



Left Ventricular Non-Compaction Cardiomyopathy (LVNC)

CONTACT US

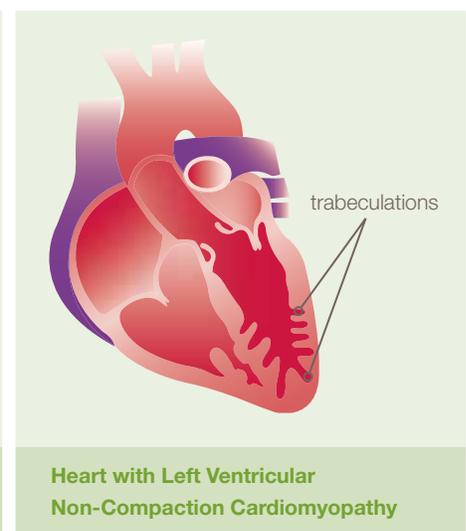
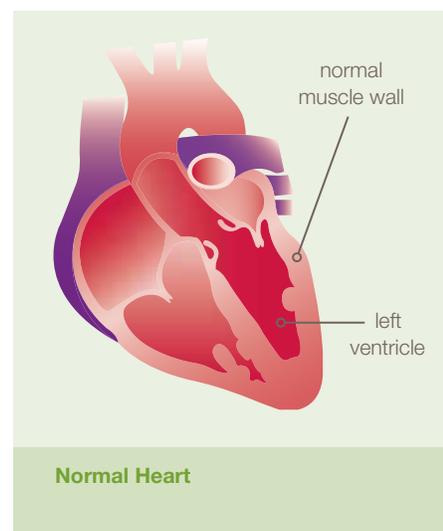
To learn more about treatment of Left Ventricular Non-Compaction Cardiomyopathy at the Heart Institute at Cincinnati Children's Hospital Medical Center, call **513-803-1746**.

Definition

In left ventricular non-compaction cardiomyopathy (LVNC) the lower left chamber of the heart, called the left ventricle, contains bundles or pieces of muscle that extend into the chamber. These pieces of muscles are called trabeculations. During development, the heart muscle is a sponge-like network of muscle fibers.

As normal development progresses, the trabeculations become compacted transforming the heart muscle from sponge-like to smooth and solid. LVNC occurs when compaction does not occur. These trabeculations typically occur at the bottom of the heart called the apex but can be seen anywhere in the left ventricle. Individuals with LVNC may also have another type of heart muscle disease (hypertrophic cardiomyopathy, dilated cardiomyopathy or restrictive cardiomyopathy).

LVNC is being diagnosed more frequently in both children and adults secondary to increased awareness of the condition. The initial diagnosis may be made at any age and many patients often go undiagnosed until later in life.



Symptoms

Many individuals with LVNC experience no symptoms. For those who have heart failure or an abnormal heart rhythm, symptoms can include:

Older children and adults

- Shortness of breath and fatigue
- Feeling dizzy or light-headed
- Fainting or passing out (syncope)
- Feeling abnormal heart beats (palpitations)
- Unexplained weight gain or swelling

Newborns and babies

- Excessive sweating with feeds or activity
- Difficulty feeding or poor growth

Although some people with LVNC experience no symptoms, they may still be at risk for complications associated with the diagnosis. The most serious complication of LVNC is sudden cardiac arrest. Sudden cardiac arrest is a sudden loss of heart function caused by a dangerously fast heart rhythm called ventricular tachycardia. Unless emergency treatments, including CPR and defibrillation, are initiated immediately after the onset of symptoms, sudden cardiac death can occur.

Most people with LVNC have a low risk for sudden cardiac arrest. However, it is important for anyone at risk to be followed by a cardiologist with experience in caring for individuals with cardiomyopathy.

Causes

LVNC is largely a genetic condition. When an individual is diagnosed with LVNC, there is a 20 to 40 percent chance that an underlying genetic cause will be identified. Most commonly, the genetic cause results in a condition that runs in a family and affects only the heart. It is possible that parents, siblings, and even aunts/uncles or cousins of an individual diagnosed with LVNC may also have this heart muscle condition. If they haven't had cardiac screening, family members may not know they are affected.

There are other genetic causes of LVNC which can be associated with other health problems. These include genetic syndromes, metabolic, and mitochondrial disorders. LVNC can also be seen in association with congenital heart disease (structural change to the heart). There are not any known acquired causes of LVNC.

Diagnosis

The diagnosis of LVNC is based on cardiac testing, family history, medical history, and physical exam.

- **Cardiac testing:** An echocardiogram is the most common test used to diagnose LVNC, as the trabeculations within the left ventricle and overall squeeze of the heart can be measured with this test.
- **Family history:** A family tree should be constructed with specific attention to cardiomyopathy, rhythm problems, sudden cardiac or unexplained death, cardiac surgery or presence of other cardiac disease in relatives.
- **Medical history:** A history of heart failure symptoms including feeding issues, heart rhythm problems and exercise intolerance.

