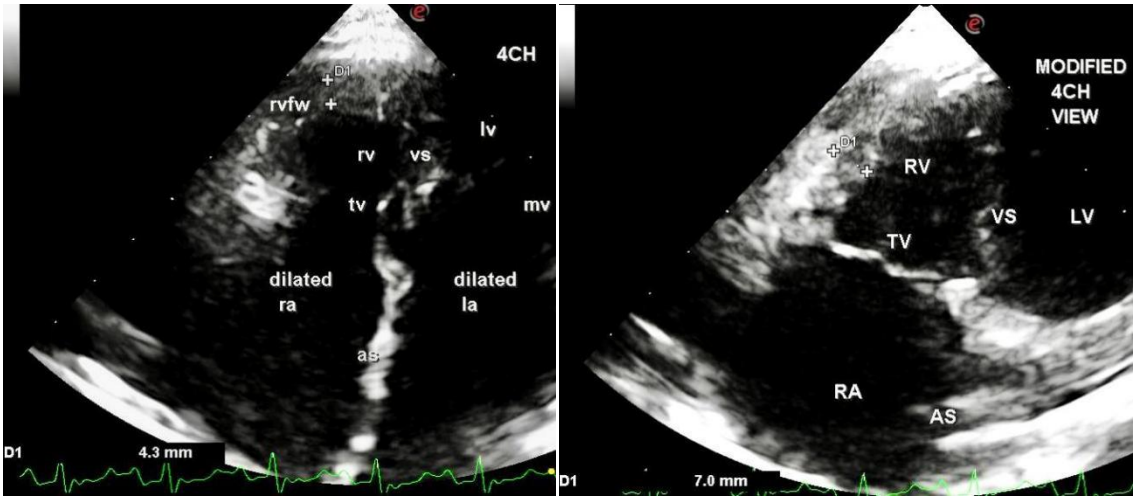
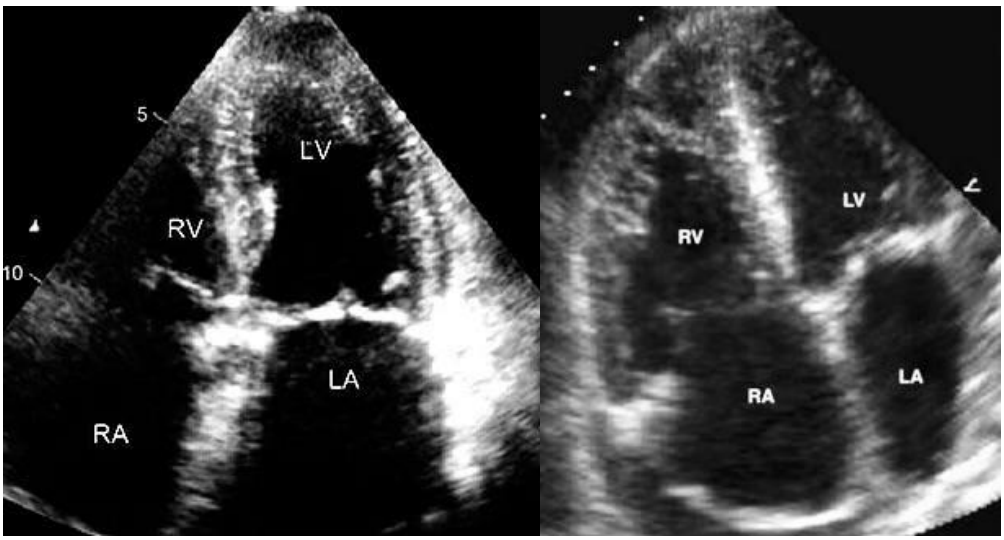


Figure 15 Parasternal LX view. There is biventricular concentric hypertrophy with smaller biventricular internal dimensions (diastolic left ventricular internal dimension was 22.6 mm and right ventricular diastolic internal dimension was 17.2 mm).



