

# Left Ventricular Noncompaction

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## Abstract

Left ventricular noncompaction (LVNC) is a cardiomyopathy characterized anatomically by deep trabeculations in the ventricular wall, which define recesses communicating with the main ventricular chamber. Major clinical correlates include systolic and diastolic dysfunction, associated at times with arrhythmias and systemic embolic events. The frequency (incidence, prevalence) is not well known; however, as the annual incidence of unclassified cardiomyopathy among children 0-10 years of age is 0.17 per 100,000 children, one can estimate the incidence of LVNC in this age group as 0.12 per 100,000. Definition and diagnostic criteria are still being debated for this recently characterized condition. Diagnosis can be made by echocardiography; current echocardiographic criteria for diagnosis typically include: presence of multiple echocardiographic trabeculations, multiple deep intertrabecular recesses communicating with the ventricular cavity, a 2-layered structure of the endomyocardium with an increased noncompacted to compacted ratio. Clinical presentation is similar to other cardiomyopathies in that it includes depressed systolic and diastolic function, systemic embolism, and tachy-arrhythmias. ECG features include biventricular hypertrophy with extreme QRS voltages, isolated or diffuse T-wave changes; Wolf-Parkinson-White with premature atrial and ventricular contractions. Medical treatment depends on the functional abnormalities (e.g., heart failure) and associated comorbidities, including systemic embolism and arrhythmia (Weiford *et al.* 2004). Aspirin therapy has been recommended for all patients to decrease the risk for systemic embolism. The mechanisms that lead to LVNC are unclear. A genetic basis in at least some cases is generically suggested. The search for genes associated with LVNC is ongoing, and there is evidence for a role of mutations of the following genes: *G4.5*, *Alpha-dystrobrevin*, *RyR2*.

## Key words

cardiomyopathy, endocardial morphogenesis, ventricular noncompaction, ventricular trabeculation

## Disease name and synonyms

- Left ventricular noncompaction - LVNC  
 - (Isolated) noncompaction of the ventricular myocardium -- (I)NVM

- Non-compaction of the left ventricular myocardium -- INLVM  
 - Left ventricular hypertrabeculation  
 - Spongy myocardium

Current usage appears to prefer the term left ventricular noncompaction (or non-compaction), though nomenclature is still debated. Some authors prefer the term noncompaction of ventricular myocardium (omitting 'left'), as the condition appears to affect both ventricles in several patients. Other authors use the term hypertrabeculation, which describes the morphology, rather than noncompaction, which suggest a specific embryologic mechanism. The prevailing preference not to use embryologic terms also has led to the declining use of the term spongy myocardium for this condition. In this text we will use the prevailing term, left ventricular noncompaction (LVNC).

In the World Health Organization's International Classification of Disease (ICD) coding system, LVNC is currently classified under 'unclassified cardiomyopathy'.

### Definition

LVNC is a cardiomyopathy characterized anatomically by deep trabeculations in the ventricular wall, which define recesses communicating with the main ventricular chamber. Major clinical correlates include systolic and diastolic dysfunction, associated at times with arrhythmias and systemic embolic events. (Weiford *et al.* 2004)

Definition and diagnostic criteria are still being debated for this recently characterized condition. Although described in case series nearly 15 years ago, (Chin *et al.* 1990) LVNC has increasingly caught the attention of the medical community in recent years. Approximately half of all articles published since 1990 (PubMed search terms, 'noncompaction' and 'non-compaction') appeared in the literature in the last 19 months (from January 2003 through August 2004, date of last review). Among these, reports from six case series provide considerable information, (Chin *et al.* 1990; Ichida *et al.* 1999; Oechslin *et al.* 2000; Pignatelli *et al.* 2003; Ritter *et al.* 1997; Stollberger *et al.* 2002) though definition and diagnostic criteria were not identical.

Some definitional issues under discussion include, for example, whether or not the presence of congenital heart anomalies should exclude a diagnosis of LVNC; whether cases diagnosed in young children vs. those diagnosed in adults represent meaningful subgroups with respect to etiology, genetics, clinical presentation (e.g., type of arrhythmia), and prognosis; and whether echocardiographic criteria (e.g., the compacted to noncompacted ratio) should differ by age at diagnosis.

