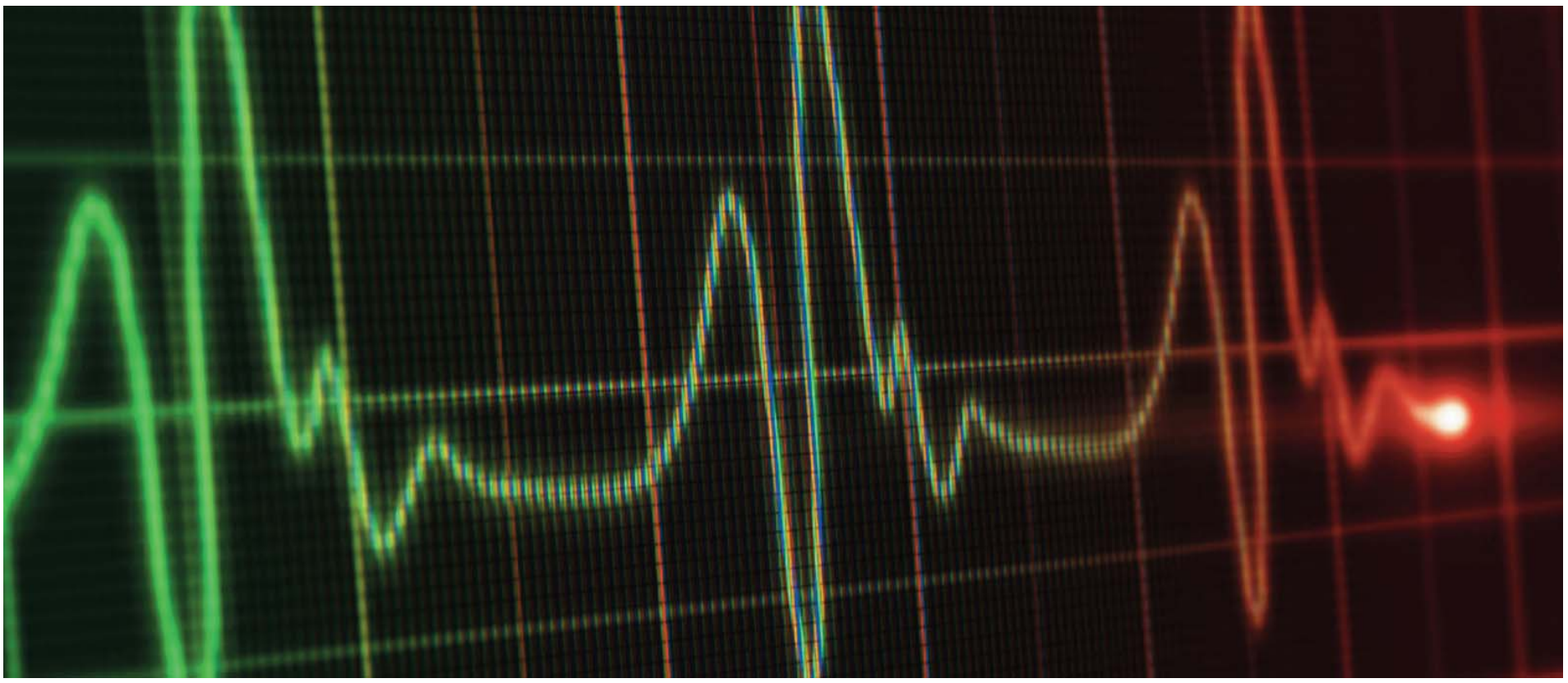

Brugada syndrome – The silent killer

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Brugada syndrome is a rare and a possibly life-threatening inherited disease that puts patients at risk of developing abnormal and irregular heartbeats in the lower heart chambers (ventricles). This abnormality is referred to as ventricular arrhythmia. The onset of Brugada syndrome usually manifests during adulthood, although it can develop at any time throughout life. Signs and symptoms liked to arrhythmias involving sudden death could present from early infancy to late adulthood. Sudden death typically occurs approximately at the age of 40. Brugada syndrome could explain some instances related to sudden infant death (SIDS), which is the greatest cause of death in infants younger than one year. Characteristics of SIDS include a sudden and unexpected death which occurs during sleep.

A sudden unexplained nocturnal death syndrome (SUNDS) is a condition defined by unexpected cardiac arrest among young individuals which usually presents at night during sleep. This condition was primarily described in southeast Asian populations, where it happens to be the greatest cause of death. Additionally, it was established that SUNDS, together with the Brugada syndrome, are the same medical disorder.