(A) (B)

Figure 16 Apical 4CH view displaying RV concentric hypertrophy. (A) RV free wall thickness was 4.3 mm in diastole and (B) 7.9 mm in systole.



(A) (B)

Figure 17 Apical 4CH view (A and B) illustrates the characteristic features of restrictive cardiomyopathy. Conspicuous dilatation of right and left atrium, smaller sized left and right ventricles, concentric hypertrophy of LV and RV.

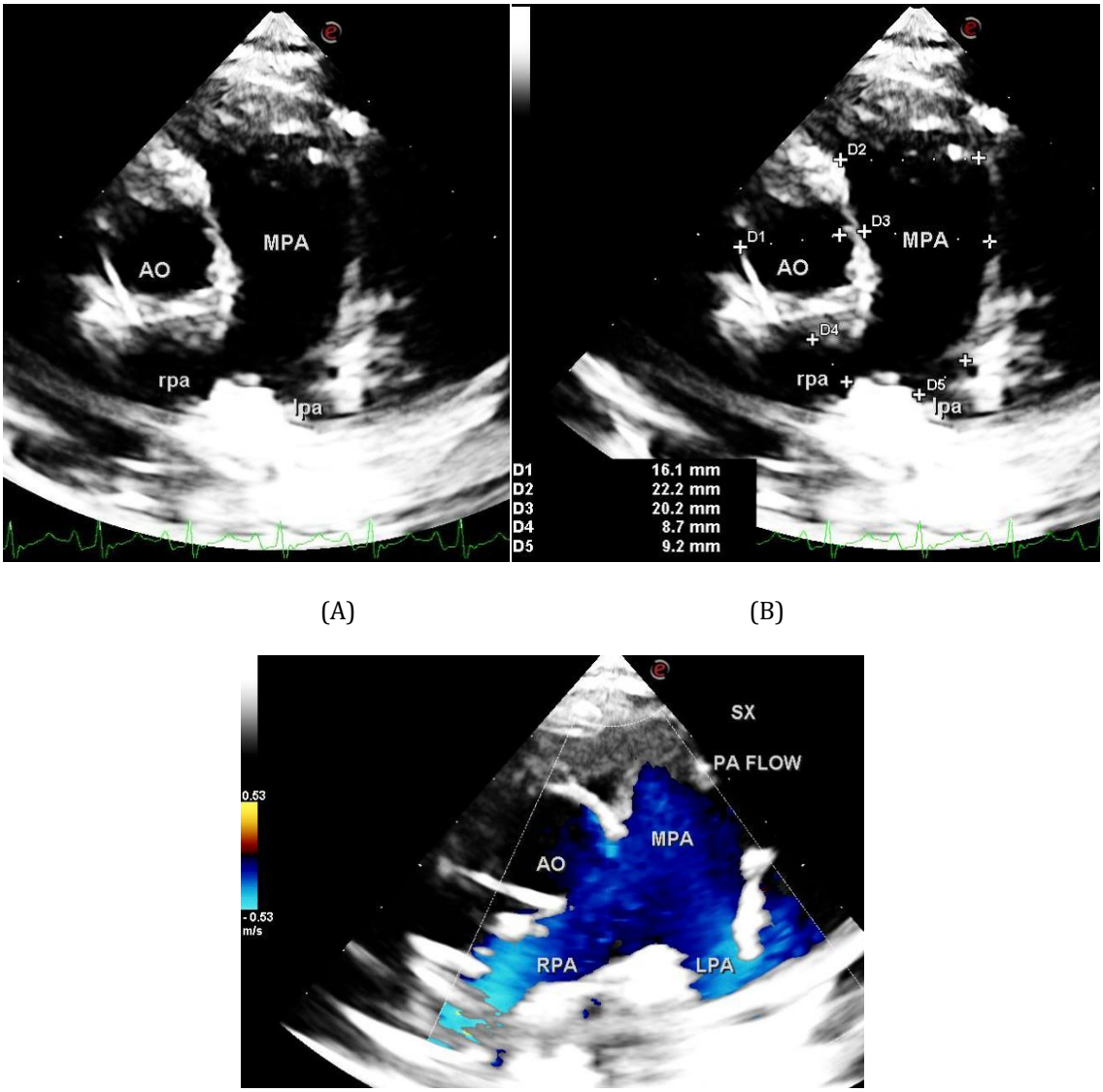


Figure 18 SX view showing dilated pulmonary arteries. (A) The pulmonary valve annulus and main pulmonary artery are markedly dilated; (B) The dimensions of dilated pv annulus, main pulmonary artery, left pulmonary artery, right pulmonary artery and aorta were estimated; (C) On color flow mapping of the pulmonary artery, characteristic dilatation of pulmonary arteries were portrayed.

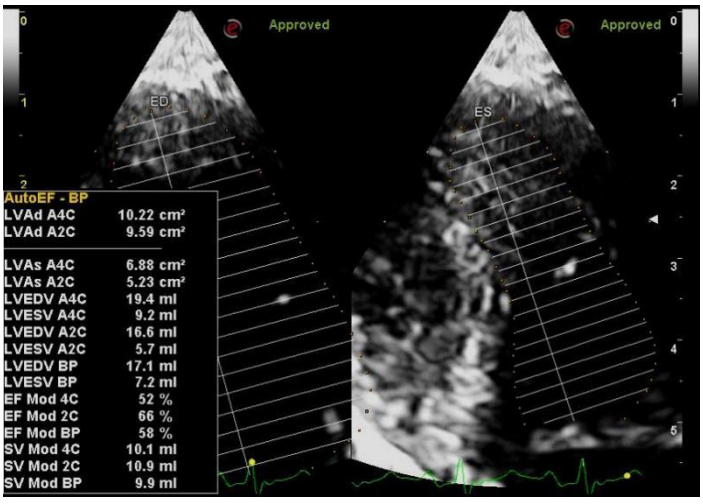


Figure 19 Biplane Simpson's method for estimation of LVEF. LVEF was 58 %.