### Diagnostic Criteria

Diagnosis can be made by echocardiography. Current echocardiographic criteria for diagnosis typically include the following three:

- Presence of multiple echocardiographic trabeculations, particularly in the apex and free wall of the left ventricle.
- Multiple deep intertrabecular recesses communicating with the ventricular cavity, as demonstrated by color Doppler imaging.
- A 2-layered structure of the endomyocardium with an increased noncompacted to compacted ratio (suggested as >2.0 in adults, >1.4 in children)

The proportion of ventricular wall affected (>50%) is also used in the definition by some authors (Nugent *et al.* 2003). Several authors exclude a diagnosis of LVNC when congenital heart anomalies are also present. Others exclude cases where with congenital heart anomalies include obstruction of the semilunar valves and coronary sinusoids (e.g., pulmonary atresia with intact ventricular septum). Some authors include cases with other congenital anomalies such as ventricular septal defects. (Pignatelli *et al.* 2003)

Magnetic resonance studies can be of diagnostic value, particularly when good echocardiographic images cannot be obtained. (McCrohon *et al.* 2002; Weiford *et al.* 2004). Several authors have underscored the potential for missing or misdiagnosing LVNC (see also section on differential diagnosis), and emphasized the importance of training and awareness on the part of the echocardiographer. (Stollberger and Finsterer 2004; Tamborini *et al.* 2004; Weiford *et al.* 2004)

ECG findings include signs of marked biventricular hypertrophy, with extreme QRS voltages reminiscent of those observed in [Pompe's disease](#); the presence of isolated or diffuse T-wave inversions; and arrhythmias, including Wolf-Parkinson-White syndrome with or without supraventricular tachycardia, ventricular arrhythmias, or conduction abnormalities, including heart block (Maile *et al.* 2004; Nihei *et al.* 2004; Stollberger and Finsterer 2004; Taniguchi *et al.* 2004; Weiford *et al.* 2004). Muscle biopsies and metabolic studies can be helpful when LVNC is diagnosed as part of a genetic or metabolic syndrome such as [Barth syndrome](#).

### Differential Diagnosis

Researchers have repeatedly suggested that LVNC is considerably underdiagnosed or misdiagnosed as hypertrophic or dilated cardiomyopathy. Considerations that have been cited for differential diagnosis include the following (Weiford *et al.* 2004):

- Normal heart with prominent left ventricular trabeculations: however, the normal variants have up to 3 trabeculations (Tamborini *et al.* 2004; Weiford *et al.* 2004)

- [Apical hypertrophic cardiomyopathy](#)
- [Dilated cardiomyopathy](#)
- [Arrhythmogenic right ventricular dysplasia](#)
- Endocardial fibroelastosis
- [Cardiac metastases](#)
- Left ventricular thrombus

A careful echocardiographic study inclusive of color Doppler evaluation by a trained and aware echocardiographer can in many cases provide the diagnosis. Magnetic resonance can also provide additional and at times crucial information for the diagnosis. Some authors suggest specific magnetic resonance and electrophysiologic studies to help identify people at increased risk for ventricular arrhythmias. (Weiford *et al.* 2004).

ECG and chest radiographs can complete the diagnostic picture. In children, further findings such as dysmorphic features, additional cardiac anomalies, and systemic or biochemical findings suggestive of a metabolic or genetic condition can be present (e.g., [urinary 3-methylglutaconic acid excretion](#), neutropenia in [Barth syndrome](#)).

### Etiology

The mechanisms that lead to LVNC are unclear, but it is widely suggested that the basic morphogenetic abnormality may be an arrest of normal compaction of the loose interwoven mesh of myocardial fibers in the embryo. However, there is little direct evidence that indeed this is the mechanism involved, and some authors caution against this interpretation (Bleyl *et al.* 1997).

Etiologic studies of LVNC are increasing rapidly. A genetic basis in at least some cases is generically suggested by the finding that a proportion of first-degree relatives of affected people also have a cardiomyopathy (dilated or hypertrophic). In familial cases, which account for approximately 18% to 50% of cases in published case series, the inheritance pattern varies with the majority of familial cases following an autosomal dominant pattern but with several families showing X-linked or mitochondrial transmission (Chin *et al.* 1990; Digilio *et al.* 1999; Ichida *et al.* 1999; Oechslin *et al.* 2000; Pignatelli *et al.* 2003; Ritter *et al.* 1997; Stollberger *et al.* 2002).

The search for genes associated with LVNC is ongoing, and there is evidence for a role of mutations of the following genes: (Kenton *et al.* 2004)

**G4.5:** this gene is located on Xq28 and was initially described in patients with [Barth syndrome](#), some of whom were found to have

LVNC. The gene products, called taffazins, are expressed mainly in heart and muscle cells and their action is thought to take place mainly in the mitochondria. To date mutations of *G4.5* have been reported in young boys rather than adults.

