A Noncompaction Reaction

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Information about a real patient is presented in stages (bold-face type) to an expert clinician (Dr. Omid Salehian) who responds to the information, sharing his reasoning with the reader (regular type). A discussion by the authors follows.

A previously healthy 26-year-old man presented to a community hospital with a 1-week history of malaise, dyspnea, and cough productive of clear sputum. He also had intermittent lower abdominal pain for the preceding 5 days. He was otherwise healthy and not taking any medication. He was an ex-smoker with a 7-pack-year smoking history who quit 2 years ago, rarely drank alcohol, and denied using any recreational drug. On physical examination, he was afebrile, with a blood pressure of 110/75 mm Hg and a heart rate of 144 bpm. His respiratory rate was 17 breaths per minute, with an oxygen saturation of 99% on room air. On cardiovascular examination, there was no jugular venous distension, and the carotid pulse was of normal volume and contour. On precordial examination, the apical impulse was not palpable, and no obvious heaves or thrills were felt. Auscultation revealed a loud first heart sound and a normal second heart sound. There was a third heart sound (S₃) audible over the lower sternal border and the apex. No additional sounds, rubs, or murmurs were heard. Abdominal examination revealed audible bowel sounds, with a soft and nontender abdomen and no organomegaly. There was no peripheral edema, and all peripheral pulses were palpable. Laboratory work, including complete blood count, electrolytes, and creatinine, was within the normal limits. A 12-lead ECG showed sinus tachycardia with minimal voltage criteria for left ventricular (LV) hypertrophy and no evidence of ischemia or infarction. A chest radiograph revealed cardiomegaly without any evidence of interstitial pulmonary edema or airspace disease.

Dr. Salehian: This young and previously healthy man presents with a very nonspecific symptom complex. On the basis of the history, one might wonder about a viral upper respiratory tract infection; however, it is important to maintain a very broad differential. The most concerning finding on physical examination is the resting tachycardia. Of course, determination of the underlying rhythm is the first step. Although the rhythm here is sinus tachycardia, with the resting heart rate >120 bpm, one should always consider atrial arrhythmias such as atrial flutter or atrial tachycardia as a distinct possibility. Sinus tachycardia has a broad differential; however, in this patient, one should consider hypovolemia and potential underlying sepsis high on the differential diagnosis. Additionally, the presence of S₃ and cardiomegaly on the chest radiograph raises the strong possibility of a cardiac cause of this patient’s symptom complex. A primary myocardi al process or valvular disease could account for the abnormalities detected here. Absence of murmur on physical examination should not rule out valvular dysfunction, because often, acute valvular regurgitation is not associated with a significant murmur. A transthoracic echocardiogram should be the next logical investigation, because it can assist in providing a rapid diagnosis of the possible cardiac origin.

Patient presentation (continued): A transthoracic echocardiogram was requested. While awaiting echocardiographic assessment, the patient developed acute onset of bilateral leg pain with associated numbness. Shortly thereafter, he was not able to move his toes and ankles. His feet became pale in color and cool to touch. There were no popliteal, dorsalis pedis, or posterior tibial pulses on either side either on palpation or with Doppler ultrasound. Urgent computed tomography (CT) angiography revealed occlusion of the distal aorta and iliac arteries (Figure 1).