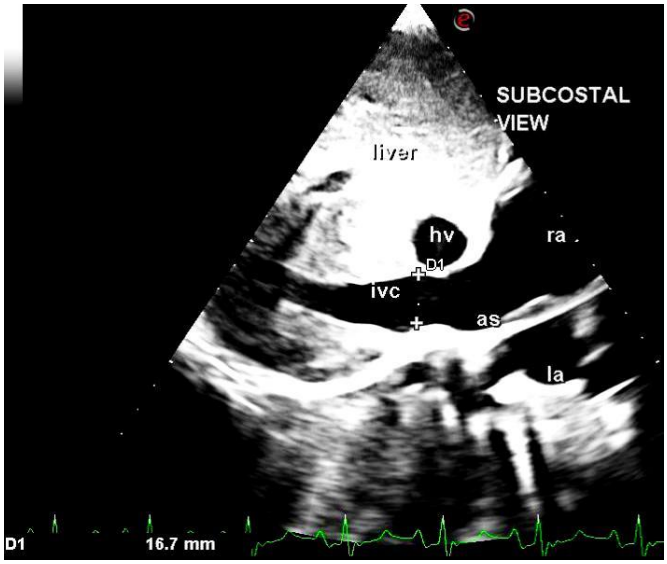
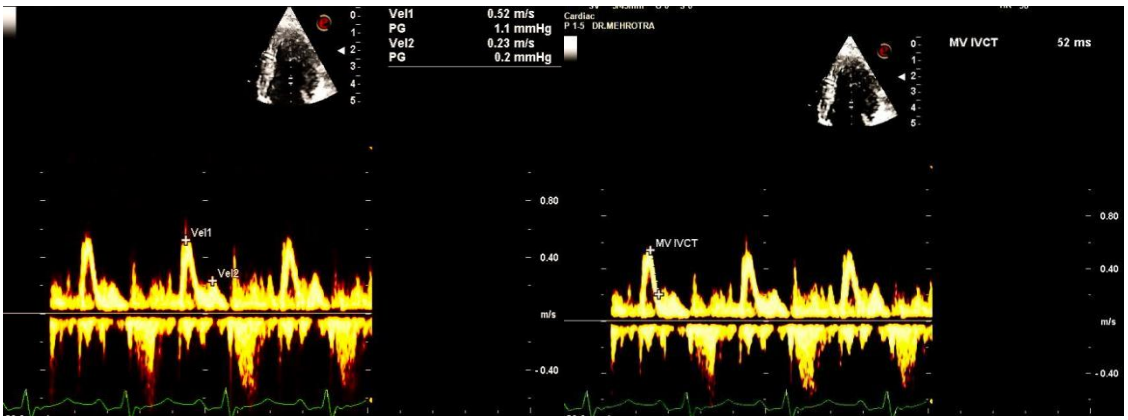
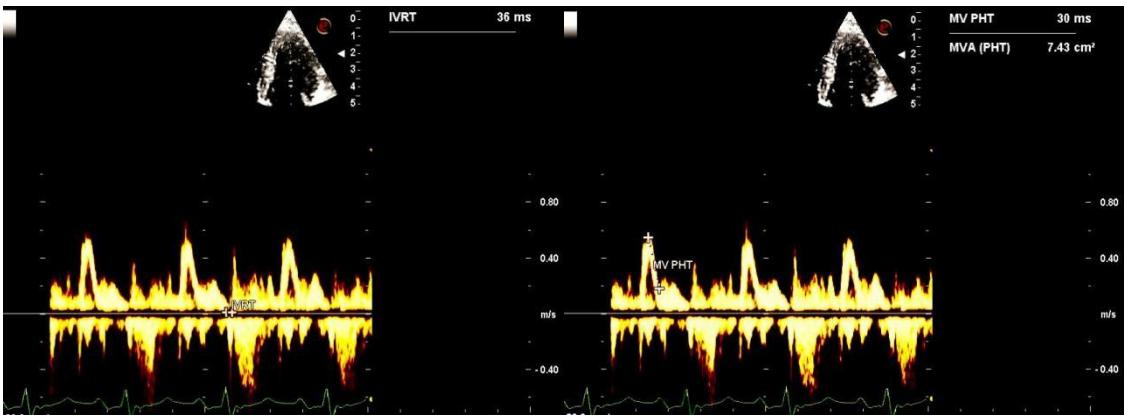


Figure 20 Subcostal view shows marked hepatic congestion accompanied by dilatation