**Alpha-dystrobrevin:** this autosomal gene was identified in a Japanese family with six members affected by LVNC (Ichida *et al.* 2001)

Other candidate genes or loci that have been suggested in the literature include the following: the ryanodine receptor 2 gene (*RyR2*): mutations in this genes were reported in people with [arrhythmogenic right dysplasia](#).

**FKBP12:** this gene modulates the release of calcium from the sarcoplasmic reticulum by the ryanodine receptor 2, and deletions for *FKBP12* in mice result in animals with feature of noncompaction and congenital heart defects. (Shou *et al.* 1998)

**Lamin A/C (LMNA):** *LMNA* mutations have been reported in a case-series of patients with [dilated cardiomyopathy](#), one of whom was reported to have features of LVNC (Hermida-Prieto *et al.* 2004)

Transcription factors including NKX2.5 and TBX5 (Hatcher *et al.* 2003): animals models in which such genes are affected can present anomalies of the ventricular structures that share features of LVNC (Bruneau *et al.* 2001; Pashmforoush *et al.* 2004)

**11p15:** this locus was suggested by genome-wide linkage analysis in one family with autosomal dominant LVNC (Sasse-Klaassen *et al.* 2004). Mutation analysis of two genes in the locus (*SOX6* and muscle LIM protein [*MLP*]) failed to identify mutations (Sasse-Klaassen *et al.* 2004).

It is not yet possible to know the quantitative contribution of these genes to LVNC and such assessment awaits the publication of large systematic screens of unselected patients preferably stratified by phenotype, age, and family history. Such studies would help compare the genetic contribution among children vs. adults, in apparently isolated vs. familial cases, and by severity/clinical phenotype. Some findings however are available in recent literature and these can be summarized as follows:

- *G4.5* mutations have been identified repeatedly in some of the larger series of LVNC but accounts for a small proportion of cases. *G4.5* mutations were identified in: 2 out of 36 cases (6%) in a case-series of young affected children evaluated in Texas In a series of 36 infants and children affected children evaluated in Texas. (Pignatelli *et al.* 2003) Both infants presented the clinical and biochemical features of [Barth syndrome](#).

- One of 48 cases (2%) of isolated LVNC from a case-series also reported from Texas. (Kenton *et al.* 2004) The mutation affected a splice site acceptor site in intron 10.

- One of 27 cases of LVNC (4%) in a case series from Japan that included 14 familial cases from 10 families and 10 sporadic cases. The mutation involved 8 and was identified also in five female carriers in the family of the affected patient.

- Other instances of G4.5 mutations were found in families with different types of cardiomyopathy, including a child with LVNC and [Barth syndrome](#) in Japan. (Ichida *et al.* 2001)

A systematic screen in the case-series from Texas evaluated 48 cases of LVNC (Kenton *et al.* 2004) for mutations in the genes G4.5, *alpha-dystrobrevin*, and *FKB12*. Except for one case with mutations in G4.5, noted above, no mutations of the other two genes were identified. The authors concluded that these genes rarely contribute to isolated LVNC, at least in this population.

In conclusion, the relatively small contribution of known mutations to the disease, compared to the higher proportion of familial cases suggests that other elusive genes remain to be identified. A careful evaluation of cases of LVNC associated with structural chromosomal abnormalities could help identify further candidate loci. Of note is that at least one child with 22q11 deletion has been diagnosed with LVNC (Pignatelli *et al.* 2003)

### Clinical Description

Clinical presentation is similar to other cardiomyopathies in that it includes depressed systolic and diastolic function, systemic embolism, and tachy-arrhythmias.

The most common presentation reported in the literature has been tachypnea due to low cardiac output. (Pignatelli *et al.* 2003; Weiford *et al.* 2004) More rarely infants and young children can present with cyanosis, syncope, dysmorphic features, and failure to thrive. In a series of 36 infants and children with LVNC, 14 (39%) presented with tachypnea indicative of low cardiac output, whereas 15 (42%) were either asymptomatic or referred because of an abnormality at the ECG or chest radiography. (Pignatelli *et al.* 2003)

