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A Review of Left Ventricular Non-compaction Cardiomyopathy (LVNC)

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Abstract

Left ventricular non-compaction (LVNC) is a rare congenital phenotype defined by the presence of prominent left ventricular trabeculae, deep intertrabecular recesses (continuous with the ventricular cavity), and a thin compacted layer. The most common presentation of LVNC is dyspnea (60%), followed by palpitations (18%), chest pain (15%), syncope (9%), and prior stroke (3%). LVNC presenting with acute myocardial infarction (MI) has rarely been reported in the literature. A forty-one-years old female presented with substernal chest pain and exertional dyspnea. On physical examination, she was alert without any distress, her lungs and heart examination were within normal limits. Peripheral pulses were palpable and regular, and +1 peripheral pitting edema was noted. EKG showed normal sinus rhythm with premature atrial contractions (PACs), left axis deviation, and ST-segment and T wave changes suggestive of inferior wall ischemia. Troponin I level was found to be elevated, which peaked within 24 hours, $Troponin_{max}$ 110.08 ng/ml. Transthoracic echocardiography showed moderate LV dilatation with severely reduced EF (15–20%), and diffuse LV hypokinesis with a grade III restrictive pattern. There was heavy trabeculation of LV involving 2/3rd LV endocardium and wall thickness with sinusoidal tunnels perpendicular to LV wall. These morphological findings met the diagnostic criteria of LVNC/NCM. LVNC presenting with acute myocardial infarction (MI) can be related to poor outcomes, however, more data is needed to establish the clinical implication of this presentation. Asymptomatic LVNC can be observed while symptomatic LVNC should be treated with standard guidelines of HF.

Keywords: Cardiomyopathy, Myocardial infarction, Heart failure, Noncompaction

1. Introduction

Left ventricular non-compaction (LVNC) is a rare congenital phenotype defined by the presence of prominent left ventricular trabeculae, deep intertrabecular recesses (continuous with the ventricular cavity), and a thin compacted layer. It is suggested that the origin of LVNC is attributable to arrested compaction or failure in the solidification of the myocardial primordium during embryogenesis. Thickened myocardium consists of two distinct layers; a thin compacted epicardial layer and a thickened non-compacted endocardial layer. Isolated LVNC does not imply LV dysfunction and it can be observed in asymptomatic individuals with normal LV size and wall thickness. However, LVNC can present with dilated, restrictive, or hypertrophic cardiomyopathy.[1,2] The prevalence of LVNC in the general population is not known, an estimated prevalence of 0.014–1.3% has been noted among patients undergoing echocardiography.[3] NCM can be sporadic or familial. However, a higher prevalence (3–4%) has been reported among patients with heart failure.[4] Almost 12–50% of reported cases have a positive family history of non-compaction cardiomyopathy with variable inheritance patterns including autosomal dominant, X linked, and autosomal recessive inheritance.[5] The most common presentation of LVNC is dyspnea

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(60%), followed by palpitations (18%), chest pain (15%), syncope (9%), and prior stroke (3%). At baseline, 31% of the patient population had New York Heart Association (NYHA) functional class III or IV heart failure. A systemic review illustrated that the majority (56%) of the patient population diagnosed with LVNC was initially referred for heart failure symptoms, and up to 11% were diagnosed via screening.[6] In the absence of census criteria, the diagnosis of LVNC should be considered in patients with distinctive trabeculations on cardiac echocardiography even without heart failure symptoms. In clinical practice, an imaging (echocardiography & cardiac magnetic resonance imaging) based ratio between compacted and non-compacted (NC) layers are currently used to diagnose this rare phenotype.[7,8] In addition to heart failure, thromboembolic events and arrhythmias are other major complications of this rare phenotype. LVNC presenting with acute myocardial infarction (MI) has rarely been reported in the literature.

2. Methods and results

We queried PubMed with "Isolated Noncompaction of the Ventricular Myocardium" [Mesh] with an additional filter of 'case reports' in the last 20-years between 2012 and 2022, and we found a total of 201 cases. On further screening, we included only cases of left ventricular non-compaction in adults. We reviewed a total of 60 cases of LVNC - their presentation and treatments are tabulated below.

3. Discussion

LVNC has been defined as genetic cardiomyopathy (CMP) by the American Heart Association (AHA) and categorized as nonclassified CMP by the European Society of Cardiology. However, with evolving evidence, LVNC is considered as a distinct CMP. LVNC is classified into the following subgroups: iLVNC associated with normal systolic and diastolic function; LVNC associated with LV dysfunction and dilatation at onset; LVNC morphology in patients fulfilling the criteria for hypertrophic CMP, restrictive CMP, or dilated CMP. In addition, LVNC can present as a transient phenomenon in conditions like myocarditis or systemic lupus erythematosus (SLE). Different genes (more than 10) that primarily code for cytoskeletal, sarcomeric and mitochondrial proteins have been implicated in genetic mutations of LVNC. The Heart Failure Society of America 2018 guidelines recommended obtaining a family history up to three generations and screening first-degree relatives of diagnosed LVNC patients.[9] NCM is mostly diagnosed in the pediatric group, but isolated left ventricular noncompaction cardiomyopathy has been reported in the elderly.[10]