of IVC and hepatic vein. ivc dimension was 16.7 mm; ivc, inferior vena cava; hv ,hepatic vein ; as, atrial septum; ra, right atrium; la, left atrium.

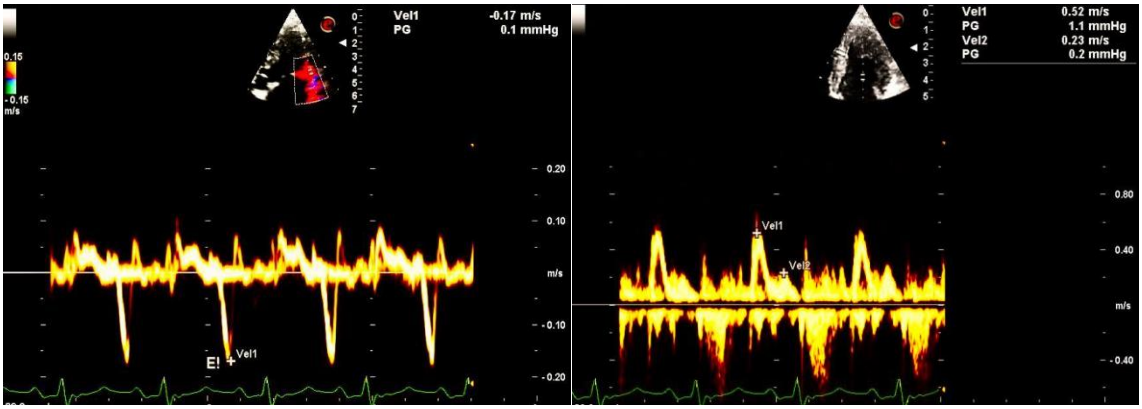


(A) (B)



