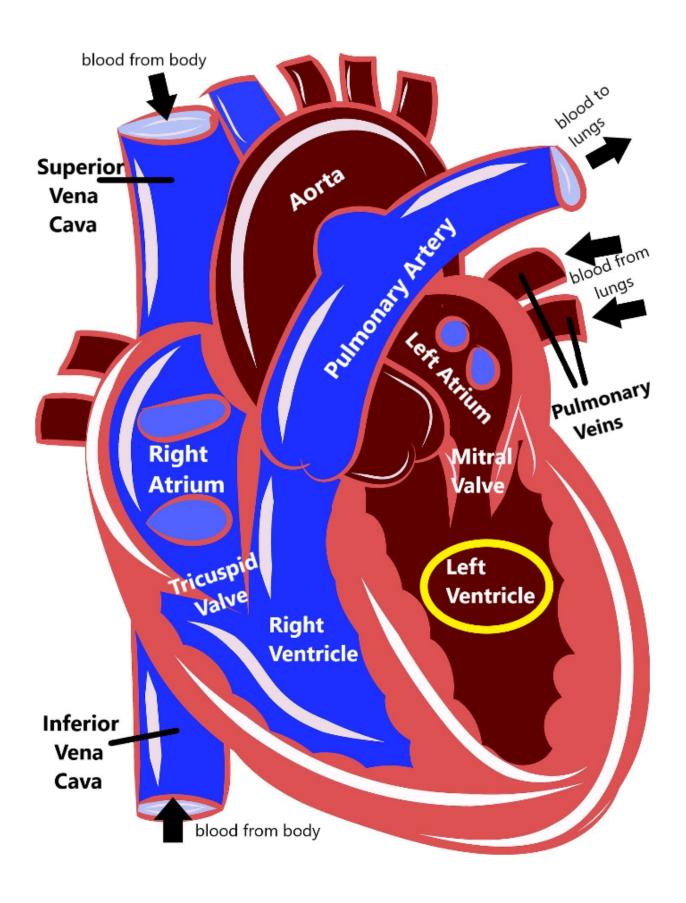


The muscles in our heart are usually smooth and firm, which is how they most efficiently pump blood around our bodies. Left ventricular noncompaction (LVNC) is a heart condition that causes the muscle of one of the bottom chambers of the heart (the left ventricle) to not develop correctly.







Instead of having smooth and firm muscle, the left ventricle in people with LVNC has muscle that is spongy, weak, and thick. This muscle cannot contract and relax how heart muscles normally should, so it is not able to pump blood as well out of the left ventricle into the rest of the body.

Some people who have LVNC may experience signs and symptoms, including:

- Blood clots
- A heart arrhythmia (abnormal heartbeat)
- <u>Heart palpitations</u>
- Extreme fatigue, particularly during exercise
- Shortness of breath, dizziness, and fainting
- Lymphedema

Other people who have LVNC may have no symptoms at all, but could be at a higher risk for sudden cardiac death. Although some individuals with LVNC may not have significant health issues, about two-thirds will go on to develop heart failure.

Causes

Only about 20-40% of cases of LVNC have a underlying genetic cause that can currently be found. Some cases of LVNC may be the result of other underlying health conditions (such as some metabolic or mitochondrial genetic disorders), but a cause cannot be identified for many people with LVNC.

There are likely many genes that can cause or increase the chance for someone to develop LVNC, but the ones that are currently most commonly tested for are MYH7, MYBPC3, SCN5A, TPM1, ACTC1, LMNA, and TNNT2.

It is estimated that approximately 8 to 12 of every one million people have LVNC. It is possible that it is more common than this, and that people who are affected that haven't shown any symptoms just don't know that they have it.

Diagnosing LVNC

While genetic testing for LVNC can be helpful to establish a diagnosis, medical providers may also use other medical tests, such as an echocardiogram or your family history. Red flags in the family history that may increase the chance for LVNC include people with:



- cardiomyopathies
- arrhythmias
- sudden or unexplained death
- feeding issues in babies and young children
- extreme fatigue during exercise (exercise intolerance)

Many families may have these red flags in their family history and DO NOT have LVNC. However, someone with a strong pattern of these or other heart issues may be at a higher chance to have LVNC, and may benefit from talking about it more with a specialist, such as a cardiologist or a genetic counselor.

Medical Management for LVNC

Treatment for LVNC can sometimes vary depending on the individual person and their specific health concerns. Certain medications, such as aspirin to help prevent blood clots, and implantable devices, such as a pacemaker or implantable cardiac defibrillator may be part of the treatment plan. Some patients with LVNC may need a heart transplant. It also may be recommended that people with LVNC avoid high-intensity or endurance sports, and to monitor their breathing and energy levels during routine exercise.

Click <u>here</u> to learn more about scheduling a genetic counseling appointment for questions about pediatric or adult genetic conditions.

Additional Resources

Children's Cardiomyopathy Foundation

Heart Failure Society of America (HFSA)