3.1. Presentation

Heart failure was the most common presentation and a common reason for hospitalization in NCM.[6] Of the cases we reviewed, 63.3% of LVNC cases were males, and 36.6% were females. The most common presentation was heart failure (31 cases, 51.6%) followed by arrhythmias (12 cases, 20%), angina (5 cases, 8.3%), and stroke (7 cases, 11.6%).(Fig. 3) Rare presentations included 1 case of dissecting aneurysm, 1 of left ventricular thrombus, 2 of dizziness, and 1 of sepsis.

3.2. Diagnosis

The diagnosis of LVNC is primarily based on morphological findings on transthoracic echocardiography (TTE) or cMRI. Jenni criteria and Chin or Stöllberger criteria are the two most commonly used and widely accepted criteria to diagnose LVNC with TTE. The sensitivity and specificity of morphological criteria are unknown. In the absence of a census on diagnostic criteria and nonspecific symptoms associated with LVNC, preventing the overdiagnosis of LVNC should be a priority. NCM is most commonly incidentally seen on TTE when working heart failure and cMRI can be used for anatomical delineation or when TTE is inconclusive.

3.3. Prognosis

The prognosis of LVNC is primarily dependent on the severity of the disease (NYHA functional class) at presentation. Depressed LV function and late gadolinium enhancement (LGE) on cMRI are the worst prognosis signs as will be seen in our patient. A systematic review showed 14% mortality at 39th month follow-up and ~50% of deaths were from sudden cardiac arrest (SCA). Cardiovascular complications at presentation, NYHA class III or greater is a stronger predictor of cardiovascular death with a hazard ratio of 20.6 and 8.8, respectively.[11] Patients who are asymptomatic at the time of the initial presentation have a better prognosis. Follow up evaluation of the patients we reviewed revealed that 58.3% of patients showed an improvement; 10% deteriorated; 6.6% mortality was observed and 25% of patients did not have a follow-up (Fig. 4).

3.4. Management

It is theorized that cardiac embolic events occur due to thrombus formation in trabeculations in patients with severe systolic and diastolic dangiotensin-receptor blocker(s), or ARB(S)ysfunction and can present with stroke or rarely as a coronary embolism.[12] Limited information regarding the specific treatment of LVNC is available, and therapy is mostly based on HF symptoms, LVEF, the burden of thromboembolism, and the presence of arrhythmias. Heart failure should be managed as per standard guidelines. Anticoagulation should not be initiated in LVNC unless another indication (atrial fibrillation, intracardiac thrombus, or prior cardioembolic episode) is present. If a patient had an episode of sustained ventricular tachycardia or survived SCD, an ICD should be placed as a secondary prevention of SCA.[13] However, the role of ICD in primary prevention of SCD in LVNC with EF<40% is unclear. Patients presenting with HFrEF cardiac transplant should be considered earlier in their disease progression as successful cardiac transplants have been reported. In our review heart failure medication such as beta blockers, angiotensin-converting enzyme inhibitors(ACEI), angiotensin-receptor blockers(ARBs) and diuretics lead to improved ejection fraction and functioning class in most patients. Anticoagulants were used in patients where thrombosis was present during diagnosis. Below is a table representing management strategies that were employed and the outcome from each of them (Table 1). Screening TTE of first degree relatives is recommended.

4. Case presentation

A forty-one-years old female presented with substernal chest pain and exertional dyspnea. She also reported occasional palpitations and generalized weakness. The patient's family history was not obtained although it would have been extremely pertinent. Her past medical history was significant for chest pain three years ago. However, diagnostic workups including serial EKGs, troponin I, and stress test were non-conclusive at that time. At admission, her vital signs were: blood pressure 93/63 mmHg, heart rate 58 beats/minute, respiratory rate 16 breaths/minute, and oxygen saturation were above 95% on room air. On physical examination, she was alert without any distress, her neck examination did not reveal any bruit or jugular venous distension. Heart examination showed regular S1 and S2, and no added sound or murmur was auscultated. A lung examination showed good bilateral air entry without any rhonchi or rales. Peripheral pulses were palpable and regular, and +1 peripheral pitting edema was noted. EKG showed normal sinus rhythm with premature atrial contractions (PACs), left axis deviation, and ST-segment and T wave changes suggestive of inferior wall ischemia. Troponin I level was found to be elevated, which peaked within 24 hours, Troponin$_{max}$ 110.08 ng/ml and gradually declined over

Table 1. Clinical outcomes based on management instituted for LVNC.

