Left Ventricular Noncompaction Cardiomyopathy Managed with Cardiac Resynchronization Therapy: A Case Report

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Received on: 06 March 2022; *Accepted on:* 04 May 2022; *Published on:* 31 August 2022

Abstract

Although "spongy myocardium" was first observed by Grant in 1926, left ventricular noncompaction (LVNC) cardiomyopathy was originally described by Engberding and Bender in 1984.[1] Left ventricular noncompaction cardiomyopathy is characterized by abnormal ventricular myocardial protrusions with a thin layer of properly compacted myocardium.[2,3] It is a rare entity, with a prevalence less than 0.02%.[4] We report a case of LVNC cardiomyopathy with dyssynchrony, which was successfully managed with cardiac resynchronization therapy (CRT).

Keywords: Cardiac resynchronization therapy, Cardiomyopathy, Conduction abnormality, Dyssynchrony, Heart failure with reduced ejection fraction, Left ventricular noncompaction.

Bengal Physician Journal (2022): 10.5005/jp-journals-10070-7081

Introduction

Although "spongy myocardium" was first observed by Grant in 1926, LVNC cardiomyopathy was originally described by Engberding and Bender in 1984.[1] Left ventricular noncompaction cardiomyopathy is characterized by abnormal ventricular myocardial protrusions with a thin layer of properly compacted myocardium.[2,3] It is a rare entity, with a prevalence less than 0.02%.[4] We report a case of LVNC cardiomyopathy with dyssynchrony, which was successfully managed with CRT.

Clinical Presentation

A 42-year-old nondiabetic, nonhypertensive housewife came to the outpatient department (OPD) with a 3-month history of exertional shortness of breath, fatigue, and episodes of paroxysmal nocturnal dyspnea without any fever, angina, transient loss of consciousness, skin rash, joint pain, bleeding from any site, altered urine characteristics, or over-the-counter drug intake. There was no significant medical history. On clinical examination, she was tachypneic, with a pulse rate of 62/minute (regular), an elevated jugular venous pressure (JVP), pedal edema, a BP of 100/66 mm Hg, bibasal rales, soft heart sounds, LV-S3, and no lymphadenopathy.

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How to cite this article: Mukhopadhyay M, Som P, Mondal R, *et al*. Left Ventricular Noncompaction Cardiomyopathy Managed with Cardiac Resynchronization Therapy: A Case Report. Bengal Physician Journal 2022;9(2):35–40.