(C) (D)

Figure 21 Mitral inflow velocity pattern of diastolic dysfunction. (A), E wave velocity was 0.52 m/sec and A wave velocity was 0.23 m/sec; (B), MV deceleration time was 52 ms;(C), Isovolumic relaxation time was 36 ms; (D), MV pressure half time was 30 ms.



(A) **(B)**

Figure 22 Tissue Doppler Imaging. (A) Lateral E' was 17 cm/sec and (B) Mitral E was 52 cm; E/E' ratio was 3.0:1.

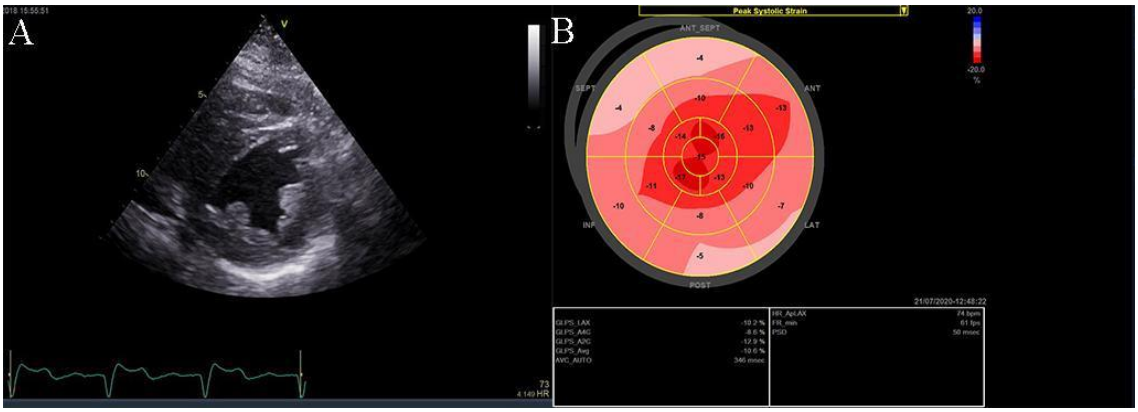


Figure 23 LV strain echocardiography. (A) Short axis view showing left ventricular hypertrophy; (B) LV speckle tracking echocardiography in the apical views reveals a significantly reduced GLS (global longitudinal strain) of - 8.6 %.

- Summary of 2Dimensional echocardiography
- On summing up, we have demonstrated numerous features of restrictive cardiomyopathy in our index patient:
- Dilated left and right atrium
- Smaller sized left and right ventricles
- Mild thickness of left and right ventricular walls
- Markedly reduced MV deceleration time, pressure half time, pressure half time, IVRT, E and A wave velocities alongwith substantially increased MV E/A ratio to 3.0:1.
- Decreased E' velocity - 17 cm/sec (normal: > 20 cm in children [11])
- LV strain illustrated markedly reduced GLS to - 8.6 %
- Dilated main & branch pulmonary arteries
- Normal LVEF with severely reduced RVEF 36 %

Dilated main & branch pulmonary arteries, without any RV inflow or outflow obstruction and in the presence of severely reduced RVEF, is suggestive of severe pulmonary hypertension.
Presence of hepatic congestion alongwith dilated IVC and hepatic vein indicates congestive heart failure.