Management administered	Improvement in clinical status	Decline in clinical status	No follow up	Stable
AC and HF medication	5	1	4	1
HF medication only	8	1	1	
AC only		1	4	
Anti-arrhythmic only		1		
PCI and HF medication	1		1	
Thrombectomy and AC	1	1		
ICD only		1		
Vasopressor therapy and diuretics		1		
Pacemaker implantation and HF medication	1		1	
CABG	1	1		
Radio-frequency ablation	3			
Mitral valve repair and HF medication	1			
Mitral valve repair	1			
Traditional medicine	1			
LV restoration surgery	1			
Transplant			1	
Observation only	3		1	3
Unknown			5	

* Two patients died close to presentation and were not included in this table.
AC - Anti-coagulation; HF - Heart failure; PCI - Per-cutaneous coronary interventions; CABG - Coronary artery bypass grafting; ICD - Implantable cardioverter defibrillator; LV - Left ventricle.

2 days. Transthoracic echocardiography showed moderate LV dilatation with severely reduced EF (15–20%), and diffuse LV hypokinesis with a grade III restrictive pattern. There was heavy trabeculation of LV involving 2/3rd LV endocardium and wall thickness with sinusoidal tunnels perpendicular to LV wall. These morphological findings were suggestive of LVNC/NCM. Transesophageal echocardiography (TEE) revealed mild spontaneous echo contrast in the left atrial appendage, however, no thrombus was visualized. Left heart catheterization showed a filling defect of the distal right posterior descending artery (PDA), consistent with a thrombus. Otherwise, normal coronary arteries with severe diffuse cardiomyopathy, out of proportion to coronary artery disease (CAD) were seen.

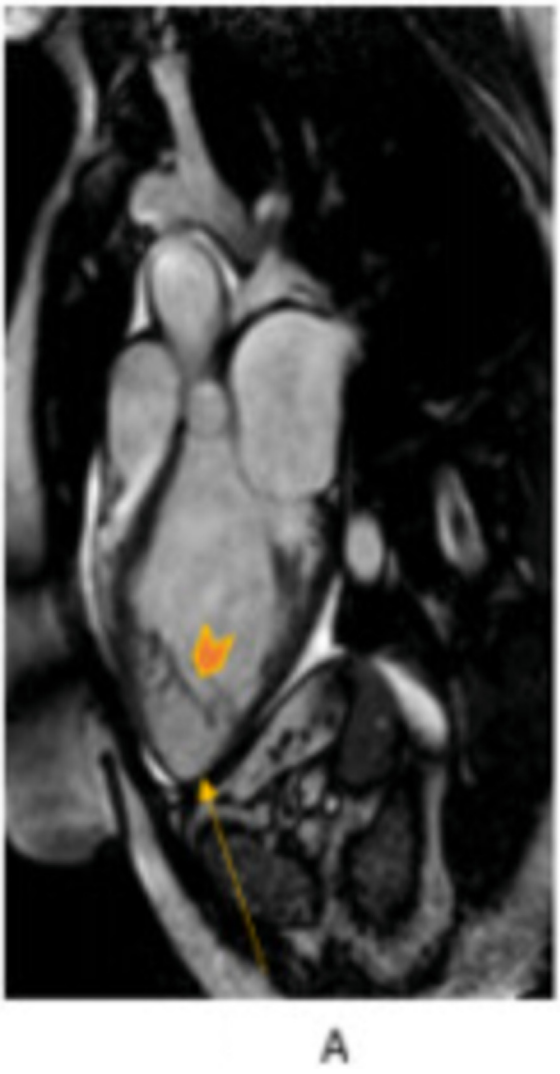
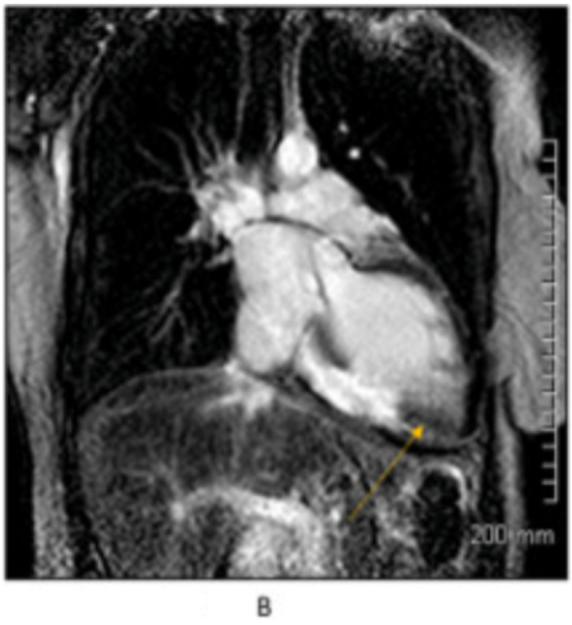


Fig. 1. Three chamber view demonstrates moderate left ventricular dilation with compaction (arrow) to non-compaction (arrow head) ratio of less than 1:2, consistent with left ventricular non-compaction.