Source of support: Nil

Conflict of interest: None

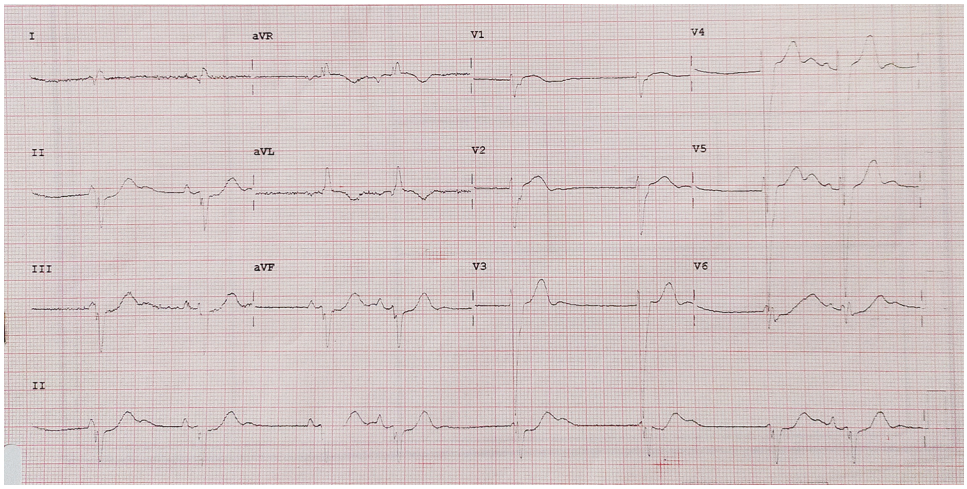

Fig. 1: Pre-CRT ECG

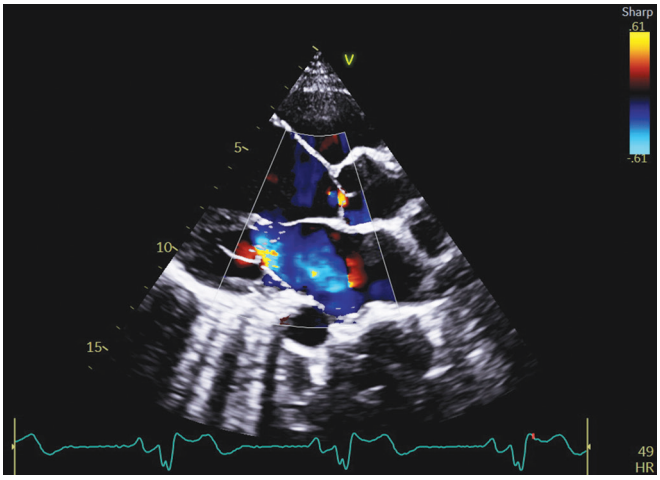

Fig. 2: Pre-CRT echo

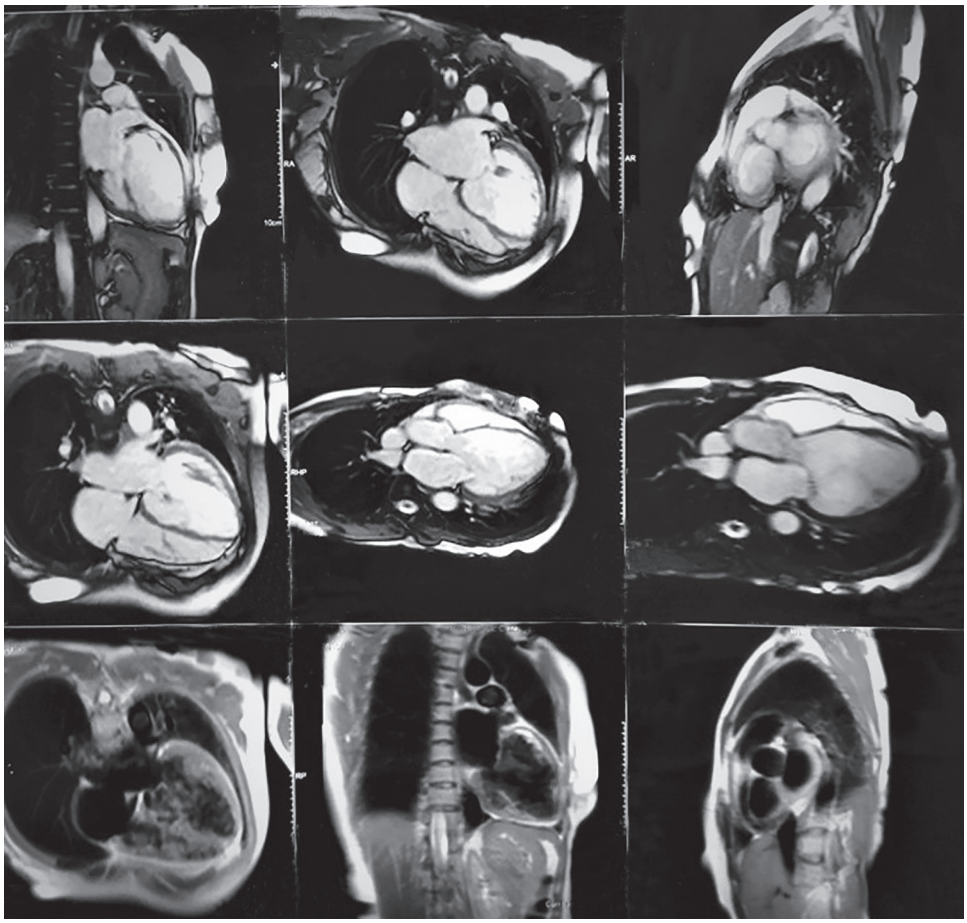

Fig. 3: Cardiac MRI image

There was neither abdominal organomegaly nor any focal neurodeficit. Hence, she was provisionally diagnosed to have decompensated left ventricular failure (at NYHA III).

