Brugada syndrome is a genetic condition that interferes with the heart's normal electrical rhythm (called an <u>arrhythmia</u>) in the lower part of the heart (the ventricles).



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Most people with Brugada syndrome do not have symptoms until adulthood, although in rare cases they can occur as early as infancy. These symptoms include fainting, difficulty breathing, seizures, and sudden death, and often happen when the person is resting or asleep.

The risk for sudden death from Brugada syndrome is highest at around age 40, although it is also suspected to play a part in some instances of Sudden Infant Death Syndrome (SIDS), which is a significant cause of death in children under a year old. Brugada syndrome was also at one time called Sudden Unexplained Nocturnal Death Syndrome (SUNDS).

Causes

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Brugada syndrome can be inherited/genetic, or can be something that someone develops because of something they were exposed to (acquired). The acquired form of Brugada syndrome has been linked primarily to certain medications, having abnormally high levels of calcium, or having abnormally high or low levels of potassium.

Although there are approximately 23 genes that have been suspected to cause hereditary Brugada syndrome, harmful changes (called pathogenic variants) in the <u>SCN5A</u> gene are the most common cause that we know of thus far (causing 15-30% of all cases of Brugada syndrome).

Brugada syndrome is thought to affect approximately 1 in 2000 (0.05%) people. 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. Some studies have estimated that Brugada syndrome is the cause for 4-12% of all unexpected sudden deaths, and for up to 20% of sudden deaths in people who don't have any obvious heart problems.

Most people with hereditary Brugada syndrome have a parent who also has it, but 1% of people are the first ones in their family to have it (called *de novo*).

Diagnosing Brugada syndrome

While genetic testing for Brugada syndrome can be helpful to establish a diagnosis, medical providers may also use other medical tests, such as an <u>ECG</u>, an <u>echocardiogram</u>, or your <u>family history</u>. Some red flags in the family history that may increase the chance for Brugada syndrome include people with:

• Repeated fainting or passing out

- Ventricular fibrillation: when the lower chambers of the heart (ventricles) quiver instead of beating like normal
- Self-terminating polymorphic ventricular tachycardia: when the ventricles begin to beat too fast, but then fix themselves
- <u>Cardiac arrest</u> (sudden heart failure)
- Sudden cardiac death

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

Medical Management for Brugada syndrome

Treatment for Brugada syndrome can sometimes vary depending on the individual person and their specific health concerns, and should be discussed with a medical provider who is familiar with Brugada syndrome. Some options for medical management can include medication, regular monitoring with an <u>ECG</u>, and a <u>surgically-implantable defibrillator</u>.

People who have Brugada syndrome should also be careful to avoid certain medications (such as antipsychotic or antidepressant drugs that block sodium, or certain drugs to treat <u>arrhythmias</u>), anesthesia, and high fevers, when possible.

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

Additional Resources

Sudden Arrhythmia Death Syndromes (SADS) Foundation

Arrhythmia Alliance