B

Fig. 2. Contrast late enhancement study show heavy trabeculation of the left ventricular apex, linear focus of hypointensity (arrow) consistent with infract/scar.

The patient was treated conservatively and a cardiac MRI was obtained. Cardiac MRI(cMRI) revealed moderate left ventricle dilatation with severe dysfunction and heavy trabeculations of LV apex and mid-LV endocardium with a compact to non-compact wall ratio of less than 1:2 consistent with non-compaction cardiomyopathy (Fig 1). Contrast enhancement study showed transmural contrast late enhancement of mid inferior wall, a finding consistent with transmural myocardial infarction (Fig. 2).

4.1. Treatment & follow up

The patient was treated conservatively with dual antiplatelet, heparin, and statin therapy. She was not able to tolerate beta-blocker and heart failure standardized regime due to low blood pressure. She was offered an implantable cardiac defibrillator (ICD) but she refused. Her clinical course deteriorated aggressively with multiple admissions due to heart failure exacerbations. Three months later, she got readmitted with cardiogenic shock requiring ionotropic support. Her signs and symptoms did not improve with vasopressors. Treatment escalation to extracorporeal membrane oxygenation(ECMO) was discussed with the patient, however, the patient refused. She died due to sudden cardiac arrest, no cardiopulmonary resuscitation was performed (the patient was DNR).

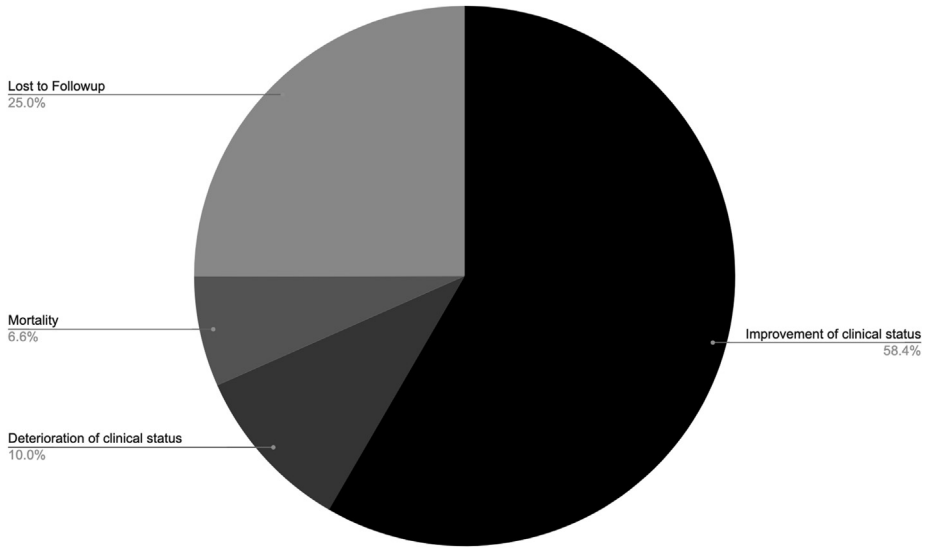


Fig. 3. Outcomes.

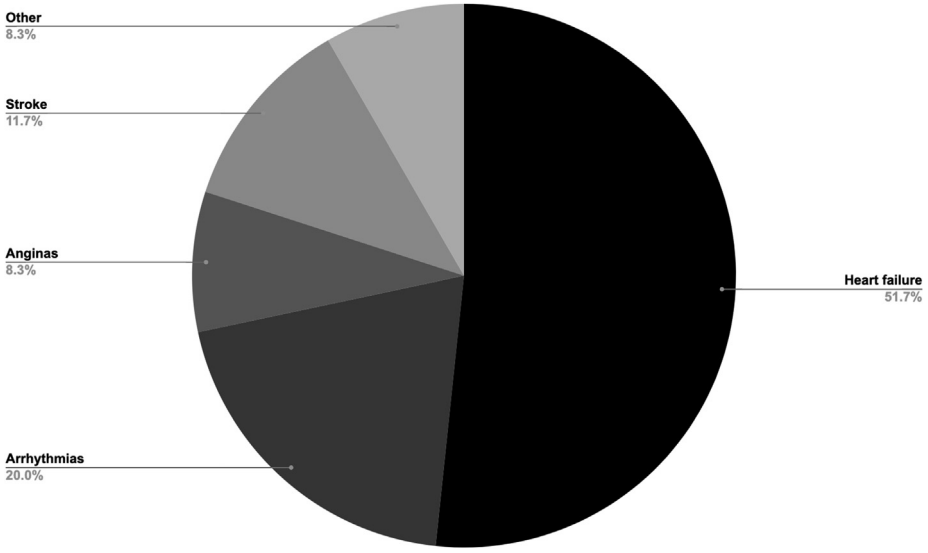


Fig. 4. Presentation.