Investigation Profile

Baseline investigations showed a normal complete hemogram with ESR. Blood sugar, lipid profile, liver function test, thyroid-stimulating hormone test, and renal profile with electrolytes including calcium were normal. She was seronegative for HIV and hepatitis B and C viruses. Both prothrombin time/international normalized ratio (PT/INR) and activated partial thromboplastin time (aPTT) were normal. Routine urine examination was noncontributory. A chest X-ray showed an increased CT ratio with pulmonary venous congestion without any mediastinal widening. Electrocardiogram (ECG) (Fig. 1) is suggestive of SA node dysfunction, a left anterior hemiblock pattern with a notched QRS and wide QRS (160 ms), S in V1, and a predominant positive QRS in I and aVL. Initial echocardiography with Doppler study

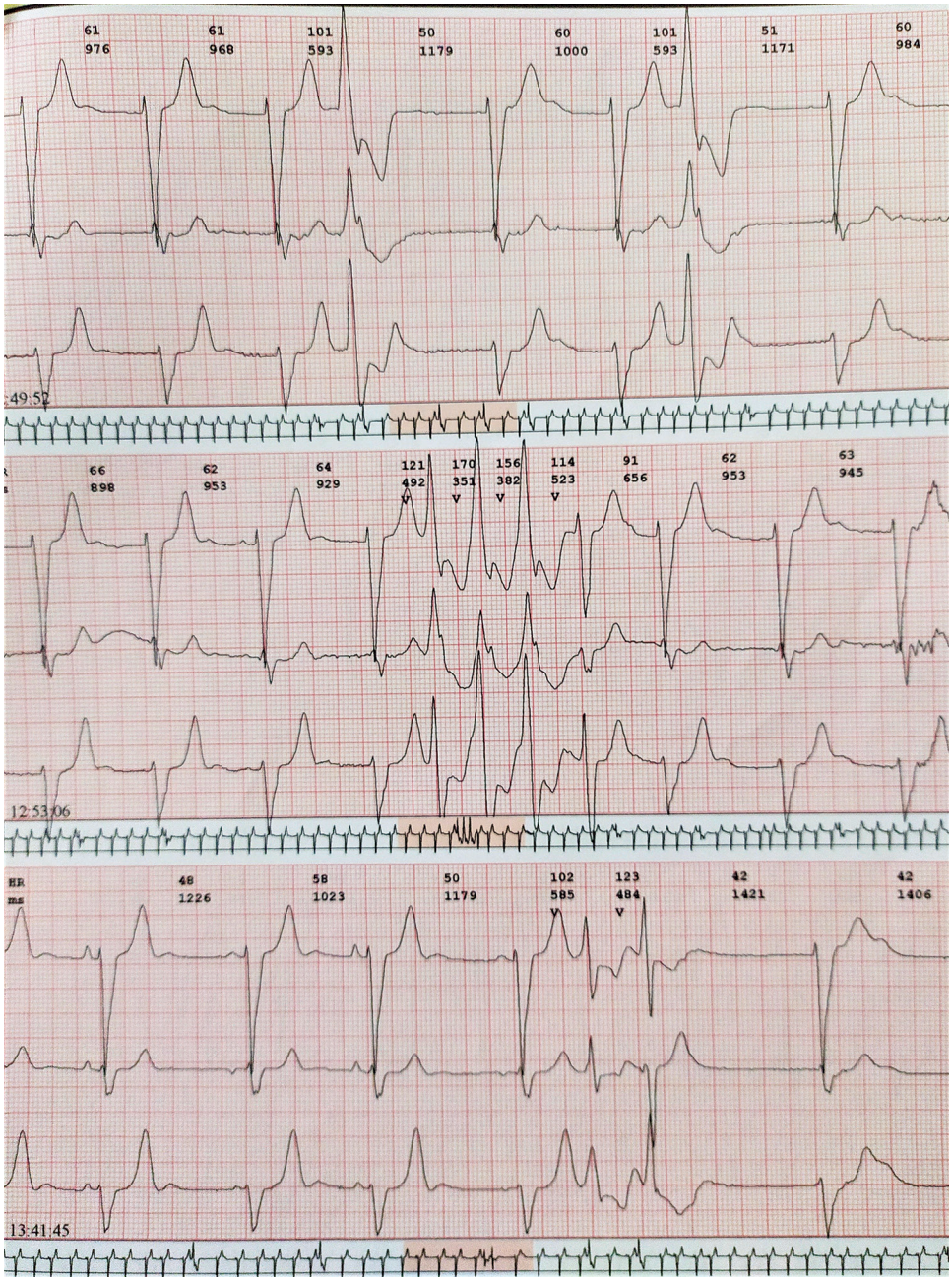

Fig. 4: Holter monitoring showing ventricular premature complex (VPCs) and NSVT

(Fig. 2) confirmed the diagnosis of dilated cardiomyopathy with LV systolic dysfunction (left ventricular ejection fraction (LVEF) 25%); there was thinning of the interventricular septum (IVS). The RV systolic function was preserved, with a mean pulmonary artery pressure of 30 mm Hg. Coronary angiography revealed normal epicardial coronary arteries, with good TIMI 3 flow. Antinuclear antibody (ANA), antineutrophil cytoplasmic antibodies (ANCA), rheumatoid factor (RA), C-reactive protein (CRP), and serum angiotensin converting enzyme (ACE) all were normal. Contrast-enhanced computed tomography thorax revealed no mediastinal lymphadenopathy. Cardiac magnetic resonance imaging (MRI) (Fig. 3) showed areas of marked thinning of the IVS, but the atrioventricular septum was normal, trabeculations were noted in the mid-LV cavity, and apical sections with noncompaction: compact wall thickness in the end diastole was 4.5:1 (>2.3:1), scarring was found in the anterolateral zone, and CMR-based LVEF was 22%. Hence, the final diagnosis was LVNC cardiomyopathy with reduced ejection fraction with conduction abnormality.

Management and Follow-up

She was given standard conservative therapy for heart failure with ACE inhibitor, diuretics, spironolactone, and beta-blocker. But she could not tolerate standard medications even at a lowest possible effective dose for a reasonable period and returned back with extreme fatigue, recurrent pre-syncope, a pulse rate of 45/minute, and a BP of 84/50 mm Hg. Conduction abnormalities and hypotension were hampering essential drug optimization

