

Left ventricular non-compaction

An introduction to LVNC

- LVNC is a condition where the muscular wall of the left ventricle of the heart appears 'spongy'.
- It can affect the heart's ability to work efficiently as a pump, and affect the electrical signalling of the heart.
- Although it cannot be cured, treatment is available for people who experience symptoms.

What is left ventricular non-compaction (LVNC)?

LVNC is a condition of the heart where the walls of the left ventricle (the bottom chamber of the left side of the heart) are non-compacted. This causes channels to form in the heart muscle, called trabeculations. This gives the left ventricle a characteristic 'spongy' look (a bit like honeycomb). Although it usually affects the left ventricle, it can also affect the right ventricle.

LVNC is described by the American Heart Association as a type of genetic cardiomyopathy (heart muscle disease caused by a change in the person's genetic make-up). However, WHO (World Health Organisation) describes it as an 'unclassified' type of cardiomyopathy. It may also be associated with dilated or hypertrophic cardiomyopathy. There is currently no universally agreed diagnostic criteria for the condition.

How does LVNC happen?

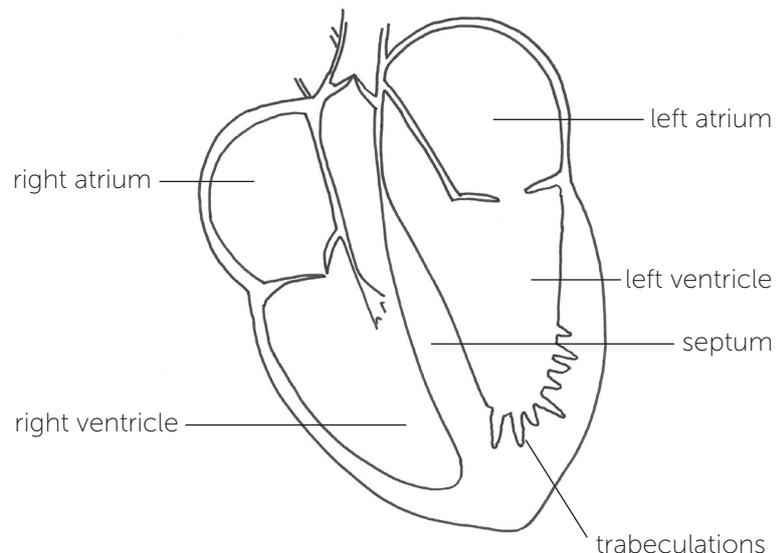
LVNC is a condition that usually develops before birth, when a baby is still an embryo, developing in its mother's womb. This means that a person is usually born with the condition. However, some people may develop the condition after birth (and then it is called 'acquired' LVNC).

The heart is made up of a thick layer of muscle cells (called the myocardium). In the developed heart, the muscle cells in the myocardium are tightly bound together and 'compacted'. When an embryo's heart develops, the fibres of heart muscles start by being 'non-compacted' before compressing to form the thick muscular wall. LVNC happens when this part of the normal development of the heart (called the 'remodelling phase') is interrupted. This interruption means that part of the myocardium does not compact but remains non-compacted.

What causes it?

LVNC is often a genetic condition (caused by an altered or 'mutated' gene). Where it is genetic, it can be inherited (passed from parent to child) and therefore can run in families. For this reason, screening of the first degree family members (parents, siblings and children) of someone with LVNC is recommended, to see if they also have the condition.

The structure of a heart with LVNC (as if you were looking in a mirror).



Who gets LVNC?

How common LVNC is, is not really known, although it appears to affect more men than women. More people are diagnosed with LVNC nowadays (compared to previously) because cardiac imaging is more advanced now (see 'how is it diagnosed'), and the areas of non-compaction can be seen and diagnosed.

LVNC is sometimes diagnosed when someone has dilated or hypertrophic cardiomyopathy and they have imaging as part of the diagnosis or regular monitoring of their condition. The imaging may show that they have LVNC as well.