5. Conclusions

LVNC and noncompaction cardiomyopathy (NCM) are two distinct entities. A diagnosis of LVNC should be considered in a patient who meets the morphological criteria on TTE even without clinical symptoms. However, if echocardiography is inconclusive, a cMRI should be obtained that will provide anatomic delineation. Screening (TTE) of first-degree relatives is recommended. Asymptomatic LVNC can be observed while symptomatic LVNC should be treated with standard guidelines of HF. LVNC presenting with acute myocardial infarction (MI) can be related to poor outcomes, however, more data is needed to establish the clinical implication of this presentation. Patients who have poor prognostic features should be considered for ICD and heart transplants earlier in the course of their disease.

Declarations

Consent for publication

The patient signed informed written consent for the publication that can be reproduced for editorial board review.

Ethics approval and consent to participate

Not applicable.

Availability of data and materials

Not applicable.

Appendix.

Author	Title	Age/Gender	Presentation	Management	Follow-up
Hugo Jesús Zetina-Tun, Guillermo Careaga-Reyna et al.	[Heart transplantation for the treatment of isolated left ventricular myocardial non-compaction. First case in Mexico]	20M	Heart failure	Transplanted	Survival to 15 months revealed class I New York Heart Association, and endo-myocardial biopsies did not reveal evidence of acute rejection.
Masanori Takamatsu, Keiji Kamohara, et al.	Effect of Noncompacted Myocardial Resection on Isolated Left Ventricular Noncompaction	65M	Heart failure	Left anterior descending artery and diagonal artery contained thrombi that were located and removed. Non-compacted trabeculae were resected.	Symptoms improved to New York Heart Association class I by his discharge on post-operative day 34. At follow-up echocardiography, the thrombus and non-compacted layer had disappeared
SBruno Ramos Nascimento,-Daniel Furtado Vidigal, et al.	Complete Atrioventricular Block As the First Manifestation of Noncompaction of the Ventricular Myocardium	32F	Complete AV block.	Dual-chamber pacemaker implantation	At 18-month follow-up, the patient is in NYHA functional class I, using diuretics and angiotensin-converting enzyme inhibitor.
Valentina Loriaa, Christian Colizzib, et al.	Left Ventricular Non-compaction: Cause or Consequence of Myocardial Disease? A Case Report and Literature Review	57F	Heart failure	Discharged against medical advise	Discharged against medical advise
Masaomi Chinushi, Kenichi Iijima, et al.	Suppression of Storms of Ventricular Tachycardia by Epicardial Ablation of Isolated Delayed Potential in Noncompaction Cardiomyopathy	65M	VT1 storms	Radiofrequency ablation.	2 weeks after RF ablation - no VTs with sotalol 80 mg daily
Wataru Miyake, Mayu Minemoto, et al.	Case Report of Left Ventricular Noncompaction Cardiomyopathy Characterized by Undulating Phenotypes in Adult Patients	57F	Heart failure	Percutaneous coronary intervention and heart failure medication	In the subsequent years, LVNC became less clear and LV function gradually improved
Wataru Miyake, Mayu Minemoto, et al.	Case Report of Left Ventricular Noncompaction Cardiomyopathy Characterized by Undulating Phenotypes in Adult Patients	36M	Heart failure	Diuretics and beta-blockers were used to treat congestive heart failure, together with anticoagulant for thrombolysis	6 months later improving LVEF to 54.3%
Giselle A. Baquero, Dustin J. Colegrove, et al.	Isolated Left Ventricular Noncompaction Causing Stroke in a 30-Year-Old Woman	30F	Stroke	Anticoagulation with warfarin	Six months later - no significant changes. 28 months later - substantial decline in LV systolic function and marked noncompaction of the posterolateral, inferior, and anterior walls.