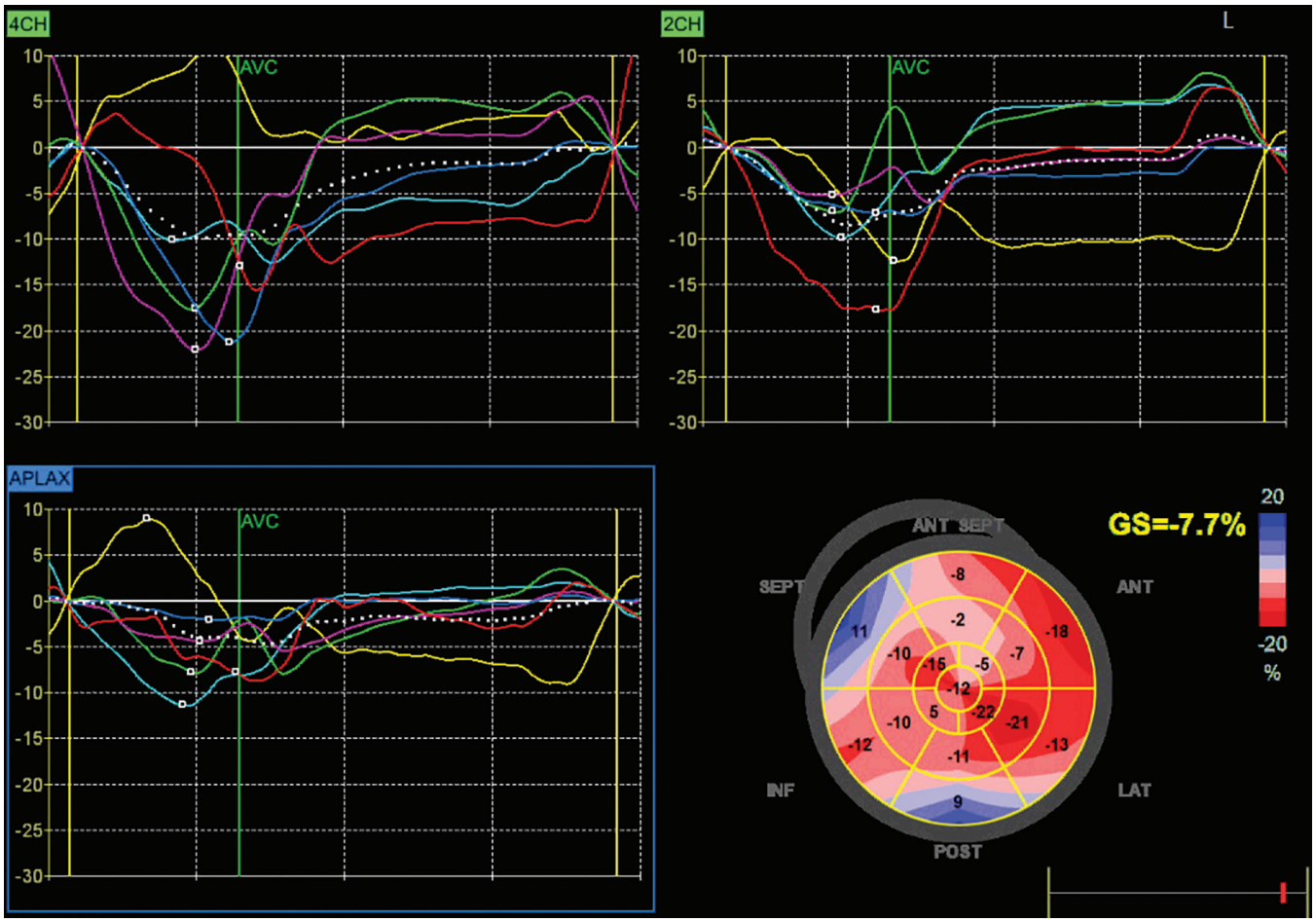

Fig. 5: Pre-CRT 2D LV strain imaging showing dyssynchrony

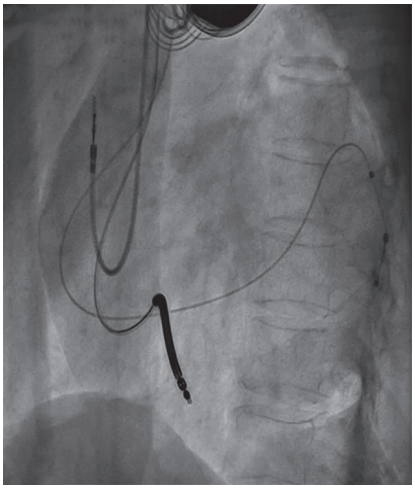

Fig. 6: CRT-D lead position under fluoroscopy in left anterior oblique (LAO) projection