Dr. Salehian: The constellation of symptoms of pain and paresthesia and the pallor, cool extremities, absence of pulse, and paralysis on physical examination are the cardinal manifestations of acute limb ischemia. In this patient, rapid development of bilateral lower-extremity signs points toward acute ischemia as the result of obstruction proximal to the aortic bifurcation. The role of imaging in the diagnosis of acute limb ischemia can be debated, and one has to balance the time required to perform and interpret the test against the risk of ongoing ischemia and requirement for urgent revascularization. In the majority of hospitals, CT angiography can be obtained in a timely fashion and can provide valuable information for the surgical planning. In this case, CT angiography provided both valuable anatomic information about the location of the obstruction and also offered information about the potential cause. Once the
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Diagnosis of acute aortic occlusion is made, it is important to act quickly and institute appropriate therapies. The vascular surgery team should be notified as soon as acute limb ischemia is being considered on clinical grounds so that the team can be mobilized and further investigations as needed by the team can be organized. Systemic anticoagulation with intravenous unfractionated heparin should be instituted after exclusion of aortic dissection while awaiting definitive surgical intervention. The differential diagnosis of acute aortic occlusion is fairly narrow and can be categorized as follows: Extrinsic compression (eg, trauma), disruption of the aortic wall (eg, aortic dissection), embolic phenomenon (intracardiac thrombus, valvular vegetations, or cardiac neoplasm), in situ thrombosis (eg, heparin-induced thrombocytopenia), and primary aortic neoplasms (eg, angiosarcoma). In patients in whom there has been previous aortic surgery or endovascular repair, thrombosis of the stent or graft should also be considered. Given the presence of cardiomegaly and S3 on auscultation, one should consider a cardioembolic source as the most likely possibility. Hence, proceeding with cardiac imaging after initiation of antithrombotic therapy is warranted. The timing and type of cardiac imaging depend on the urgency of the situation. Transthoracic echocardiography would be the logical initial imaging, because it is portable, widely available, and can be performed quickly even in an unstable patient at the bedside.

**Patient presentation (continued):** Intravenous infusion of unfractionated heparin was started immediately, and the patient was transferred urgently to our tertiary care facility for operative management. Given the potential for a cardioembolic source, an urgent transthoracic echocardiogram was performed in the operating room before surgery.

**Dr. Salehian:** A cardioembolic source should be considered likely given the physical examination and chest radiograph findings. The underlying process could be a dilated cardiomyopathy from acute myocarditis, valvular heart disease, or any other cause of cardiomyopathy. Cardiac tumors, valvular vegetations, or left atrial thrombi with paradoxical atrial fibrillation also need to be considered as potential embolic sources. A transthoracic echocardiogram would be useful to assess myocardial and valvular function, as well as the presence of other cardiac masses with embolic potential. However, the absence of an intracardiac mass on transthoracic echocardiography does not exclude a cardioembolic source, because there might only have been a single mass that has embolized, or the mass or its remnants could be located in chambers that are not visualized well on transthoracic echocardiography. This is typically the case for thrombi that arise from the left atrial appendage. Given this, there should be a low threshold in performing a transesophageal echocardiogram in cases in which the probability of cardioembolic source is high and the transthoracic echocardiogram is inconclusive.

**Patient presentation (continued):** The transthoracic echocardiogram demonstrated a severely dilated LV characterized by deep trabeculations in the mid and apical portions (Figure 2A and 2B; Movies I and II in the online-only Data Supplement). There was severe global hypokinesis, with a calculated ejection fraction of 13%. In addition, a mobile echogenic mass highly suggestive of thrombus was seen at the LV apex (Figure 3A and 3B; Movies II and III in the online-only Data Supplement). After echocardiographic assessment, the patient successfully underwent catheter-based embolectomy of the aorta and iliac arteries by use of a bilateral femoral approach with reperfusion to the affected vessels. Postoperatively, the patient’s hemodynamic status deteriorated, with findings suggestive of cardiogenic shock. He was transferred to the coronary care unit for vasopressor support. The patient was restarted on unfractionated heparin infusion immediately after surgery.

**Dr. Salehian:** The echocardiogram revealed dilated cardiomyopathy with evidence of deep trabeculations and deep intertrabecular recesses that resulted in a thickened myocardium,
with 2 layers consisting of compacted and noncompacted myocardium. Continuity between the LV cavity and the deep intratrabecular recesses was also noted. These findings are suggestive of LV noncompaction cardiomyopathy (LVNC), which is a rare genetic cardiomyopathy. Cardiac magnetic resonance imaging (MRI) is currently considered superior for diagnosis of LVNC and can be performed when the patient is stabilized and can be transferred safely. The presence of mobile echodensity at the LV apex and associated severe LV systolic dysfunction in the setting of acute aortic occlusion is indicative of cardiac thromboembolism as the cause of the obstruction. Systemic anticoagulation should be continued indefinitely unless there is life-threatening bleeding.