For a definitive determination of primary (idiopathic) vs secondary restrictive cardiomyopathy, endomyocardial biopsy is necessary. However, the parents refused to give their consent for the procedure.

3.3. Final Echocardiographic diagnosis

- Pediatric restrictive cardiomyopathy
- Severe pulmonary hypertension

- Congestive heart failure

4. Discussion

In RCM, the usual mode of inheritance is autosomal dominant while autosomal recessive, X-linked, and mitochondrial patterns of inheritance are not uncommon [12]. Etiology can be idiopathic, genetic, inborn error of metabolism, or systemic infiltrative disease. Mechanism of restrictive phenotype is varied ranging from myocardial fibrosis and hypertrophy to infiltration of the myocardium (like amyloid, iron overload, storage disorders, and endomyocardial fibrosis) [12].

4.1. Restrictive Cardiomyopathy - classification

Several classification have been put forward by many authors. However, there are two effective classifications which are prevalently employed in clinical practice. The classification of Ditaranto et al [13] is shown in Figure 24.

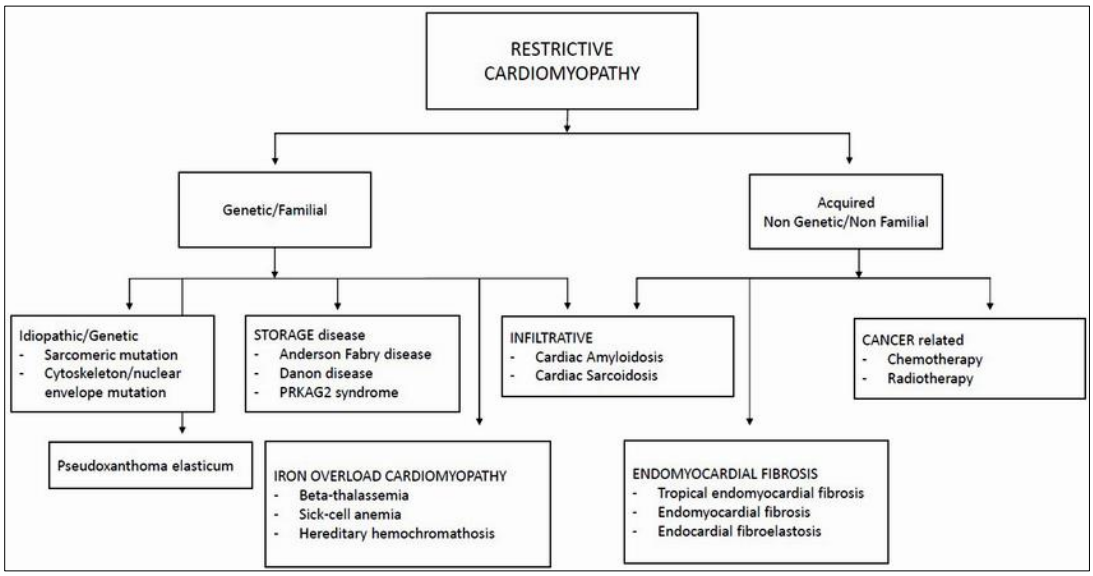


Figure 24 Classification of restrictive cardiomyopathy [13].

Pereira et al [14] proposed a simple classification of RCM (Figure 25).

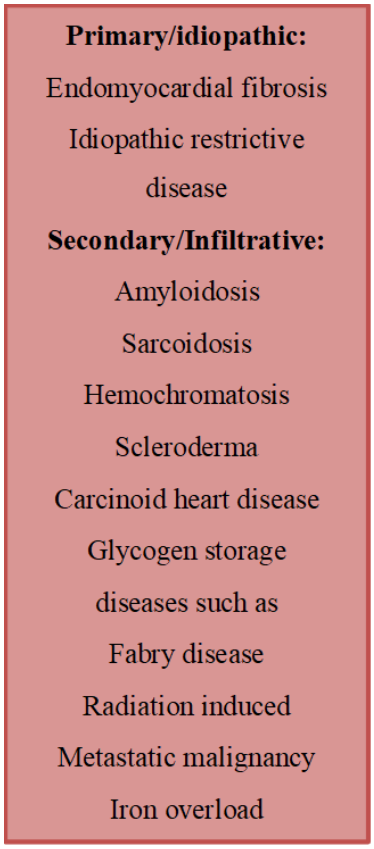


Figure 25 Classification of restrictive cardiomyopathy [14]