Monu Rani, Rajesh Rajput, et al.	Non-compaction of ventricular myocardium with polycystic kidney disease with cardiogenic cerebral embolism	65M	Stroke	diuretics and oral anticoagulation for secondary stroke prevention and systemic embolism.	His brachial monoparesis was improving, and the patient has been doing fine on pharmacotherapy.
Marwa Elnazeir, Lisa Nobel, et al.	Left Ventricular Non-compaction Cardiomyopathy as a Potential Cause of Bilateral Posterior Cerebral Artery Stroke – a Rare and Unique Clinical Occurrence	63M	Stroke; Heart failure	tPA; eventually apixaban, aspirin and statin	3-month followup - residual visual field deficits. Cardiac outcome improved EF of 40%
Takafumi Ogawa, Sho Ogata , et al.	Left ventricular non-compaction with pulmonary capillary hemangiomatosis-like lesions: case report	52M	Heart failure	Diuretic and vasopressor therapy	Died from a worsening of the disease
Bianca Olivia Cojan-Minzat, Paul-Mihai Boarescu, et al.	Catheter ablation of a right posterior accessory pathway in a patient with left ventricular noncompaction: A case report	32M	Arrythmia	Catheter ablatio	15 months of follow-up - no more episodes of palpitations or syncope.
Arun Gopi, Placid Sebastian, et al.	A rare malady with a rarer complication	19M	Heart failure	Tirofiban bolus followed by infusion for 24 h and continued on heparin.	Not available
Yuechun Shen, Xinchun Li, et al.	Myocardial Noncompaction Presenting With Myocardial Bridge	34M	Angina	Lifestyle cares, trimetazidine(improves myocardial glucose utilization through inhibition of fatty acid metabolism) Shensongyangxin. traditional Chinese medicine, containing ginseng, ophiopogon japonicus, Schisandra, and red sage root	EF improved to 77%. Other parameters from echocardiograph and electrocardiograph are not changed.
Claudia Stöllberger, Josef Finsterer et al.	Embolism from non-compaction to different vascular territories	26M	Emboli leading to stroke, renal artery occlusion and PAD	Thrombolysis and anticoagulation	Not available
Takashi Igarashi, Shinya Takase, et al.	Left ventricular non-compaction complicated by mitral valve prolapse: report of a case	31M	Heart failure	Mitral valve repair for MVP and ascending aorto-bifemoral bypass	LVEF was increased to 41.8 % post-op
Heon Lee, Seok-Yeon Kim, et al.	Isolated Non-Compaction of the Left Ventricle in a Patient with New-Onset Heart Failure: Morphologic and Functional Evaluation with Cardiac Multidetector Computed Tomography	72M	Atrial fibrillation and heart failure	Routine treatment for cardiac failure	EF improved to 40% on echocardiography

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Author	Title	Age/Gender	Presentation	Management	Follow-up
Ling Han, Jing-Gang Luo, et al.	Left Ventricular Non-compaction Combined With EpinephrineSecreted Pheochromocytoma Inducing Heart Failure	28F	Phaeochromocytoma and heart failure symptoms.	HF treatment with bed rest, sodium nitroprusside, and diuretics,	After two months, re-hospitalized, and resection of the pheochromocytoma was performed. All oral drugs were ceased after the operation. F/u echo continued to show LVNC, however, the systolic function of the myocardium was significantly improved, and LVEF was increased to 56%.
Yusuf Izzettin Alihanoglu, Ismail Dogu Kilic, et al.	Sustained ventricular tachycardia in a patient with isolated non-compaction cardiomyopathy	25F	Heart failure	Amiodarone, anticoagulant therapy and heart failure treatment with beta-blockers, ACEI, and diuretics.	None
Arun Kannan, Anindita Das, et al.	An interesting case of cryptogenic stroke in a young man due to left ventricular non-compaction: role of cardiac MRI in the accurate diagnosis	28M	Stroke	No Rx. Since the patient remained asymptomatic, he was advised to follow up with a cardiologist.	Not available
Kana Fujikura, Dennis Finkielstein, et al.	Left ventricular non-compaction and dyssynchrony	43M	Heart failure	Dobutamine was started. Digoxin, Hydralazine, Lasix, and Captopril were initiated and titrated.	His symptoms improved from NYHA class IV to III. The condition stabilized and the patient was discharged home.
Jian-Ming Li, Ting Li, et al.	An adult patient with left ventricular noncompaction detected on radionuclide myocardial perfusion imaging	51M	Ventricular tachycardia	Not available	Not available
Huan Sun, Qini Zhao, et al.	Daily 10 mg rivaroxaban as a therapy for ventricular thrombus related to left ventricular non-compaction cardiomyopathy	43M	Tthrombosis in left ventricle	Rivaroxaban and beta-blocker	Not available
Hideyuki Tanaka, Tatsunori Kimura, et al.	Aortic valve replacement for aortic regurgitation with rare left ventricular non-compaction	74F	Heart failure	Aangiotensin-converting enzyme inhibitor, diuretic, and aortic valve replacement. Discharged on an oral anticoagulant, heart failure medication and a beta-blocking agent	Remained free of cardiac events at two years when the symptoms of heart failure improved to NYHA functional class 1. The characteristic features of non-compaction remained unchanged