**Patient presentation (continued):** A cardiac MRI (Figure 4; Movies IV and V in the online-only Data Supplement) was performed to further clarify the nature of our patient’s cardiomyopathy. This demonstrated layers of noncompacted tissue in both ventricles, more so on the left than the right. The ratio of the noncompacted to compacted layer was 5:1, which supports the diagnosis of LVNC. The thrombus previously identified on echocardiography was quantified on MRI as 9 mm in diameter. On further inquiry, it was discovered that the patient’s sister had presented with heart failure and a right hemispheric stroke shortly after delivery of her first child at the age of 20 years and was diagnosed with peripartum cardiomyopathy.

**Dr. Salehian:** MRI findings confirm the diagnosis of LVNC suspected on transthoracic echocardiogram. Cardiac MRI is currently considered the “gold standard” for diagnosis of LVNC. A ratio of noncompacted to compacted myocardium of >2.3 in diastole on cardiac MRI supports the diagnosis of LVNC. Thromboembolism is a well-known complication of LVNC, with reports of up to one quarter of patients experiencing thromboembolic complications in follow-up.

The history of heart failure complicated by cerebrovascular embolic event in the peripartum period in the sister is very interesting. One certainly needs to consider the possibility of LVNC as the underlying cause of her heart failure. Because of the rarity of LVNC, its diagnosis can be eluded despite appropriate investigations. When heart failure is observed in familial clusters, one needs to consider genetic cardiomyopathies as a potential cause. In cases with familial clustering, a screening clinical assessment, including electrocardiographic and echocardiographic evaluation, of first-degree family members should be performed.

**Patient presentation (continued):** Over the next few days, the patient showed gradual hemodynamic improvement and was eventually weaned off vasopressor support. On the fifth postoperative day, the patient complained of sudden onset of chest heaviness and shortly afterward became acutely hypotensive, dyspneic, and diaphoretic. A 12-lead ECG (Figure I in the online-only Data Supplement) showed new anterolateral ST elevation.

**Dr. Salehian:** The constellation of symptoms and ST elevation in the setting of known LV thrombus (despite systemic
anticoagulation) points to the diagnosis of acute myocardial infarction secondary to coronary artery thromboembolism. Development of cardiogenic shock is a worrisome prognostic feature. Emergent coronary angiography with a view to percutaneous intervention is warranted in parallel with consultation with the cardiac transplantation service. Hemodynamic support during coronary angiography and percutaneous coronary intervention is problematic in this case. Intra-aortic balloon counterpulsation can be initiated readily, but the data supporting its use in myocardial infarction with cardiogenic shock are limited. The presence of LV thrombus precludes the use of the Impella percutaneous LV assist device from ABIOMED (Danvers, MA). Peripheral cardiopulmonary bypass and extracorporeal membrane oxygenation are suitable options but are associated with a number of logistical challenges that result in significant time delays before they can be initiated. Furthermore, even at sites involved in advanced cardiac care, extracorporeal membrane oxygenation support may not be routinely available.

Patient presentation (continued): Emergent coronary angiography revealed complete thrombotic occlusion of the mid to distal left anterior descending coronary artery, as well as the mid segment of a large first diagonal branch of the left anterior descending coronary artery (Figure 5A). Furthermore, the left circumflex coronary artery, which arose anomalously off the right coronary artery, was subtotally occluded in its midportion, with angiographic evidence of thrombus (Figure 5B; Movie VI in the online-only Data Supplement). The patient’s hemodynamic status continued to deteriorate despite aggressive inotropic and vasopressor support.

Dr. Salehian: The coronary angiogram confirmed the clinical suspicion of thromboembolism to the coronary arteries. With progressive cardiogenic shock in the absence of immediate access to peripheral bypass, insertion of an intra-aortic balloon pump and percutaneous revascularization with aspiration thrombectomy to restore myocardial perfusion in an attempt to stabilize the patient is indicated. If available, the transplantation and cardiovascular surgical services should be consulted urgently for possible requirement of LV assist device support.

