

# Isolated Non-Compacted Right Ventricular Myocardium with Severe Pulmonary Hypertension

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**Abstract—**BACKGROUND: There have been only at least seven cases of isolated right ventricular non-compaction reported in literature, hence, there are no definite criteria for diagnosis and recommendations for management of these patients. The reported clinical manifestations include heart failure, arrhythmias and cardioembolic events. Left ventricular and biventricular non-compaction are likewise rare but more common than isolated right ventricular non-compaction. Pulmonary hypertension is associated with biventricular non-compaction more commonly but not with isolated right ventricular non-compaction.

**SETTING:** University of Santo Tomas Hospital

**CASE REPORT:** We report a case of a 25 year old female who presented with progressive dyspnea for one year accompanied by easy fatigability. She denied chest pain, palpitations, pedal edema, paroxysmal nocturnal dyspnea and orthopnea. She had a brother who died at a young age reportedly due to a cardiac illness which was not fully worked-up. Her 2D echocardiogram showed excessive prominent trabeculations and deep inter-trabecular recesses in the right ventricular wall with depressed right ventricular systolic function by tricuspid annular plane systolic excursion and fractional area change. Color Doppler studies also showed severe tricuspid regurgitation, pulmonic regurgitation and severe pulmonary hypertension. Secondary causes of pulmonary hypertension like connective tissue diseases, left heart disease, chronic thrombotic/embolic disease, anomalous cardiac and pulmonary shunts and lung disease were excluded. She was given diltiazem, sildenafil, digoxin, warfarin overlapped with subcutaneous enoxaparin and oxygen supplementation which provided symptomatic relief.

**Index Terms—**Isolated ventricular non-compaction, Non-compacted right ventricular myocardium, Pulmonary hypertension, Ventricular Hypertrabeculation.

## I. CASE REPORT

A 25 year old female consulted due to progressive dyspnea for the past year. She denied chest pain, palpitations, pedal edema, paroxysmal nocturnal dyspnea and orthopnea. She has no known medical conditions prior to symptom onset. She delivered via normal spontaneous delivery with no peripartum complications to a baby boy four years before consultation. Family history revealed that she had a brother who died at a young age due to a cardiac illness which was not fully worked-up. His clinical presentation was not known to the patient. On examination, she was tachycardic with a regular pulse rate of 105 beats/minute, a blood pressure of 90/60 mmHg, and afebrile at 36.7°C. She had an elevated jugular venous pressure with a prominent ‘y’ descent. There were no malar rash, no oral ulcers and no skin rashes. Cardiovascular examination showed displacement of the left ventricular apex beat at the 5<sup>th</sup> left intercostal space anterior axillary line, presence of an RV heave and a PA lift. There was a grade 4/6 holosystolic murmur heard best at the 4<sup>th</sup> left intercostal space over the parasternal line and a 4/6 decrescendo murmur at the 2<sup>nd</sup> left intercostal space parasternal line. Respiratory system examination showed fine bibasilar crackles on both lower lung fields. There were no limitations in movement of her joints and no arthritis noted. Her pulses were full and equal. There were no pedal edema and no cyanosis noted.

Initial consideration at that time were congenital heart disease, r/o pulmonary embolism, right ventricular hypertrophy, sinus rhythm, tricuspid regurgitation, pulmonic regurgitation, pulmonary hypertension, in congestive heart failure, NYHA Class IV D.

Twelve lead electrocardiogram (Appendix 1: Figure 1) showed sinus tachycardia with right axis deviation and right ventricular hypertrophy. Chest X-ray (Appendix 1: Figures 2A

Manuscript submitted March 10, 2014.

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and 2B) showed cardiomegaly with prominent pulmonary vascular markings and prominent pulmonary arteries. Arterial blood gas showed partially compensated metabolic alkalosis with inadequate oxygenation at room air (Appendix 2: Table 1). Transthoracic 2D echocardiogram showed (Appendix 1: Figure 3.1-3.3) dilated and hypertrophied right ventricle with non-compaction of the right ventricular apex and mid segments with depressed systolic function with fractional shortening of 19% and tricuspid annular plane systolic excursion (TAPSE) of 1.4 cm. The left ventricle was normal in size with good wall motion and contractility and normal resting systolic function with grade 1 diastolic dysfunction. There was systolic and diastolic interventricular septal flattening suggestive of volume and pressure overload (Appendix 1: Figure 3). The right atrium was dilated with no evidence of thrombus. There was minimal pericardial effusion with no evidence of tamponade. She had severe tricuspid regurgitation, pulmonary regurgitation and severe pulmonary hypertension with a pulmonary arterial pressure of at least 70 mmHg by tricuspid regurgitant jet and 68 mmHg by tricuspid regurgitation regression formula.