Humberto Morais	Aneurysm of the right sinus of Valsalva dissecting into the interventricular septum, submitral aneurysm and left ventricular non-compaction: three rare diseases in the same patient	24M	Dissecting aneurysm	Diuretics, angiotensin-converting enzyme inhibitors, and digitalis, with dramatic improvement in symptoms. He was proposed for surgery but refused and was discharged in New York Heart Association class I.	The patient died one year after the first admission.
Nikhil Kumar, Christopher A Troianos, et al.	Left Ventricular Assist Device Insertion in a Patient With Biventricular Noncompaction Cardiomyopathy, Ebstein Anomaly, and a Left Atrial Mass: A Case Report	21F	Heart failure a large LA mass.	Medical management for heart failure and the placement of an intra-aortic balloon pump -> AFib with Thrombus in the right upper pulmonary vein. The entire mass was removed - > LVAD and surgical removal of thrombus	Stable post-op
Rafał Dąbrowski, Maciej Sterliński, et al.	Cardiac resynchronization therapy in an elderly patient with left and right ventricular noncompaction	57M	Heart failure	Cardiac resynchronization therapy (CRT-D) was introduced. 1 episode of ven- tricular tachycardia terminated by a cardioverter intervention. Heart failure medication on discharge	A follow-up of 32 months have shown good response to CRT-D treatment.
Josef Finsterer, Claudia Stöllberger et al.	Left ventricular hypertrabeculation/noncompaction in an Eritrean war invalid with neuromuscular disease	52M	Heart failure	Not available	Not available
Alessio Lillia, Marco Chioccioli, et al.	Coronary microfistulae associated with non compacted myocardium: A rare cause of myocardial ischemia unraveled by a multimodality imaging approach	61M	Chest pain	Metoprolol twice-a-day and was discharged without symptoms.	18-months follow-up visit - doing well and does not report further episodes of chest pain.
Rahul D. Sawant, Leisa J. Freeman , et al.	Pregnancy and treatment outcome in a patient with left ventricular non-compaction	37F	Heart failure	Suppression of lactation with bromocriptine, together with standard heart failure management	Stable
Felix K Wegner , Gerhard-Paul Diller , et al.	a 'tiger heart' distinct variant of left ventricular non-compaction	30M	Asymptomatic,with arrhythmia	Observation	After 52 months, the patient remained stable and asymptomatic.
Gonzalo Navarrete , Eduardo Pozo ,et al.	Spongious Ischemic Myocardium	38M	Heart failure	Inotropes, diuretics, vasodilator, and coronary artery bypass surgery.	Six months after surgery, under optimal medical therapy, the patient was New York Heart Association functional class I.
Nay Aung , Filip Zemrak ,et al.	LV Noncompaction Cardiomyopathy or Just a Lot of Trabeculations?	51F	Dizziness	Conventional heart failure medical therapy and implantable cardioverter-defibrillator.	2 years later demonstrated progressive LV disease after that ICD was placed

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Author	Title	Age/Gender	Presentation	Management	Follow-up
Nay Aung , Filip Zemrak ,et al.	LV Noncompaction Cardiomyopathy or Just a Lot of Trabeculations?	32M	Palpitations	Observation	2 years later demonstrated an increase in LV internal dimensions and decline in LV systolic function
Dipesh Pradhan , Chen Liping, et al.	An unusual case of myocardial noncompaction involving both ventricles	18M	Heart failure	ACE inhibitor, aldosterone antagonist, and thiazide diuretic	Not available
Jamil Hajj-Chahine , Géraldine Allain,et al.	Aortic root replacement in a patient with left ventricular noncompaction	57F	Heart failure	Coronary artery bypass graft	6 months later, she was doing well, and the echocardiogram revealed a good functioning prosthetic valve; the left ventricle ejection fraction increased to 50%.
Michael E Pitzer , Peter H Seidenberg , et al.	Asymptomatic Left Ventricular Noncompaction — Implications for Athletic Participation	18M	asymtomatic	Observation	The patient was cleared for participation in football with close monitoring and repeat echocardiography to be completed in 1 year
Giovanni Pulignanoa , Maria Denitza , et al	Noncompaction and embolic myocardial infarction: The importance of oral anticoagulation	67F	Heart failure	IV inotropes and diuretics, implantable cardioverter-defibrillator and was discharged in NYHA class II, under standard heart failure therapy including oral anticoagulation (OAC),	2 years after stopping anticoagulation , EF fell to 15%, Coronary angiography revealed a thrombotic occlusion of the mid segment of the left anterior descending artery,death followed 25 days later
Daniele Muser , Gaetano Nucifora , et al	Clinical Spectrum of Isolated Left Ventricular Noncompaction: Thromboembolic Events, Malignant Left Ventricular Arrhythmias, and Refractory Heart Failure	17M	Stroke	Implantable cardioverter-defibrillator	Five years later, he underwent cardiac transplantation for worsening heart failure
Daniele Muser , Gaetano Nucifora , et al	Isolated noncompaction of right ventricle–a case report	19M	Heart failure	Treated with heart failure medications, anticoagulants, and drugs to control rapid ventricular rate	Not available
Margaret Kapor Manus , Satyajeet Roy , et al	Non-compaction cardiomyopathy in an asymptomatic athlete	20M	Asymptomatic	Observation	6-months later he remains asymptomatic
Bryan E-Xin Tan , Mallory Balmer-Swain , et al	Rare case of mitral annulus disjunction and non-compaction-like myocardium	74M	Heart failure	Not available	Not available
Johann Altenberger , Georg Hasenauer ,et al	Disappearance of left ventricular hypertrabeculation/ noncompaction and sudden death in a patient with Turner mosaic syndrome	5M	Acute coronary syndrome	Coronary artery bypass grafting	Death within 6 month