repeatedly. Her 24-hour Holter monitoring (Fig. 4) revealed the presence of nonsustained ventricular tachycardia (NSVT). She underwent advanced 2D LV strain echocardiography (Fig. 5) with dyssynchrony assessment. It revealed a septal-to-posterior wall mechanical delay of 160 ms, an MR dp/dt of 522 Hg/second, an interventricular mechanical delay of 48 ms, a QRS-isovolumetric contraction onset of 38 ms, a QRS-peak S of 219 ms, a 2D LV global longitudinal strain (GLS) score of 7.7%, an LV <5, and an LV Tei index of 1.3. Considering the presence of dyssynchrony, she was planned for cardiac resychronization therapy defibrillator (CRT-D). Cardiac resychronization therapy defibrillator (Fig. 6) implantation was carried out successfully with good electrical parameters. Post-CRT ECG (Fig. 7) revealed complete atrioventricular synchrony with 100% capture, narrow QRS (100 ms), R in V1, negative QRS complex in I and aVL, and QRS in lead II is more negative than that in lead III. Post CRT, her quality of life (QoL) improved significantly (NYHA I), drug optimization became possible, LVEF increased to around 45% in biplane (Figs 8 and 9), the 2D LV GLS score improved to 12%, MR dp/dt improved (>1000 mm Hg/second), LV filling pressure normalized (E/e' = 9.5), LV became 6, and the LV Tei index became 0.7. A regular OPD basis follow-up has been uneventful over 2 years. A regular follow-up CRT programming revealed (Cardiac Compass) less fluid overload and increasing physical activities over time (Figs 10 and 11).

Discussion

By the end of fourth week of embryonic cardiac development, physiological trabeculations increased the cardiac output and surface area for oxygen and nutrient absorption.[1] Trabeculation is followed by compaction. Defect at any of these stages can result in persistence of trabeculations. Cardiac imaging has