Venous duplex scan of the lower and upper extremities (Appendix 3 and 4) were done to rule out venous thrombosis and showed normal results. Complete Blood count (Appendix 2: Table 4) showed leukocytosis with predominance of segmenters.

She was given furosemide 20 mg intravenously once followed by 20 mg orally once daily and anticoagulation with subcutaneous Enoxaparin 0.4 ml subcutaneously every 12 hours. Transesophageal echocardiogram (Appendix 1: Figure 4) did not demonstrate anomalous shunts and showed intact interatrial septum and interventricular septum. The right atrium was noted to be dilated with an echodensity noted attached to its walls suggestive of thrombus formation; the suspicious thrombus was seen to extend up to the proximal superior vena cava in the short axis view.

Secondary causes for pulmonary hypertension like connective tissue diseases, left heart disease, chronic thrombotic/embolic disease, anomalous cardiac and pulmonary shunts and lung disease were excluded. D-dimer, Protein C and Protein S activities, anti-cardiolipin Immunoglobulin antibodies and anti-nuclear antibodies (Appendix 2: Table 5) to rule out possible pulmonary embolism and connective tissue disease were done and showed negative results. She was likewise referred to Rheumatology specialist for evaluation of pulmonary hypertension and was assessed to have no connective tissue disease that warrants further examination. She was started on diltiazem 30 mg/tablet, 1 tablet q8 hours, sildenafil 50 mg/tablet, ¼ tablet OD, and digoxin 0.25 mg/tablet, 1 tablet OD. Warfarin 2.5 mg/tablet, 1 tablet OD was overlapped with subcutaneous enoxaparin 0.4 mL/ q12 hours, and she was given oxygen supplementation at 2-3 lpm/nasal cannula.

## II. DISCUSSION

Non-compaction of the ventricular myocardium is a rare type of cardiomyopathy with an estimated prevalence of 0.05% in adults and 0.14% in pediatric population and occurs more commonly in males with a 5.7 to 1.2 male to female ratio. <sup>[1]</sup> It is a cardiomyopathy thought to be caused by arrest of normal embryogenesis of the endocardium and myocardium. This abnormality is often associated with other congenital cardiac defects, but it is also seen in the absence of other cardiac anomalies. <sup>[2]</sup> The diagnosis of isolated ventricular non-compaction includes the exclusion of other cardiac anomalies other than hypertrabeculation of the ventricular myocardium with communication of the trabeculae into the ventricular lumen.

The World Health Organization/International Society and Federation of Cardiology Task Force reported in 1995 that noncompaction of ventricular myocardium is a rare cardiac malformation considered as an unclassified cardiomyopathy. <sup>[3, 4]</sup> In 2006, it has been classified by the American Heart Association as a genetic cardiomyopathy. <sup>[13, 14]</sup> In the same year, Espinola-Zavaleta et. al. published an echocardiographic study that identified 53 cases out of 125,438 echocardiograms studied to have non-compacted ventricular myocardium. Seventy four percent of the identified cases had isolated ventricular non-compaction while the rest had other accompanying congenital abnormalities. Two-thirds of all the identified cases had left ventricular involvement while the other third had biventricular involvement. None were identified to have isolated right ventricular non-compaction. Because of its rare occurrence, there is paucity of data regarding its diagnosis and management. Diagnostic criteria that were used in previous case reports followed those used for left ventricular non-compaction by Oechslin et. al. and Jenni et. al. which we also applied in our case.