The differentiation of constrictive pericarditis from RCM is challenging because the two condition share a common clinical manifestation, and similar echocardiographic findings. However, the treatment options are different, particularly pericardiectomy can effectively cure constrictive pericarditis. Therefore this differentiation is crucial [13, 14] (Table 4).

Table 4 Differential diagnosis of constrictive pericarditis and restrictive cardiomyopathy [13, 14].

	Constrictive pericarditis	**Restrictive cardiomyopathy**
Clinical examination	Kussmaul's sign, usually present	Kussmaul's sign, may be present
	Pulsus paradoxus (may be present)	Pulsus paradoxus (infrequent)
	Pericardial knock	S3; Systolic murmur due to mitral and tricuspidal regurgitaion
Biomarkers		
NT-proBNP	Normal or slightly abnormal	Abnormal
2-Dimensional echocardiography		
LV ejection fraction	Normal	Normal or slightly decreased
Pericardium appearance	Thickened/bright	Normal
Interventricular septum movement	Abnormal	Normal
Interventricular septum position	Varies with respiration	Normal

Doppler echocardiography		
E/a ratio	Increased (≥ 2)	Increased (≥ 2)
E/a ratio response to Valsalva maneuver	Variation > 25%	Minimal variation
E-wave decelerating time, ms	Decreased (≤160)	Decreased (≤160)
E' septal, cm/s	≥ 8	< 8
E' septal/lateral	Septal > lateral	Lateral > septal
S', cm/s	> 5	< 5
Mitral valve inflow propagation, cm/s	Normal or increased (≥ 100)	< 55
Hepatic vein flow	Diastolic flow reversion during expiration	Diastolic flow reversion during inspiration
2-Dimensional speckle tracking echocardiography		
Circumferential strain	Decreased	Normal
Radial strain		
Basal	Decreased	Normal
Apical	Normal	Normal
Longitudinal strain		
Septal	Normal	Decreased
Lateral	Decreased	Decreased
Basal segments	Normal	Decreased
Apical segments	Decreased	Decreased
Global	Decreased	Decreased
Twist motion	Decreased	Normal
Apical	Decreased	Normal
Left atrial reservoir strain		
Lateral	Decreased	Decreased
Septal	Increased	Decreased
CT scan/MRI		
Pericardial thickening	Present	Absent
Cardiac catheterization		
Right and left ventricular end-diastolic pressures comparison	Equal or ≤ 5 mm Hg	Usually left > right
Dip-plateau waveform	Typically present	Can be present
Doppler values are based on adult normative data, as discussed in the text. LV indicates left ventricle; and RCM, restrictive cardiomyopathy.		

Restrictive cardiomyopathy (RCM) is a heart muscle disease that leads to diastolic dysfunction with preserved systolic function in the early stages. The cardiac muscle is affected by abnormal stiffness affecting the ventricle; this leads to increased diastolic pressure in the atrium, causing hypertrophy [15]. RCM represents approximately 2.5-5% of all cases of cardiomyopathies in children. RCM is the least common of three original subtypes of cardiomyopathies: hypertrophic, dilated, and restrictive. Possible infiltrative diseases have been identified in adults (Amyloidosis, Sarcoidosis, Hereditary Haemochromatosis, and Fabry's disease) [15].

There are no defined diagnostic criteria for RCM in children; however, the left atrium size is often used as a reference for diagnosis. Some guidelines recommend measuring the mitral valve E/A ratio, which is > 2 in advanced diastolic dysfunction stages.