- **Physical exam:** A thorough physical exam will be done to evaluate for signs of a cardiac problem. Assessment of skeletal muscle strength will be performed since problems with skeletal muscle may occur with cardiac muscle disease.

Management and Treatment

The treatment for individuals with LVNC is focused on treating associated types of heart muscle disease such as a thick heart muscle (hypertrophic cardiomyopathy) or poorly squeezing heart muscle. The primary goal is to improve cardiac function and to prevent symptoms. Medications are used to treat symptoms, reverse heart damage and to prevent ongoing damage to the heart muscle. If an individual has decreased heart function, they are at an increased risk for a clot to form in between the trabeculations. A blood thinner, such as aspirin, may be recommended for some individuals to prevent blood clots forming. The formation of a clot places an individual at risk for stroke or other organ damage that can occur if a clot leaves the heart. Individuals with LVNC need to be followed by a cardiologist on a regular basis.

Lifestyle changes are recommended for some individuals. Exercise is generally important, however, individuals with LVNC may experience fatigue and shortness of breath with exercise. It's important for these individuals to limit their activity if they become tired, dizzy or have difficulty breathing. In some cases, participating in competitive or endurance sports may place an individual at increased risk for heart rhythm problems and may be discouraged.

Some people with LVNC are at increased risk for sudden cardiac arrest. For these individuals, an implantable cardioverter-defibrillator (ICD) may be recommended. In some patients with LVNC, a pacemaker (cardiac resynchronization therapy) may be combined with the ICD. Mechanical pacing from different areas of the heart may improve the heart's ability to supply blood to the body.

If cardiac function continues to worsen despite medications, a mechanical device may be necessary to help the heart pump and supply blood to the body. The device that would be used for young children would be the Berlin heart device. For older children and adults there are multiple devices that could be surgically placed to provide cardiac support.

In some cases, if the condition is severe, a heart transplant may be considered.

Screening for Family Members

All first degree relatives of an individual who has LVNC should undergo routine cardiac evaluation. This includes parents, brothers, sisters, and children. If a gene mutation causing LVNC in an individual is known, screening is recommended for those family members proven to also carry the gene mutation. Routine cardiac screening would not be recommended for family members who did not inherit the gene mutation.

The timing of routine cardiac screening for family members should be discussed with a cardiologist and genetics professional who have experience in caring for individuals with cardiomyopathy. How frequently an individual should be evaluated is based on published guidelines and the individual's age, personal medical and family history.

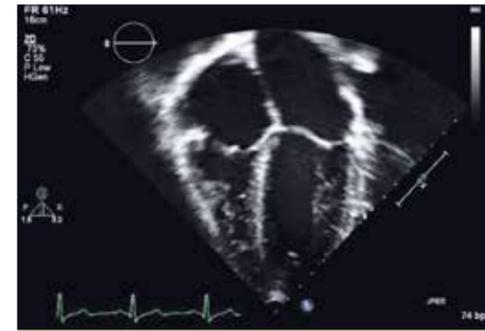


Fig 1.1

Normal Heart

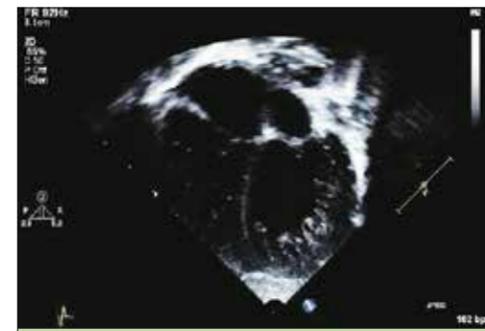


Fig 1.2

Heart with Left Ventricular Non-Compaction Cardiomyopathy

RESOURCES FOR FAMILIES

The Cardiomyopathy Association:

www.cardiomyopathy.org

Children's Cardiomyopathy Foundation:

www.childrenscardiomyopathy.org



Cincinnati Children's is ranked #8 in Cardiology and Heart Surgery, and third among all Honor Roll hospitals in the 2012–13 *U.S. News & World Report* listing of Best Children's Hospitals.

Genetic Testing

Genes are packages of genetic information that tell our bodies how to develop and function. LVNC can be caused by a change or mutation in a gene that encodes proteins important for the contraction or squeeze of the heart muscle. A parent that carries a gene mutation will have a 50 percent chance of passing it on to each child.

Genetic testing is available to look for mutations in the genes known to cause LVNC. This type of testing is done on a blood sample. It is best to begin genetic testing in a family with an individual who is known to have cardiomyopathy. If a gene mutation is found in someone with LVNC, genetic testing for the same mutation can be offered to close family members. Finding a gene mutation in other family members will help identify family members who may be at risk to develop heart muscle disease and who should have cardiac screening.

A genetic counselor and/or geneticist can assess the family history and provide information about the chance that there is a genetic predisposition to cardiomyopathy. A genetics professional can also explain and facilitate genetic testing and interpret results for patients and families.

For immediate consultations, physician referrals and patient care follow-up, call the National Physician Priority Link at **888-987-7997**.

www.cincinnatichildrens.org