Isolated ventricular non-compaction is a rare congenital cardiomyopathy with a high morbidity and mortality due to malignant arrhythmias and pump failure. Areas affected by non-compaction are characterized by increased trabecularization and deep inter-trabecular spaces. <sup>[5]</sup>

Isolated noncompaction of the ventricular myocardium, first described by Chin et. al. in 1990, is characterized by persistent embryonic myocardial morphology found in the absence of other cardiac anomalies to explain the abnormal development. In such cases, the resultant deep recesses communicate only with the ventricular cavity, not the coronary circulation. <sup>[2]</sup>

The diagnosis of noncompaction of the ventricular myocardium can be made by 2-dimensional and color doppler echocardiography. <sup>[2]</sup> The echocardiogram is in fact, considered the diagnostic procedure of choice, and diagnosis is based on established criteria. <sup>[6, 4]</sup>

Multiple prominent ventricular trabeculations with deep intertrabecular recesses are seen. Color Doppler imaging

demonstrates blood flow through these deep recesses in continuity with the ventricular cavity. Isolated ventricular myocardium is diagnosed when the above criteria are satisfied and coexisting cardiac lesions, such as semilunar valve obstruction and coronary artery anomalies, are excluded. [2] Our patient demonstrated presence of deep intertrabecular recesses with communication only with the ventricular cavity and most prominent in the apical segment of the right ventricular wall. The semilunar valves were normal and the interatrial and interventricular septa were intact.

The main diagnostic criterion of noncompaction, that is, the only one that is accepted and recognized, is evaluation of the ratio between the spongiosus and the compact thickness of the ventricular wall of  $>2$ . This ratio is easy to calculate for the left ventricle; on the contrary, it is more difficult to calculate for the right ventricle. [7] Our patient demonstrated a 4.33:1 non-compacted to compacted layer ratio of the right ventricular myocardium.

Isolated ventricular non-compaction can be accurately diagnosed by echocardiography, as seen by the agreement with necropsy findings. Although other modalities such as computer tomography, magnetic resonance imaging, and ultrafast computed tomography may also be helpful, no diagnostic criteria for these modalities have yet been proposed. [8]

The most commonly used echocardiographic criteria for the diagnosis of isolated ventricular non-compaction in adults follows the proposal by Oechslin et al.

Diagnostic Criteria for Isolated ventricular non-compaction by Oechslin et al:

1. Absence of coexisting cardiac abnormalities (other than 2-4 below) by definition.
2. Typical 2-layered structure of the myocardium with a thin, compacted outer (epicardial) band and a much thicker, noncompacted inner (endocardial) layer consisting of trabecular meshwork with deep endocardial spaces (the maximum end systolic of the noncompacted to compacted myocardium of  $>2$  is characteristic). Measure in parasternal short axis at end systole.
3. Predominant segmental location of the abnormality (i.e., noncompacted myocardium is predominantly [ $<80\%$ ] found in the apical and midventricular areas of both the inferior and lateral wall).
4. Color Doppler echocardiographic evidence of deeply perfused intertrabecular recesses (without communication with coronary circulation). [9]

Two dimensional echocardiography with color and doppler studies is the standard diagnostic procedure for isolated ventricular non-compaction. This technique allows the determination of localization and extent of isolated ventricular non-compaction, atrial and ventricular sizes, and both systolic and diastolic ventricular function. Magnetic resonance

imaging (MRI) can provide multiple tomographic sections of the ventricles in patients with ventricular and cardiac deformities and has been applied in children with congenital heart disease. Echocardiography and magnetic resonance imaging findings demonstrated a good correlation as far as localization and extent of isolated ventricular non-compaction were concerned. Magnetic resonance imaging, however, offers no additional information compared to echocardiography, except for the detection of a thrombus which can be hidden in the spongeliike myocardium of isolated ventricular non-compaction. [5]

Diagnostic criteria of isolated ventricular noncompaction are focused on description of the left ventricle. Most notably, the absence of coexisting cardiac abnormalities has been supposed as one of the criteria for diagnosis of isolated ventricular non-compaction. [10]

Isolated right ventricular non-compaction is a rare entity with little data on diagnosis and management strategies. Diagnosis is often delayed probably contributing to the adverse prognosis. [11]

The most common clinical presentations of ventricular non-compaction described in literature were heart failure, arrhythmias and cardioembolic events. Table 6 (Appendix 2) compares the different presentations of the reported cases of isolated right ventricular non-compaction. It occurs most commonly among males on the third decade of life and presents with heart failure symptoms.

Our patient demonstrated a two-layered right ventricle in the apical region, mid and superior regions of the right ventricular free wall which was most pronounced at the apical region on 2D echocardiogram. Deep inter-trabecular spaces communicating with the right ventricular cavity were present. Transesophageal echocardiography confirmed the absence of other cardiac anomalies. Following the same criteria used for left ventricular non-compaction as proposed by Oechslin et. al., above findings confirmed that she has an isolated non-compacted right ventricle. However, she was also found to have severe pulmonary hypertension for which no other identifiable causes have been identified. Our patient was given medications for heart failure and pulmonary hypertension as well as oral anticoagulation.