David C Peritz , Aaron Vaughn ,et al	Hypertrabeculation vs Left Ventricular Noncompaction on Echocardiogram A Reason to Restrict Athletic Participation	18M	Mumur	Observation	The patient returned to full athletic participation, and reevaluated annually.
Ashraf Alazzoni , Vidhya Nair , et al	noncompaction reaction	26M	Heart failure and ischemic limb	Intravenous infusion of unfractionated heparin echocardiographic assessment, the patient successfully underwent catheter-based embolectomy of the aorta and iliac arteries	On the fifth postoperative day the pateint developed ventricular fibrillation followed by pulseless electric activity, which did not respond to full and prolonged resuscitative effort
Hany Younan , Raed Alroughani , et al		29F	Stroke	Started on oral anticoagulants.	Unavailable
Madan Raj Aryal , Madan Badal , et al	Left ventricular non-compaction presenting with heart failure and intramural thrombus	28M	Heart failure	Lisinopril 5 mg daily and furosemide 40 mg, and intravenous heparin Low-dose metoprolol	3 months revealed stable LVEF of 15%. and ICD placed
Mareomi Hamada , Yoshimi Takamura , et al	Left ventricular non-compaction mimicking hypertrophic obstructive cardiomyopathy	30F	Angina	Cibenzoline and after 3 months - LV restoration surgery	After the operation, her systolic murmur and clinical symptoms disappeared. The extent of LVNC decreased and intertrabecular recesses disappeared
Costin Nicolae Ionescu , Diane Turcot	Left ventricular non-compaction and aneurysm revealed by left ventriculography	60M	Syncope	Oral anticoagulation and (ICD)	Unavailable
Alberto Cecconi , María José Olivera , et al	Epicardial lipomatous hypertrophy with ventricular septum separation and myocardial non-compaction: a new cardiomyopathy?	60F	Angina	Monitor	The patient was asymptomatic at the 6-month follow-up
Tomoaki Hirose , Tetsuji Kawata , et al	Aortic valve replacement in a patient with left ventricular noncompaction	62F	Heart failure	Medical management of heart failure and aortic valve replacement	2 years after the operation showed trivial mitral regurgitation and improvements in LV ejection fraction to 59%
Mariola Szulik , Tomasz Kukulski, et al	Long QT syndrome and left ventricular non-compaction	22F	Arhytthmia	Amiodarone, and lignocaine	Death on the 5th day
David Schibilsky , Karin Klingel ,et al	Successful Surgical Treatment of an Electrical Storm in a Patient With Atypical Non-compaction Cardiomyopathy	34M	Arhythmia	Amiodarone, sotalol, and finally ranolazine. Finally had Intraoperative epicardial ablation	Resolution after Six months
Josef Finsterer , Martina Dumser	Noncompaction in a septic heart, missed on echocardiography	88F	Sepsis	Antibiotics and pressors for sepsis	Multiorgan failure and death

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Author	Title	Age/Gender	Presentation	Management	Follow-up
Giuseppina Novo , Gregory Dendramis, et al	Left ventricular non-compaction presenting like a double-chambered left ventricle	24F	Asymptomatic	Observation	At 1 year follow up - remains asymptomatic
Zsófia Szentpáli , Tamas Szili-Torok , et al	Primary electrical disorder or primary cardiomyopathy? A case with a unique association of noncompaction cardiomyopathy and cathecolaminergic polymorphic ventricular tachycardia caused by ryanodine receptor mutation	39F	Heart failure	Dual chamber pacemaker-defibrillator implantation with further titrating of β-blocker therapy and ablation	resolution of the symptoms within 6 months
Anita Sadeghpour , Shadi Faghihi, Azin Alizadehasl	Can hypertrophic cardiomyopathy and noncompaction left ventricle coexist in a single patient?	50F	Heart failure	Unavailable	Unavailable
Sherif Moustafa David J Patton, et al	Unusual myocardial late gadolinium enhancement in isolated noncompaction cardiomyopathy	19M	Heart failure	Oral anticoagulation, in addition to the standard therapy for heart failure, based on the notion of mural LV thrombi revealed by the CMR. Cardiac resynchronization therapy/defibrillator was performed 6 months later	Unavailable
Wei-Ting Chang , Yi-Shan Tsai et al	Left ventricular non-compaction cardiomyopathy: an under-recognized disease diagnosed by echocardiography and computed tomography	38F	Heart failure	Angiotensin-converting-enzyme inhibitors, diuretics, amiodarone, and anticoagulants.	4 days later due to a right middle cerebral extensive infarction and died of intractable increased intracranial pressure
Rafał Dąbrowski, Maciej Sterliński , et al.	Cardiac resynchronization therapy in an elderly patient with left and right ventricular noncompaction	67 M	Heart failure	Cardiac resynchronization therapy and ramipril, bisoprolol, furosemide and spironolactone	Follow-up at 32 months showed good response to cardiac resynchronization therapy

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