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Brugada syndrome : An introducion

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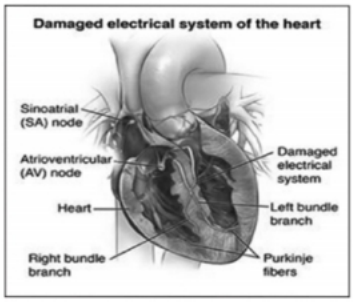
Brugada syndrome is a genetic disorder that can cause a dangerous irregular heartbeat. When this happens, the lower chambers of your heart (ventricles) beat fast and irregularly. This prevents blood from circulating correctly in your body. This can be dangerous and may lead to fainting or even death, especially during sleep or rest.

The disease has been called sudden, unexplained nocturnal death syndrome because people with it can often die in their sleep.

Definition

A cardiac disorder characterized on electrocardiogram (ECG) by ST segment elevation with a coved aspect on the right precordial leads, and a clinical susceptibility to ventricular tachyarrhythmias and sudden death occurring in the absence of overt myocardial abnormalities.



Incidence

Brugada syndrome is rare. It affects about 5 of every 10,000 people worldwide. Symptoms often start during adulthood. But the disorder can develop at any age, including infancy. The average age of death related to the disease is 40 year old.

Etiology

- The first genetic association with Brugada syndrome discovered was a loss-of-function mutation in the cardiac voltage-gated sodium channel gene SCN5A. It is thought to be found in 15-30% of Brugada syndrome cases.
- Mutations in calcium and potassium channels, associated channel proteins, and desmosomal proteins have also been linked with the disease.
- Brugada syndrome is inherited in an autosomal dominant pattern. However, affected individuals may demonstrate variable expressivity and reduced penetrance.
- Additionally, many environmental and genetic factors may influence the phenotype, including temperature, medications, electrolyte abnormalities, and cocaine.

Brugada syndrome may be caused by:

- Structural abnormality in your heart, which may be hard to detect.
- An imbalance in chemicals that help send electrical signals through your body (electrolytes).
- Use of certain prescription medications or cocaine

Risk factors

Family history of Brugada syndrome: This condition is often passed down through families (inherited). If other family members have had Brugada syndrome, you're at an increased risk of having it, too.

Being male: Men are more frequently diagnosed with Brugada syndrome than women (male hormone testosterone may contribute to the difference between genders).

Race: Brugada syndrome occurs more frequently in Asians than in other races.

Fever: A fever doesn't cause Brugada syndrome, but it can irritate the heart and trigger fainting or sudden cardiac arrest in someone with Brugada syndrome, especially in children.

Complications

Sudden cardiac arrest: If not treated immediately, this sudden loss of heart function, breathing and consciousness, which often occurs while sleeping, is fatal. With fast, appropriate medical care, survival is possible.

Fainting: If you have Brugada syndrome and you faint, get emergency medical attention.

Sudden death is the most serious complication of Brugada syndrome.

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This often occurs while the person is sleeping.

Symptoms of brugada syndrome

Many people with brugada syndrome do not have any symptoms and do not realize they have it.

Some people experience:-

Fits (Seizures)

Palpitations

Fainting

Sudden death or cardiac arrest

Chest pain, breathlessness or dizziness

Rapid heartbeat

Difficulty breathing (especially at night)

Cardiac arrest (heart failure) may be the first sign of Brugada syndrome

You can get symptoms at any time, but they are sometimes triggered by something such as a high temperature, drinking lots of alcohol, or dehydration.

Diagnosis

Genetic (DNA) testing: Your doctor will analyze the sequence of the SCN5A gene for any mutations. Mutations in this gene often cause the genetic version of Brugada syndrome.

If the diagnosis is unclear or if you are found to have the Brugada ECG pattern but have no symptoms, you may have a test called a "drug challenge." This is done in a hospital setting with professional supervision. You are given a medicine that blocks sodium channels. Then several ECGs are done to look for changes in your ECG.

Electrocardiogram (ECG): This test measures the electrical currents in your heart and displays them as wavy lines on a screen. Doctors examine the display to look for any irregularities. Brugada syndrome ECG results have identifiable patterns.

Medication-Assisted ECG: Your doctor may give you medication to make it easier for him or her to identify Brugada syndrome on the ECG screen.

Electrophysiology (EP) testing : Your doctor inserts catheters into your heart through veins in your legs (femoral veins) to measure the electrical activity of your heart. This is a rare test reserved for unclear diagnoses.

Echocardiogram: An echocardiogram uses sound waves to create images of your heart. This test cannot diagnose Brugada syndrome on its own, but it can help your doctor rule out any structural problems with your heart.

Lab tests : Your doctor looks at potassium and calcium levels to diagnose Brugada syndrome and to rule out any other potential conditions.

Diagnostic criteria

Major criteria

1. Presence of ECG marker in structurally normal hearts.
2. Appearence of ECG marker after administration of Na+ channel blockers.

Minor criteria

1. Family history of sudden cardiac death.
2. Syncope of unknown origin.
3. Documented ventricular tachycardia/ fibrillation
4. Genetic mutation of ion channels.

Diagnostic criteria based on ECG

Type 1

- Coved ST segment elevation >2mm in >1 of V1-V3 followed by a negative T wave.
- This is the only ECG abnormality that is potentially diagnostic.
- It is often referred to as Brugada sign.

Type 2

Brugada Type 2 has >2mm of saddleback shaped ST elevation.

Type 3

Brugada type 3 can be the morphology of either type 1 or type 2, but with <2mm of ST segment elevation.

Treatment

Brugada syndrome treatment depends on your risk of a serious abnormal heartbeat.

You're considered at high risk if you have:

- A personal history of serious heart rhythm problems
- Fainting spells
- Survived sudden cardiac arrest

If you don't have any symptoms, you may not need any specific treatment because your risk is likely low. However, your doctor will recommend the following steps to reduce your chances of an abnormal heart rhythm.

Treat fever aggressively: Fever is a known trigger of abnormal heartbeats in people with Brugada syndrome, so use fever-reducing medications at the first sign of fever.

Avoiding drugs that may trigger an abnormal heart rhythm: Many drugs can increase the risk of an irregular heartbeat, including certain heart medications and antidepressants. Too much alcohol