Patient presentation (continued): Given the patient’s hemodynamic instability, he was intubated and mechanically ventilated. The cardiovascular surgery service was consulted urgently. A 40-mL intra-aortic balloon pump was inserted before proceeding with attempted revascularization. All lesions had an angiographic appearance consistent with embolization. The thrombus within the diagonal branch was aspirated successfully (Figure 6), thereby restoring flow. However, repeated attempts at aspiration of the clot within the mid left anterior descending coronary artery were unsuccessful. Despite these aggressive measures, the patient developed ventricular fibrillation followed by pulseless electric activity, which did not respond to full and prolonged resuscitative efforts.

A partial autopsy restricted to the heart was performed. The heart showed cardiomegaly, with a weight of 615 g. The LV showed marked dilatation, with numerous trabeculations and deep recesses (Figures 7). Histological sections showed an inner noncompacted layer and an outer compacted layer. Histological sections of the 3 main coronary arteries did not show any thrombus. However, occasional intramural and epicardial branches showed recent thrombus (Figure 8). Widespread contraction band necrosis of the myocytes was present.

Discussion
This case illustrates the importance of a systematic approach to patients presenting with lower-limb ischemia. It also highlights the need for timely clinical and investigational assessment of patients with acute aortic occlusion, which needs to be performed in concert with therapeutic management. Identifying the underlying cause of acute occlusion should be done in tandem with therapeutic intervention so as to minimize time delays, which are directly correlated with survival.

Figure 5. A, Coronary angiogram of the left system showing totally occluded first diagonal branch of the left anterior descending coronary artery and complete occlusion of the left anterior descending coronary artery in its mid segment (arrowhead), with no obvious collateral flow seen. B, Angiography of the right system anomalously left circumflex (from the right) subtotally occluded in its mid portion (arrow).
Acute Aortic Occlusion

Acute aortic occlusion results in a sudden decrease in lower-limb perfusion (and possibly internal organs, depending on the level of occlusion) that threatens the viability of the lower limbs and organs. It is associated with very high mortality rates of 50% to 80%, with significant morbidity in survivors.1 Acute aortic occlusion is extremely uncommon, and its exact incidence and prevalence are not known. The nature of symptoms largely depends on the level of occlusion and the ability to recruit collateral channels to provide flow around the occlusion. Symptoms usually develop over a period of minutes to hours. In patients without a history of claudication and vascular disease, collateral blood vessels cannot be recruited quickly enough to circumvent the occluded artery. These patients usually present more acutely with symptoms of pain, paresthesia, pulselessness distal to the occlusion, pallor, paralysis, and cool extremities (poikilothermia). These have been conveniently grouped into a mnemonic known as the 6 Ps.

Diagnosis of acute aortic occlusion can generally be made based on the history and physical examination, including assessment of pulses and the ankle-brachial index bilaterally. To assess the severity and level of occlusion, urgent noninvasive vascular imaging studies can be performed (eg, CT angiography or magnetic resonance angiography). Once the diagnosis of acute aortic occlusion has been made and aortic dissection has been ruled out, the patient should immediately receive an intravenous unfractionated heparin bolus followed by a continuous heparin infusion.2 Anticoagulation prevents further propagation of thrombus and inhibits thrombosis distally in the arterial and venous systems caused by low flow and stasis. It is important to start the heparin on the basis of the clinical evaluation after aortic dissection has been ruled out and not to delay it while awaiting the results of imaging studies.

Once the diagnosis of acute aortic occlusion is made, urgent vascular surgery consultation is warranted. Treatment involves endovascular interventions and open surgical revascularization. The goal of catheter-based endovascular interventions is to restore blood flow as rapidly as possible to a viable or threatened limb with the use of drugs (thrombolysis), mechanical devices, or both.2 Surgical approaches to the treatment include thromboembolectomy, bypass surgery, and adjuncts such as patch angioplasty and intraoperative thrombolysis. Restoration of a palpable foot pulse, audible arterial Doppler signals, and visible improvement of foot perfusion (eg, capillary refill, increased temperature, and sweat production) suggest treatment success. Therapeutic anticoagulation with heparin is reinstituted after the procedure.3