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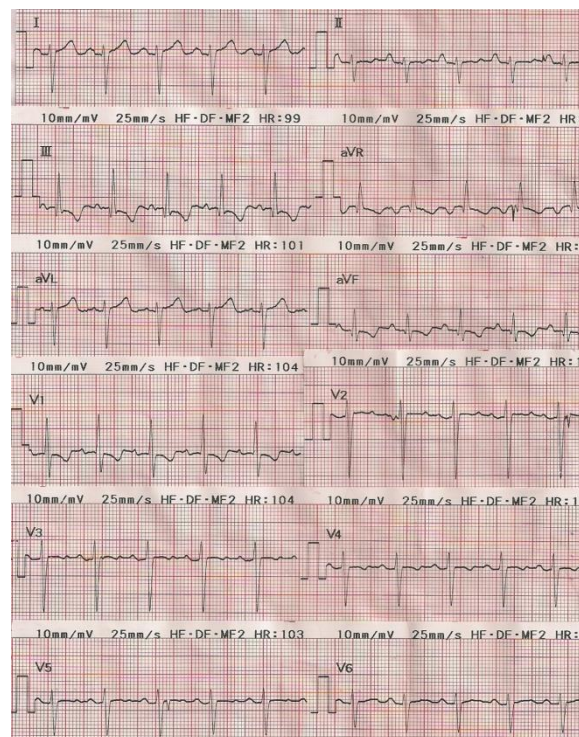
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## APPENDIX

### APPENDIX1: FIGURES



**Figure 1: 12L ECG**  
AR 103 bpm, VR 103 bpm, PR 0.20 s, QRS 0.08 s, QT 0.36 s, axis +120°; Sinus tachycardia, Right axis deviation, Right ventricular hypertrophy

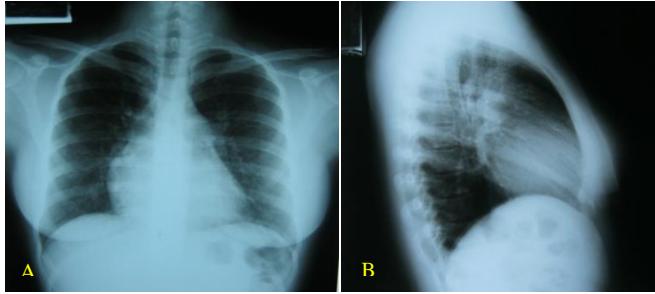
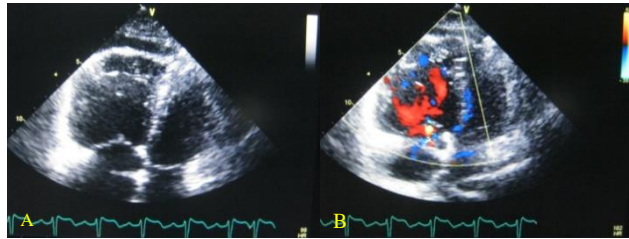


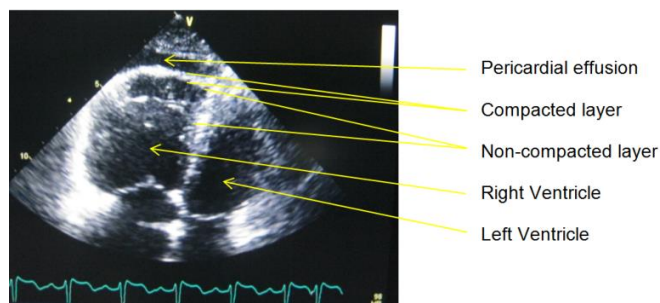
Figure 2: **Chest X-Ray** (A. Postero-Anterior View and B. Lateral View)

The lung fields are clear. There is prominence of the pulmonary arteries and its proximal branches on the left side. The heart is borderline in size. There is fullness of the left hilum, which on the lateral view may be secondary to fullness of pulmonary vessels. The diaphragm and sinuses are intact.