Based on limited data, the frequency and type of arrhythmias appear to vary by age. Among children, the more common arrhythmias include [Wolf-Parkinson-White](#) (WPW) with or without supraventricular tachycardia, as well as ventricular tachycardia. These have occasionally required radiofrequency ablation and the implantation of an intracardiac defibrillator (Pignatelli *et al.* 2003; Weiford *et al.* 2004). The rare occurrence of sudden death in reported

case series could be due to ventricular arrhythmias. Among adults, various forms of bundle block through complete atrioventricular block have been described (Taniguchi *et al.* 2004)

### Diagnostic methods

As noted above, ECG abnormalities are frequent. In many patients, ECG features include biventricular hypertrophy with extreme QRS voltages, noted to be similar to those seen in [Pompe's disease](#); isolated or diffuse T-wave changes; WPW with premature atrial and ventricular contractions. Chest radiographs can show an enlarged heart and sign of pulmonary congestion.

Echocardiography is most often the basis for diagnosis. Echocardiographic criteria that have been proposed include the presence of multiple echocardiographic trabeculations, multiple deep intertrabecular recesses communicating with the ventricular cavity, involvement of the apical or middle portion of the ventricle, and a 2-layered structure of the endocardium with an increased noncompacted to compacted ratio (>1.4 in children according to some, >2.0 in adults according to others). The use of color Doppler helps identify these key features. The importance of awareness and training on the part of the echocardiographer has been repeatedly noted.

Magnetic resonance studies also have a role in diagnosis, and can provide convincing evidence in cases where the echocardiographic findings are uncertain. (McCrohon *et al.* 2002)

Genetic testing for known mutations can provide additional data for counseling and research. Findings consistent with Barth syndrome include 3-methylgluconic aciduria and neutropenia. Skeletal biopsy can provide evidence for inclusions and myopathic changes.

### Frequency and epidemiology

The frequency (incidence, prevalence) is not well known and there has been a scarcity of population-based studies of LVNC. A retrospective cohort study of cardiomyopathy in Australia identified 314 of pediatric cardiomyopathy, of which 29 (9.2%) had a diagnosis of LVNP. (Nugent *et al.* 2003) These 29 cases represented 69% of 'unclassified' cardiomyopathy in the study. From this study one can speculate as follows:

Because the annual incidence of unclassified cardiomyopathy in the study among children 0-10 years of age was 0.17 per 100,000 children, one can estimate the incidence of LVNC in this age group as 0.12 per 100,000 (or 1 in 850,000 children 0-10 years of age per calendar year)

The annual incidence of unclassified cardiomyopathy was highest among children under 1 year of age (1.18 per 100,000 children under 1 year of age). If LVNC accounted in this age group for the same overall proportion of cases, one could estimate an annual incidence among infants of 0.81 per 100,000, or 1 in 123,000 infants per calendar year.

Other recent epidemiologic studies of cardiomyopathies did not report separately cases of LVNC (Arola *et al.* 1997; Lipshultz *et al.* 2003) so no further data on incidence is currently available. The observed prevalence will likely increase with the improvement of cardiac imaging and the increased recognition of this condition by echocardiographers. Many authors now suggest that LVNC has a higher prevalence and more varied prognosis than was originally thought (McCrohon *et al.* 2002; Weiford *et al.* 2004)

With respect to other epidemiologic characteristics, several case-series and review of the literature (Pignatelli *et al.* 2003; Weiford *et al.* 2004) show a slight excess of males, with a male proportion ranging from 56% to 82%. This is taken as evidence for a contribution to LVNC of X-linked genes, including G4.5.

#### Management including treatment

Medical treatment depends on the functional abnormalities (e.g., heart failure) and associated comorbidities, including systemic embolism and arrhythmia (Weiford *et al.* 2004). Aspirin therapy has been recommended for all patients to decrease the risk for systemic embolism.

Some recommended treatments for selected clinical presentations include:

decreased ventricular systolic function: afterload reduction and/or beta-blocker

hypertrophic phenotype, no depressed ventricular function: beta-blocker or calcium blocker

mitochondrial abnormalities: some groups recommend use of thiamine, coenzyme Q10, riboflavin, and carnitine in selected cases (Pignatelli *et al.* 2003)

Several authors have remarked on the 'undulating' clinical course of some patients, in which the clinical and functional findings improve and then worsen over time (Weiford *et al.* 2004). Cardiomyopathy can worsen to the point that several children have required cardiac transplantation. (Pignatelli *et al.* 2003)

#### Unresolved questions

Controversies exist about diagnostic criteria, nomenclature, prognosis, origin, pathogenesis, and the necessity to classify LVNC as a distinct entity and cardiomyopathy by the World Health Organization.

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