While managing patients with acute aortic occlusion, it is crucial to think about the possible causes. Acute aortic occlusion can be the result of extrinsic compression (eg, trauma), disruption of the wall (eg, aortic dissection), embolic phenomenon (intracardiac thrombus, valvular vegetations, or cardiac neoplasm), in situ thrombosis (eg, heparin-induced thrombocytopenia), or primary aortic neoplasms (eg, angiosarcoma). The most common pathogenesis by far is emboli, with the heart playing a major role. Common causes of cardiac embolism are LV thrombi from recent myocardial infarction or
severe cardiomyopathy, atrial fibrillation with thrombi originating in the left atrium, large valvular vegetations, or cardiac neoplasm. Therefore, careful clinical assessment with particular attention to cardiovascular examination, ECG, and a transthoracic echocardiogram (depending on the degree of suspicion) are needed as part of the workup of patients who present with an acute aortic occlusion. Emboli from thoracic aortic ulcers, as well as large aortic aneurysms, may also lead to distal aortic occlusion. Most emboli that affect the lower extremities lodge at the iliocostal or femoral bifurcation but can present with more proximal occlusion if there is focal narrowing of the aorta, as is commonly seen in patients with chronic vascular disease.

It is important to stress that clinical and investigational assessment of a patient with acute aortic occlusion should be performed in a timely manner to avoid delays to definitive management with catheter-based or surgical therapies.

LV Noncompaction
One of the rare causes of LV systolic dysfunction is LVNC, also known as spongy myocardium and LV hypertrabeculation. It is a rare primary genetic cardiomyopathy with a prevalence close to 0.01%. Before the fifth to eighth week of development, ventricular myocardium exists as a loose arrangement of muscle fibers with deep separating recesses. With maturation, this spongy myocardium is transformed into compact musculature. It is thought that LVNC occurs from an aberration in this process that results in a thickened ventricular wall composed of 2 distinct layers, an epicardial layer that is condensed tissue and a thicker endocardial layer arrested in the spongy form. This noncompacted layer is characterized by prominent trabeculae and deep intertrabecular recesses in direct communication with the ventricular cavity.

Several studies have examined the epidemiology of LVNC. The largest study in a pediatric population showed that LVNC accounts for up to 10% of all childhood cardiomyopathies, with a prevalence of 0.12 per 100,000 in the general pediatric population. Other studies have arrived at similar numbers. The prevalence of noncompaction cardiomyopathy in the adult population remains unclear, because observational studies have only been conducted in patients referred for echocardiographic assessment and are prone to significant bias. Earlier studies identified a prevalence of LVNC between 0.05% and 0.014% of all transthoracic echocardiograms, but with improved imaging and increasing awareness (which can lead to overdiagnosis), the prevalence has increased in more recent reports.

Noncompaction cardiomyopathy can be either sporadic or familial with variable inheritance patterns. Autosomal dominant inheritance is more common than X-linked or autosomal recessive inheritance. Mutations have been reported in at least 9 genes in patients with this condition (Table). LVNC is anatomically characterized by the presence of ≥3 trabeculations, because up to 70% of normal hearts have some degree of LV trabeculation. Although generally, all LV myocardial segments can be affected, the most frequently affected segments are the apex and the distal and middle segments of the inferior and lateral walls (80% of the cases). The middle and basal septum with the outflow tract are usually spared.

The major clinical manifestations of LVNC are heart failure (60%–80%), atrial and ventricular arrhythmias (including up to 30%–40% prevalence of ventricular tachycardia), and systemic cardioembolic events (up to 20%–40%). Diagnosis is usually established by echocardiography or cardiac MRI; however, CT and left ventriculography are other imaging modalities that may raise the initial clinical suspicion.

There are multiple proposed echocardiographic criteria available based on observations from different centers. The most widely accepted set of criteria, proposed by Jenni and colleagues, are the following: (1) A thickened LV wall consisting of 2 layers (a thin, compacted epicardial layer and a markedly thickened endocardial layer with numerous prominent trabeculations and deep recesses, with a maximum ratio of noncompacted to compacted myocardium ≥2:1 at end systole in the parasternal short-axis view); (2) color Doppler evidence of flow within the deep intertrabecular recesses; and (3) prominent trabecular meshwork in the LV apex or midventricular segments of the inferior and lateral wall.