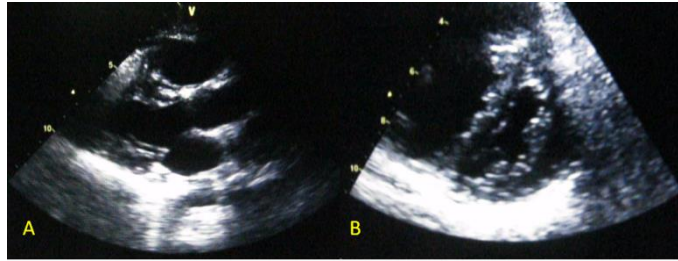


**Figure 3.1** Transthoracic 2D Echocardiogram showing Plain 4 Chamber View (A) and Color Doppler imaging (B) demonstrating blood flow through these deep recesses in continuity with the ventricular cavity. The right ventricle is dilated with thickened walls with depressed systolic function by TAPSE (1.4 cm) and Fractional area change (19%). The non-compacted to compacted myocardial layer ratio is 4.32: 1 at the right ventricular apex.

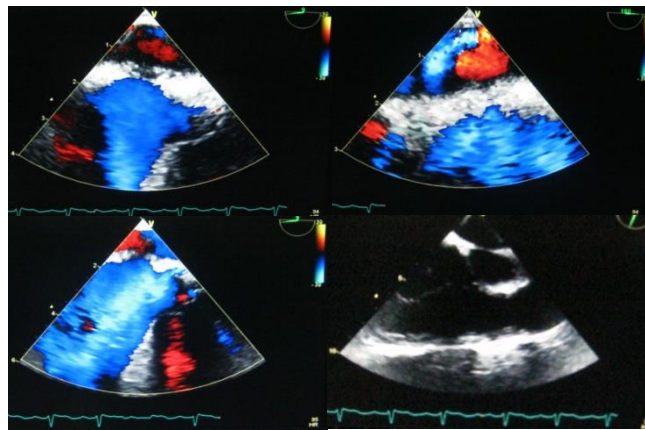
TAPSE = tricuspid annular plane systolic excursion



**Figure 3.2:** Transthoracic 2D Echocardiogram (4 Chamber View) showing layers of non-compacted myocardium and compacted myocardium on the right ventricle.



**Figure 3.3 Transthoracic 2D Echocardiogram Parasternal Long Axis (A) and Parasternal Short Axis (B) Views.** The transthoracic 2D echocardiogram showed Non-compacted right ventricular myocardium with depressed systolic function by TAPSE and Fractional Area Change; Concentric left ventricular remodeling with good wall motion and contractility and normal resting systolic function with grade 1 diastolic dysfunction. Ejection fraction was 63%. Dilated right atrium (40) with no evidence of thrombus. Minimal pericardial effusion (echo free space measuring 0.96 cm anterior to the right ventricle). Doppler Studies showed severe tricuspid regurgitation, pulmonic regurgitation and severe pulmonary hypertension by tricuspid regurgitation jet (70 mmHg) and tricuspid regurgitation regression formula (67.58 mmHg); TAPSE = tricuspid annular plane systolic excursion



**Figure 4** Transesophageal Echocardiogram showed intact interatrial septum and interventricular septum. The right atrium is noted to be dilated.

**APPENDIX 2: LABORATORY AND ANCILLARY RESULTS**

**Table 1: Arterial Blood Gas**

	<b>December 31, 2012</b>	<b>January 1, 2013</b>
<b>pH</b>	7.581	7.585
<b>pCO2</b>	27.2 mmHg	29.0 mmHg
<b>pO2</b>	71.5 mmHg	73.6 mmHg
<b>Temp</b>	37.0°C	37.0°C
<b>FiO2</b>	21.0%	21.0%
<b>BP</b>	760.0 mmHg	759.5 mmHg
<b>HCO3</b>	25.5 mmol/L	27.5 mmol/L
<b>O2sat</b>	96.5%	96.8%
<b>BE</b>	6.0 mmol/L	7.7 mmol/L
<b>TCO2</b>	26.3 mmol/L	28.4 mmol/L
<b>O2CT</b>	20.3 VOL%	20.4 VOL%
<b>BB</b>	54.0 mmol/L	55.7 mmol/L
<b>SBE</b>	4.5 mmol/L	6.5 mmol/L
<b>AaDO2</b>	45.7 mmHg	41.2 mmHg
<b>a/A</b>	0.61	0.64
<b>RI</b>	0.6	0.6
<b>Interpretation</b>	Partially compensated metabolic alkalosis with inadequate oxygenation at room air	Partially compensated metabolic alkalosis with inadequate oxygenation at room air