Few echocardiography characteristics have been recognised to differentiate primary from secondary restrictive cardiomyopathy [9, 10, 13, 14] (Table 5).

Table 5 Echocardiographic features distinguishing primary from secondary restrictive cardiomyopathy

Variables	**Primary**	**Secondary**
Wall thickness of LV and RV	Usually normal or mildly increased	Commonly increased
Ventricular cavity abnormalities	Absent	May be present
LV mass	Normal	May be increased
Pericardial thickness	Normal	May be mildly increased without calcification
Interatrial thickness	Normal	May be thickened
Additional features (thickened valves and pericardial effusion)	Not seen	May be present
Exclusively confined to heart muscles	Yes	Not confined (myocardial damage caused by systemic or multi-organ disease)

Cardiac MRI may be a diagnostic alternative to demonstrate the atrial cavity dilatation and increasing thickness of the ventricular myocardium, due to infiltrative diseases; therefore, it is usually more helpful in adults than children [14, 16, 17].

Treatment is aimed at secondary causes when present. In adults, the treatment objective is to reduce the protein production that affects the myocardium in cases where genetic disorders cause RCM and supportive measures until cardiac transplantation is possible. Loop diuretics and aldosterone inhibitors are preferred over other antihypertensive medications such as ACE inhibitors and ARBs. They are not well tolerated because of their hypotensive effect. Creatinine and electrolytes must be monitored during their use.

Additionally, β-blockers and calcium antagonists are not commonly indicated because they may reduce the heart rate affecting the optimal cardiac output for patients with normal heart rates. However, they can be used in some arrhythmic presentations. Digoxin's use is limited for tachyarrhythmias management because the preserved systolic function may lead to digitalis intoxication. Amiodarone may be considered for this function [14, 18]. RCM has a poor prognosis, the worst among all cardiomyopathies. Studies show that patient mortality after 2-3 years of diagnosis, without cardiac transplantation is 50 to 75% at five years [14, 17].

4.2. Diagnostic criteria

There are no established diagnostic criteria for RCM in the pediatric age. Therefore, diagnosis can be based on a thorough clinical and physical evaluation, a complete patient and family history, and various specialized investigations. In our case, the patient arrived for the first time at the cardiac center with critical cardiac symptoms such as progressive dyspnoea, cough, and palpitations. Chronic disease evidence was appreciated at first sight due to her sickly appearance. The parents did confirm a cardiac diagnosis at a young age. A complete cardiac study was under taken because of her symptoms and signs suggestive of congestive heart failure.

The patient's X-rays, combined with clinical findings, confirmed a global cardiac illness. The electrocardiogram revealed the presence of atrial hypertrophy, which led to echocardiography to determine atrial involvement and measure how much this impacted cardiac function. The echocardiogram reported a LA size of 30 mm, accompanied by severe compromise of both her left and right atria. The E/A ratio was 3.0:1, consistent with severe grade of restriction. Perhaps, a diagnosis of restrictive cardiomyopathy was adequate according to both clinical and echocardiographic data.

5. Conclusion

Restrictive cardiomyopathy usually presents as heart failure with preserved ejection fraction and has distinct echocardiographic and hemodynamic features. Given its varied etiology and clinical presentations, it can be a diagnostic challenge and requires a high degree of suspicion. Evaluation of patients with suspected RCM is guided by their history, presenting features, and clinical suspicion. Management involves treatment of the underlying condition in secondary RCM, symptomatic therapy for systemic and pulmonary congestion, and early initiation of heart failure therapies when needed.

Compliance with ethical standards

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Disclosure of conflict of Interest

There is no conflict of interest.

Statement of ethical approval

Ethical approval was obtained from the ethical review committee of our institution-Prakash Heart Station, Lucknow.

Statement of Informed Consent

Informed consent was procured from the parents of the child.

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