A problem with all of the available criteria is that there is no gold standard other than direct tissue examination for comparison. Data about the diagnostic accuracy of each criteria are not available in the original studies, but subsequent reports suggest that the Jenni criteria confer the highest sensitivity (89%) and specificity (95%). Similarly, there are multiple cardiac MRI criteria available that, compared with echocardiography, offer improved ability to assess the myocardium to help distinguish between other forms of cardiomyopathy. The most commonly used cardiac MRI criterion is a ratio of noncompacted to compacted myocardium of ≥2.3 in diastole as assessed in 3 long-axis views.

Data on treatment of LVNC are limited, and there is no specific therapy other than those routinely used in the treatment of patients with other forms of LV systolic dysfunction. There are multiple case reports showing that patients with LVNC respond well to conventional heart failure therapies, including angiotensin-converting enzyme inhibitors, β-blockers, and diuretic therapy. In patients with advanced heart failure, use of aldosterone antagonists and consideration for cardiac resynchronization therapy in eligible patients per the current practice guidelines.
guidelines are warranted. Finally, cardiac transplantation in patients with severe forms of symptomatic LV systolic dysfunction who have undergone appropriate medical therapies should be a consideration. Intermittent therapy with intravenous β-adrenergic agents such as dobutamine or phosphodiesterase inhibitors such as milrinone may be considered in cases of end-stage heart failure with no clear mortality benefit.

Given the aforementioned risk of arrhythmias in patients with LVNC, it is reasonable to perform some form of regular rhythm monitoring (such as Holter monitoring) to detect asymptomatic arrhythmias. Despite the high rates of ventricular tachycardia, only approximately one fourth of these are sustained. The risk of life-threatening ventricular tachycardia with preserved ejection fraction is unclear, although preliminary data would suggest that the odds are low. Currently, the decision to place an implantable cardioverter-defibrillator in patients with LVNC should be in accordance with the standard indications for such therapy in those patients with nonischemic cardiomyopathy. Patients with a history of sustained ventricular tachycardia or resuscitated sudden cardiac death should receive implantable cardioverter-defibrillator therapy for secondary prevention of sudden cardiac death. Current indications for placement of an implantable cardioverter-defibrillator for primary prevention in patients with LVNC are similar to those for other causes of LV dysfunction and include the presence of LV ejection fraction <35% and New York Heart Association class II to III symptoms. Similarly, indications for placement of biventricular pacemaker for cardiac resynchronization therapy should follow the current guidelines established for other forms of cardiomyopathy.

It is estimated that the thromboembolic complications in patients with LVNC range from 10% to 37%. These events include cerebrovascular accidents, pulmonary emboli, and mesenteric infarction. There is a single case report of acute coronary syndrome thought to be secondary to thromboemboli in a 65-year-old woman with LVNC with a single

<table>
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<th>Table. Key Features of LVNC</th>
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| **Prevalence**<sup>6,10</sup> | ≈0.01% of general population  
≈3% in adult heart failure population |
| **Inheritance** | Sporadic  
Autosomal dominant more common than X-linked  
Autosomal recessive (less common) |
| **Identified genes**<sup>12–14</sup> | LIM domain binding protein 3 (LDB-3)  
α-Dystrobrevin (DTNA)  
Lamin A/C (LMNA)  
Cardiac troponin T (TNT2)  
Cardiac troponin I (TNNI3)  
α-Cardiac actin (ACTC)  
β-Myosin heavy chain (MYH7)  
Cardiac myosin binding protein C (MYBPC3)  
Tropomyosin1 (TMP1) |
| **Echocardiographic criteria**<sup>15</sup> | Ratio of noncompacted to compacted myocardium (in systole from short-axis views) >2.0  
Color Doppler evidence of deep intertrabecular recesses in direct communication with the LV  
Absence of coexisting structural cardiac abnormalities |
| **Cardiac MRI criteria**<sup>16,17</sup> | Ratio between noncompacted and compacted layers >2.3 (in end diastole)  
Trabeculated LV mass >20% of global LV mass (in end diastole) |
| **Differential diagnosis** | Apical hypertrophic cardiomyopathy  
Dilated cardiomyopathy  
Endocardial fibroelastosis  
Apical thrombus  
Hypertensive cardiomyopathy  
Tumors  
Prominent and abnormal chords |
| **Management** | As per guidelines currently established for patients with LV dysfunction with or without congestive heart failure (ACE inhibitors/ARB, β-blockers, aldosterone antagonists, digoxin)  
Device therapy (ICD and CRT) as per current guidelines for both primary and secondary prevention in patients with LV systolic dysfunction  
Anticoagulation perhaps indicated in LVNC patients with severe LV systolic dysfunction (in absence of other indications)  
VAD therapy and cardiac transplantation for patients not responding to maximal medical therapy; should follow current established guidelines |
| **Family screening** | Electrocardiographic and echocardiographic assessment of first-degree relatives |