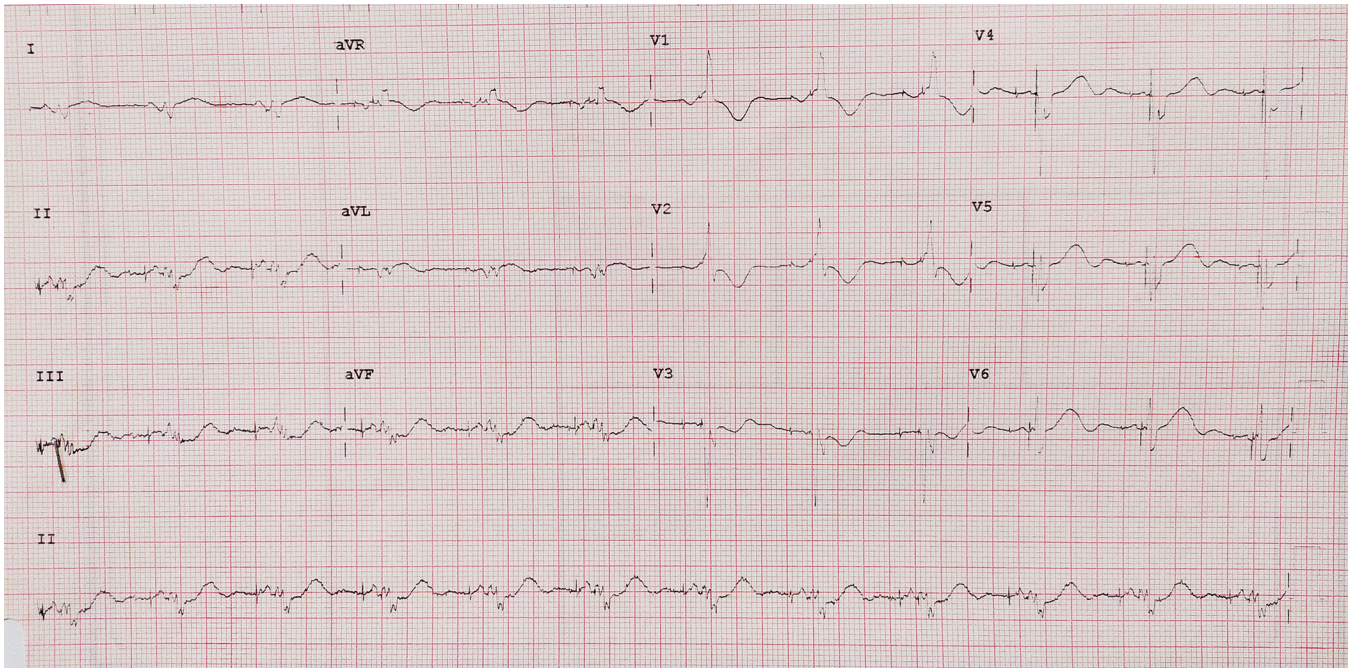

Fig. 7: Post-CRT-D ECG showing narrowing of the QRS duration

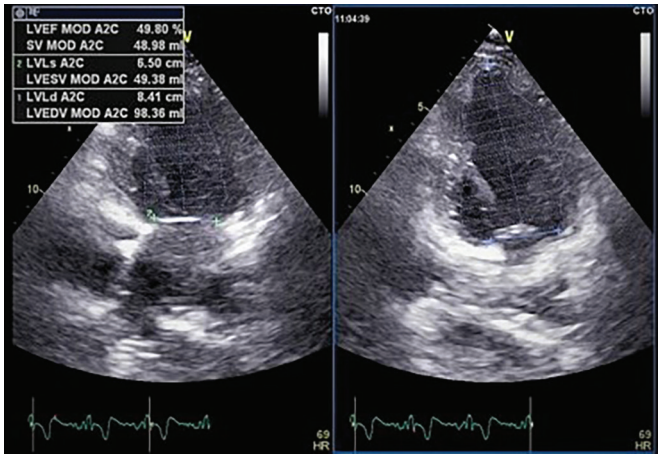

Fig. 8: Post-CRT-D echo showing improved LVEF

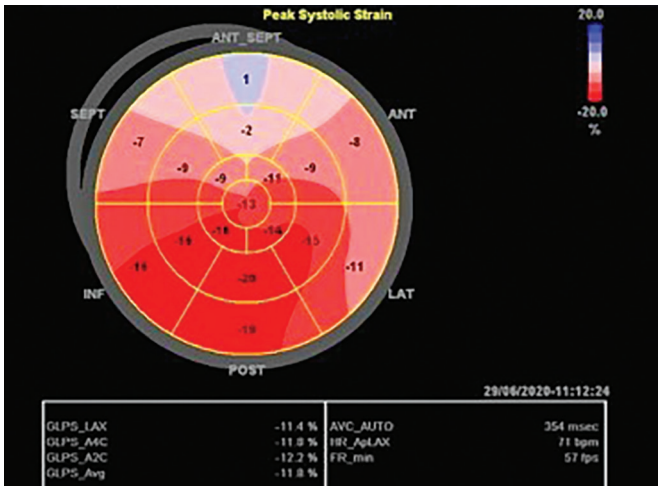

Fig. 9: Post-CRT 2D LV strain echo showing relatively more synchronous LV contraction with improvement of the LV 2D GLS score

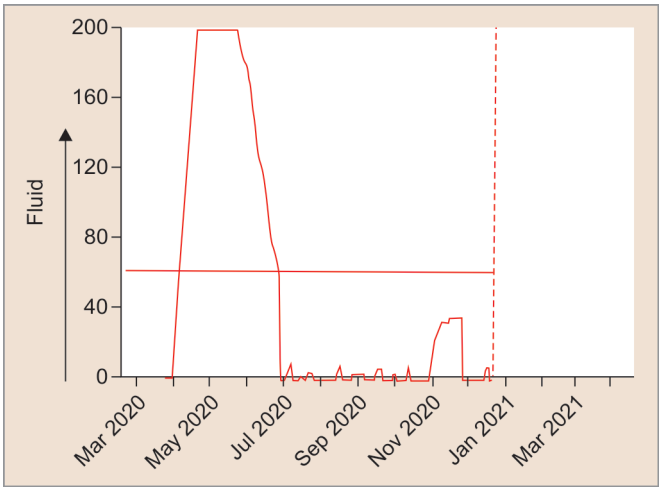

Fig. 10: CRT follow-up programming showing progressive lower fluid accumulation due to improvement of LV systolic function

proved an increased prevalence of excessive trabeculation (possibly due to hemodynamic reason) in "athlete's heart", 25% of primigravida women (reversible), chronic renal failure secondary to polycystic kidney disease, sickle cell anemia, and beta-thalassemia.[5] The clinical presentation of LVNC cardiomyopathy can be varied; for example, incidentally detected during cardiac imaging in asymptomatic individuals, overt LVNC cardiomyopathy with symptomatic heart failure, conduction abnormalities, tachyarrhythmia, and sudden cardiac death.[2] Systolic dysfunction seen in LVNC is due to hypoperfusion secondary to subendocardial microvascular abnormalities and dyssynchrony between the compacted and noncompacted myocardial layers.[6] Electrocardiogram abnormalities are noticed in over 80% of patients with LVNC, such as intraventricular conduction delay, AV block, repolarization abnormalities (early repolarization and corrected QT interval (QTc) prolongation),