**Table 2: Bleeding Parameters**

	Reference	Jan 1	Jan
<b>Prothrombin time</b>	10.3-14.1	12.7 s	27.9 s
<b>Normal control</b>		10.6 s	11.7 s
<b>PT ratio</b>		1.0	2.3
<b>INR</b>	0.8-1.3	1.0	2.3
<b>Percent Activity</b>			22.3%
<b>Activated PT</b>	27.0-45.4	43.5 s	
<b>Normal Control</b>		37.0 s	

**Table 3: Blood Chemistries**

	Reference	December 31, 2012
<b>D-dimer</b>	Less than 0.50	0.36 mg/L FEU
<b>BUN</b>	9-23	8.76 mg/dL
<b>Creatinine</b>	0.5-1.2	0.85 mg/dL
<b>Sodium</b>	137-147	137.00 mmol/L
<b>Potassium</b>	3.8-5.0	4.03 mmol/L
<b>Magnesium</b>	1.6-2.59	2.25 mg/dL
<b>Ionized Calcium</b>	1.12-1.32	1.22 mmol/L

**Table 4: Complete Blood Count**

	Reference	Result
<b>Hemoglobin</b>	120-170	161 g/L
<b>RBC</b>	4.0-6.0	5.12 x 10 <sup>12</sup> /L
<b>Hematocrit</b>	0.37-0.54	0.48
<b>MCV</b>	87 +/- 5	94.00 U <sup>3</sup>
<b>MCH</b>	29 +/- 2	31.40 pg
<b>MCHC</b>	34 +/- 2	33.40 g/dL
<b>RDW</b>	11.6-14.6	12.80
<b>MPV</b>	7.4-10.4	9.10 fL
<b>Platelet</b>	150-450	230 x 10 <sup>9</sup> /L
<b>WBC</b>	4.5-10.0	17.80 x 10 <sup>9</sup>
<b>DIFFERENTIAL COUNT</b>		
<b>Neutrophils</b>	0.50-0.70	0.82
<b>Segmenters</b>	0.50-0.70	0.82
<b>Lymphocytes</b>	0.00-0.07	0.17
<b>Monocytes</b>	0.20-0.40	0.01

**Table 5: Other work-ups**

	Reference	Result
<b>Protein C Activity</b>	70.0-140.0	91.1 %
<b>Protein S Activity</b>	60.0-150.0	74.6%
<b>Anti-Cardiolipin IgG</b>	<23	9.30 GPL
<b>Anti-Cardiolipin IgM</b>	<11	7.29 MPL
<b>Anti-Nuclear Antibody</b>		NEGATIVE

**APPENDIX 3: Peripheral Venous Duplex Scan of the Lower Extremities**

Right: No evidence of Venous thrombosis

Competent sapheno-femoral and sapheno-popliteal valves

Left: No evidence of Venous thrombosis

Deep vein valve reflux on the distal posterior tibialis vein

Superficial vein valve reflux on the distal greater saphenous vein at the thigh

Competent sapheno-femoral and sapheno-popliteal valves

**APPENDIX 4: Peripheral Venous Duplex Scan of the Upper Extremities**  
Normal venous duplex scan of the upper extremities

**Table 6 Comparison of the Clinical presentations of previously published cases of isolated right ventricular non-compaction**

Case	Age (years)	Gender	Clinical presentation		
			Arrhythmias	Heart failure	Others
Fazio, G. et. al Case 1 (2010)	N/A	N/A	Complex ventricular extrasystoles		
Fazio, G. et. al Case 2 (2010)	3				Sudden death
Maheswari, M et. al. (2012)	40	Male		Exertional dyspnea, palpitation and swelling over both lower extremities and abdomen	
Gomathi, S. et. al. (2008)	23	Male		Exertional dyspnea, facial puffiness, pedal edema and exertional palpitation	
Song, Ze Zhou (2008)	23	Male	Sinus irregularity, right axis deviation, inverted T waves, multiple left bundle branch block type extrasystolic ventricular beats and abiogenesis atrial premature beats	Chest distress and exertional palpitation	
Zhang, XJ et. al. (2009)	23	Female		Bipedal edema, anorexia, abdominal swelling and exertional shortness of Breath	
Song, Ze Zhou (2011)	69	Male		Mild chest distress and exertional palpitations	
Vicera et. al.	25	Female		Exertional dyspnea	

N/A = not available