The cause of Brugada syndrome is a mutation in one of various genes. The most prevalent gene mutation related to this condition is *SCN5A,* which is affected in approximately 30% of individuals. Some of the remaining genes contributing to Brugada syndrome give out instructions which eventually lead to production of proteins that ensure the appropriate location or function of sodium channels within the heart muscle. Ion channels such as sodium, calcium and potassium channels are of extreme importance as they help with maintenance of a regular heartbeat.

In affected individuals in whom the gene identification was unsuccessful, the cause of Brugada syndrome remains unknown. In some cases, certain medication can lead to a nongenetic (acquired) form of the disorder. Drugs which can cause an abnormal heart rhythm involve medication indented for treatment of arrhythmias, angina (chest pain), high blood pressure, depression as well as other mental illnesses. Abnormally raised levels of calcium (hypercalcemia), potassium (hyperkalaemia) and low potassium levels (hypokalaemia) have also been linked to Brugada syndrome.

Inheritance

The inheritance of Brugada syndrome is autosomal dominant, meaning only one copy of the affected gene within each cell is sufficient to cause the disorder. With regards to majority of cases an affected individual would have one parent with the condition. Other cases may be caused by appearance of new mutation in the gene. These cases present in population with no family history of Brugada syndrome.

Frequency

Brugada syndrome is estimated to affect two in 10,000 individuals worldwide, yet the exact prevalence remains unknown. This condition appears much more frequent among individuals of Asian ancestry, especially in Japanese and South Asian populations. Brugada syndrome affects both males and females however it is eight to 10 times more frequent in men. It is suspected that high testosterone levels may be accountable for this difference.

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Symptoms of Brugada syndrome

Many individuals with Brugada syndrome do not show any symptoms therefore they are not aware of it. Symptoms contributing to the syndrome include blackouts, fits (seizures), occasional noticeable heartbeats (palpitations), chest pain, shortness of breath as well as dizziness.

Triggers

It is possible to decrease the risk of developing a fast heartbeat by avoiding things which could be a trigger including high temperatures, considerable alcohol consumption, dehydration as well as certain medication (such as sodium blocking drugs).

Tests for Brugada syndrome

As with other inherited arrhythmias, Brugada syndrome could be challenging to diagnose. Electrocardiogram (ECG) is the main test for Brugada syndrome. It measures heart's electrical activity and is usually performed within clinical setting. During ECG small sensors are attached to the chest together with upper and lower limbs. They are linked to a machine which measures electrical signals delivered by the heart with each beat. There are several different conditions which can mimic Brugada pattern on an ECG, and because the implications of the diagnosis, these conditions must be ruled out. In case of an individual fainting without an unidentifiable reason (especially a younger, healthy person) that person should be checked by a cardiac electrophysiologist. Moreover, genetic testing can be performed in order to look for one of the faulty genes which cause Brugada syndrome.

Treatment

There is no cure for Brugada syndrome yet doctors commonly recommend either an electrophysiology (EP) study for condition monitoring or a special device which gets implemented near the shoulder called an implantable cardioverter defibrillator (ICD) which can avoid an arrhythmic attack.

EP study is a one/two-hour procedure which allows the doctor to assess the electrical system of the heart using special sets of catheters. The EP study is used to identify the abnormal rhythm and determine the effect of certain medication.

ICD is an implantable device; extremely small and is progressively decreasing in size with each innovation. An ICD can detect and present the type of arrhythmias which lead to fainting and are potentially fatal. If arrhythmia gets sensed by ICD an energy is sent to the heart in order to "shock" it back to the normal (sinus) rhythm.

Living with Brugada

Brugada is a serious condition that some people die from. The change of this happening is significantly reduced if it is diagnosed and treated. The first line of treatment suggested for individuals living with Brugada syndrome is lifestyle change. People should be informed and educated in order to recognise and eliminate triggers which can lead to arrhythmias. These involve reduction of alcohol consumption, avoiding certain medication and instant treatment of fever. Even though, the abnormal heart rhythms caused by Brugada syndrome are more likely to happen at rest or even during sleep, some individuals experience arrhythmias while performing excessive exercise. Therefore, it is advised that people with Brugada syndrome practise gentle exercise and avoid strenuous activities. Most people can carry on everyday activities such as working out, having sexual intercourse, getting pregnant, having children and driving.

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