ACE indicates angiotensin receptor enzyme; ARB, angiotensin receptor blocker; CRT, cardiac resynchronization therapy; ICD, implantable cardioverter-defibrillator; LV, left ventricle/left ventricular; LVNC, left ventricular noncompaction cardiomyopathy; MRI, magnetic resonance imaging; and VAD, ventricular assist device.
thrombus in the left anterior descending artery with otherwise normal coronary arteries.\textsuperscript{21} Although it would appear that the intertrabecular recesses could create local stasis that leads to development of thrombi, current evidence indicates that LVNC does not independently confer a higher risk of thromboemboli when one controls for the impact of LV dysfunction and arrhythmias. Hence, the routine use of long-term prophylactic anticoagulation in all patients with LVNC is controversial but may be justified in those with severe systolic dysfunction, atrial fibrillation, or a history of previous embolic events.

Once the diagnosis of LVNC is made, it is important to clinically evaluate the first-degree family members given the demonstrated genetic association, with up to 50\% of patients reporting a history of cardiomyopathy in the family. Assessment of family members should include electrocardiographic and echocardiographic evaluation; however, genetic studies are not recommended routinely given that the majority of genetic associations are based on case reports. In family screening of 45 patients with LVNC, 25\% of asymptomatic relatives had echocardiographic abnormalities, which included LVNC with or without systolic dysfunction and LV dilatation without diagnostic criteria for LVNC.\textsuperscript{22}

Most of the information about LVNC comes from case reports and a few case series. More studies are needed to assess the utility of available diagnostic tests, the role of routine genetic screening, and the efficacy of current treatment options and to explore alternative treatment plans in both symptomatic and asymptomatic individuals.

Disclosures

None.

References


KEY WORDS: cardiomyopathy, dilated, with left ventricular noncompaction • embolism
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SUPPLEMENTAL MATERIAL

Movie Legends:

Movie 1. Transthoracic echocardiogram in apical 4 chamber showing heavily trabeculated left ventricle with severe systolic dysfunction and a mobile echodensity suspicious of a thrombus attached to the apex.

Movie 2. Transthoracic echocardiogram in apical 2 chamber showing heavily trabeculated left ventricle with severe systolic dysfunction and a mobile echodensity suspicious of a thrombus attached to the apex.

Movie 3. Transthoracic echocardiogram in parasternal short axis at the apical level showing heavily trabeculated left ventricle with severe systolic dysfunction and a mobile echodensity suspicious of a thrombus.

Movie 4. Cardiac MRI (4 chamber) revealing heavily trabeculated left ventricle with severe global systolic dysfunction.

Movie 5. Cardiac MRI (short axis) revealing heavily trabeculated left ventricle with severe global systolic dysfunction. Mobile thrombus is seen at the apex.
Movie 6. Coronary angiogram of both the left and the right system revealing thrombotic occlusion of the left anterior descending and the diagonal branch as well as the anomalous left circumflex (from the right coronary artery).

Figure Legend:

Figure 1. Supplemental Figure 1. 12-lead electrocardiogram performed at the time of acute hypotension and chest pressure demonstrating anterolateral ST segment elevation.