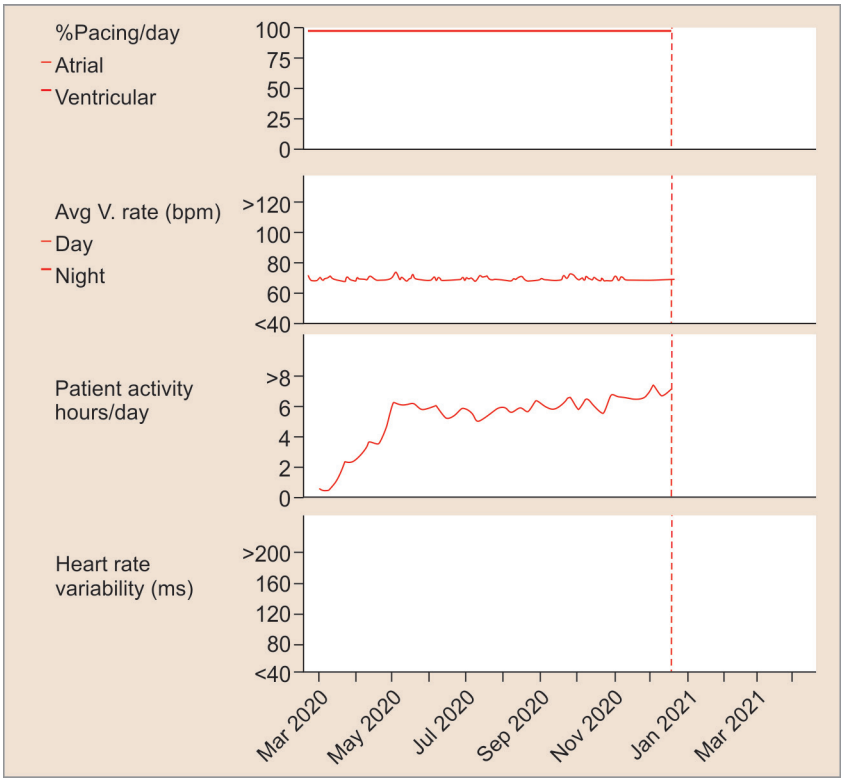

Fig. 11: Post-CRT follow-up programming showing progressively increased physical activity of patient, that is, improved QoL

supraventricular tachyarrhythmia (most frequently atrial fibrillation), and electrocardiographic evidence of left ventricular hypertrophy.[6,7] Repolarization disturbances predispose patients to malignant ventricular tachyarrhythmias and sudden cardiac death.[6,8] Cardioembolic strokes in LVNC are secondary to mural thrombi formed in the intertrabecular recesses.[2] The primary prophylactic anticoagulation is recommended for patients with LVNC cardiomyopathy with a reduced ejection fraction <40%, and/or atrial fibrillation, and patients with intracardiac thrombi.[6,9] The validated diagnostic criteria are Jenni criteria for echocardiography and Peterson criteria for cardiac MRI (most sensitive).[10,11] Our patient had LVNC cardiomyopathy with a reduced ejection fraction with conduction abnormalities and low blood pressure with bradycardia, which hampered drug optimization for heart failure treatment. Hence, early implantation of the CRT device was planned. The CRT-D was chosen over CRT-P as there was scar in CMR and NSVT in Holter monitoring, along with a history of pre-syncope. Our timely decision not only helped in drug optimization but also translated into good post-CRT LV functional improvement with sustained satisfactory QoL of our patient (Figs 10 and 11).

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