Although it may be diagnosed at any age, many people are not diagnosed until later in life, when they start to develop symptoms. Some people may have undiagnosed LVNC even if they don't have any symptoms.

What are the symptoms of LVNC?

Not everyone with LVNC has symptoms (some people never have any symptoms). Symptoms vary from one person to another depending on the amount and position of the non-compaction, and they are not specific to LVNC (they can happen with other types of cardiomyopathy, and other conditions, which can make diagnosis difficult).



Symptoms usually happen either because:

- the non-compactation reduces how well the heart can pump blood around the body (which can cause symptoms of heart failure as the heart fails to meet the needs of the body) or
- the trabeculations affect the normal electrical signalling of the heart (which can cause arrhythmias – heart rhythms disturbances).

Symptoms of LVNC include:

- breathlessness;
- fatigue (extreme tiredness);
- feeling dizzy or light-headed;
- fainting or passing out (syncope);
- feeling of abnormal heartbeats (palpitations); and
- swelling of the legs, ankles and feet.

How is it diagnosed?

If someone has a family member with cardiomyopathy or LVNC, or they have symptoms that suggest they may have a heart condition, they may be referred for various tests. This may include the following.

- Medical history and family tree – to look at symptoms and whether other family members have this condition (which can be genetic);
- Physical exam – to see what physical symptoms, if any, are experienced.
- ECG (electrocardiogram) – to look at the conduction of electrical impulses through the heart and see if arrhythmias are happening.
- Echo (echocardiogram) with colour Doppler and MRI (magnetic resonance imaging) scan – types of imaging and show the structure of the heart. They can also show the movement of blood as it flows through the heart, and can be used to see where the trabeculations are.

LVNC is usually diagnosed by comparing the areas of non-compacted heart muscle to the compacted muscle, and the amount of trabeculations.

How is LVNC treated and managed?

Although LVNC cannot be cured, treatment can control any symptoms caused by a reduction in how well the heart is working as a pump, or by problems with the electrical signalling in the heart. Treatment aims to help to improve the heart's function and to reduce any complications. Treatment is guided by the results of the diagnostic tests and the symptoms, and so is individualised to the needs of the individual and the symptoms they are experiencing.

Treatment may include any of the following.

- **Anticoagulants** – these reduce the risk of blood clots forming in the trabeculations which can lead to a stroke. Blood clots can also form due to atrial fibrillation (an abnormal heart rhythm). An example of an anti-coagulant is warfarin, which may also be prescribed when someone has reduced function in the left ventricle.
- **Treatment for heart failure** – where LVNC causes symptoms of heart failure, treatment will include:
- **diuretics** (water tablets) – to reduce the build-up of any fluid on the lungs or the ankles by encouraging the kidneys get rid of water as urine;
- **ACE inhibitors** (angiotensin-converting enzyme inhibitors) – these relax the smooth muscle around the blood vessels to reduce the workload on the heart, and reduce the volume of the blood, making it easier for the heart to work;
- **Angiotensin II Receptor Blockers** (ARBs) – these dilate (enlarge) the blood vessels which helps to reduce blood pressure. They may be used if the person is not able to tolerate ACE inhibitors; and
- **beta-blockers** – these reduce the rate and force of the heart's contraction, by reducing stimulation of adrenalin (which would normally make the heart beat faster).
- **ICDs** (implantable cardioverter defibrillator) – this may be an option if the person experiences, or is at risk of, arrhythmias. ICDs detect and correct any dangerous arrhythmias which could otherwise lead to a cardiac arrest.
- **Pacemaker** – these take over the electrical signalling of the heart and restore a normal heart rhythm. This is important for people with arrhythmias such as heart block, which makes the heart rate slow down.
- **Treatment for other types of cardiomyopathy** – if someone has LVNC alongside another type of cardiomyopathy, they may need additional treatment.

If someone does not have any symptoms, they will not usually require any treatment.

Although there is no agreed criteria for diagnosing LVNC, and still lots of 'unknowns' about the condition, research is ongoing to understand more.

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