Brugada Syndrome

Brugada syndrome (also known as **Sudden unexpected nocturnal death syndrome**, **Sudden unexplained death syndrome**, or **SUDS** is a condition that causes a disruption of the heart's normal **rhythm**. Specifically, this disorder can lead to uncoordinated electrical activity in the heart's lower chambers (ventricles), an abnormality called **ventricular arrhythmia**. If untreated, the irregular heartbeats can cause fainting (syncope), seizures, difficulty breathing, or sudden death. These complications typically occur when an affected person is resting or asleep.

Signs and Symptoms

Brugada syndrome usually becomes apparent in **adulthood**, although signs and symptoms, including sudden death, can occur any time from early infancy to old age. The mean age of sudden death is approximately 40 years. This condition may explain some cases of sudden infant death syndrome (SIDS), which is a major cause of death in babies younger than one year. It is characterized by sudden and unexplained death, usually during sleep. Sudden unexplained nocturnal death syndrome (SUNDS) is a condition characterized by unexpected cardiac arrest in young adults, usually at night during sleep. This condition was originally described in **Southeast Asian populations**, where it is a major cause of death. Researchers have determined that SUNDS and Brugada syndrome are the same disorder.

Causes

Mutations in the SCN5A gene have been identified in fewer than one-third of people with Brugada syndrome. This gene provides instructions for making a sodium channel, which normally transports positively charged sodium atoms (ions) into heart muscle cells. This type of ion channel plays a critical role in maintaining the heart's normal rhythm. Mutations in the SCN5A gene alter the structure or function of the channel, which reduces the flow of sodium ions into cells. A disruption in ion transport alters the way the heart beats, leading to the abnormal heart rhythm characteristic of Brugada syndrome. In affected people without an identified SCN5A mutation, the cause of Brugada syndrome is often unknown. In some cases, certain drugs may cause a nongenetic (acquired) form of the disorder. Drugs that can induce an altered heart rhythm include medications used to treat some forms of arrhythmia, a condition called angina (which causes chest pain), high blood pressure, depression, and other mental illnesses. Abnormally high blood levels of calcium (hypercalcemia) or potassium (hyperkalemia), as well as unusually low potassium levels (hypokalemia), also have been associated with acquired Brugada syndrome. In addition to causing a nongenetic form of this disorder, these factors may trigger symptoms in people with an underlying SCN5A mutation.

Chances of Developing Brugada Syndrome

Although Brugada syndrome affects both men and women, the condition appears to be 8 to 10 times more common in **men**. Researchers suspect that testosterone, a sex hormone present at much higher levels in men, may be responsible for this difference.

Genetics

Mutations in the SCN5A gene cause Brugada syndrome.

Heredity

This condition is inherited in an autosomal dominant pattern, which means one copy of the altered gene in each cell is sufficient to cause the disorder. In most cases, an affected person has one parent with the condition. Other cases may result from new mutations in the gene. These cases occur in people with no history of the disorder in their family.

Epidemiology

The exact prevalence of Brugada syndrome is unknown, although it is estimated to affect 5 in 10,000 people worldwide. This condition occurs much more frequently in people of **Asian ancestry**, particularly in Japanese and Southeast Asian populations.

External links

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