Brugada syndrome

Information for families

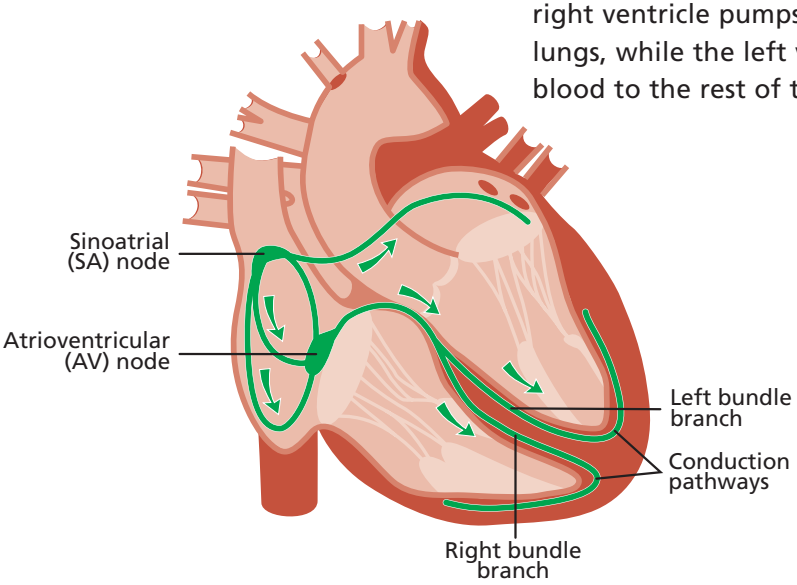
Great Ormond Street Hospital for Children
NHS Foundation Trust

This information sheet from Great Ormond Street Hospital (GOSH) explains about the medical condition Brugada syndrome, what causes it and where to get help.

The Normal Heart

The heart is a special kind of muscle which acts as a pump to keep blood moving around the body. The pumping action of the heart muscle is triggered by electrical impulses which pass through the walls of the heart, causing them to contract.

A specialised area of heart tissue called the sinoatrial (SA) node is the starting point for each electrical signal through the heart. The electrical impulse travels through the walls of the top chambers of the heart (the left and right atria), causing them to contract and squeeze blood downwards into the bottom chambers of the heart (the left and right ventricles). The impulse stops briefly at the atrioventricular node (AV node), before passing into the walls of the ventricles. As it moves through the ventricles, it causes them to contract and pump blood out of the heart. The right ventricle pumps blood to the lungs, while the left ventricle pumps blood to the rest of the body.



What is Brugada syndrome?

Brugada syndrome is an inherited condition caused by a change in a person's DNA. People with Brugada syndrome have changes in the microscopic structure of individual heart muscle cells – these changes affect the way that electrical impulses are able to pass through the heart. The condition can put affected individuals at increased risk of developing an abnormal heart rhythm, known as an arrhythmia. This means that the electrical signals passing through the ventricles may become dangerously fast (ventricular tachycardia) or may become disorganised and lose their usual pattern (ventricular fibrillation). These arrhythmias make the heart muscle contract in an abnormal, ineffective way which means that it is unable pump blood out very effectively. If left untreated, these arrhythmias can cause loss of consciousness, and may even be fatal.

What are the signs and symptoms of Brugada syndrome?

People with Brugada syndrome do not always show symptoms so it can remain undiagnosed. If someone does have symptoms, they are likely to include fainting spells (syncope) or heart palpitations (flutters). Unfortunately, in some cases, Brugada syndrome may cause sudden death in undiagnosed individuals. It is therefore important that the condition is detected early so that doctors can identify and treat those patients who have a higher risk of developing dangerous heart rhythms.

Patients with a diagnosis of Brugada syndrome should be reviewed by a specialist cardiologist, who can establish whether or not treatment is required. Some people may be referred for cardiac screening following diagnosis of the condition in a close relative – these people are sometimes diagnosed incidentally with Brugada syndrome, although they may not have had any symptoms themselves.

How is Brugada syndrome diagnosed?

An electrocardiogram (ECG) is one of the main tests for Brugada syndrome. It measures electrical activity within the heart through sticky sensor pads put on your child's chest. The results are displayed on a screen and printed on a thin strip of paper.

Some people with Brugada syndrome have a very typical pattern on their ECG at rest, and this is sufficient to make a diagnosis. In other individuals, an ECG may not show the typical abnormalities, and a different test called an ajmaline provocation test might be suggested. This test uses a medicine called ajmaline (which is introduced via a drip in the patient's arm) to provoke the typical ECG changes. In patients who do not have Brugada syndrome, ajmaline will not have any significant effect on the ECG. The ajmaline test is widely used by specialist cardiologists around the world to help diagnose Brugada syndrome in adults and teenagers. It is a very safe test and complications are rare. Further information about the ajmaline provocation test is available in a separate information sheet.

How is Brugada syndrome treated?

Although Brugada syndrome can be dangerous for some individuals, most children and adults with this condition will not need any treatment and have a low risk of developing abnormal and dangerous heart rhythms. However there are some important measures that affected patients need to take to reduce their risk of developing arrhythmias.

There are certain medicines that can increase the risk of abnormal heart rhythms in people who have Brugada syndrome. Cardiologists will therefore advise patients to avoid these drugs. Your family doctor (GP) should be made aware that there are certain medications that should not be prescribed for patients with Brugada syndrome. A regularly updated list of these medicines can be found online (www.brugadadrugs.org). Different medications are widely available, so this should not mean that any other health problems will need to go untreated.

A high temperature or fever can cause typical ECG changes in patients with Brugada syndrome. Therefore, affected individuals are advised to control fevers very carefully by taking regular paracetamol and ibuprofen. If a fever persists despite these measures, people with Brugada syndrome should attend their local hospital as a precautionary measure and have an ECG.

In individuals with cardiac symptoms or when the doctor thinks they have a higher risk of arrhythmia, treatment with a device called an implantable cardioverter-defibrillator (ICD) may be recommended. An ICD comprises a small generator about the size of a matchbox, connected to some thin wires. Both the generator and wires are inside the body so they are not visible. The generator box sits just beneath the collarbone and it is implanted while the patient is under a general anaesthetic. The ends of the wires sit inside the heart and monitor the heart rhythm constantly. If the ICD detects a dangerous heart rhythm, it can automatically deliver a pulse of electricity to 'reset' the heart and restore the normal pattern of electrical impulses. The vast majority of patients with Brugada syndrome do not require an ICD – they are only recommended when a patient is considered to be at high risk of developing a dangerous heart rhythm.

What happens next?

Children and young people diagnosed with Brugada syndrome will need regular life-long monitoring. As Brugada syndrome is an inherited condition, immediate family members of any affected individual will require clinical screening at a specialist cardiology centre.

It is not always possible to use genetic testing to diagnose Brugada syndrome, because we do not know all of the gene changes which can cause the condition. Clinical screening with ECGs and ajmaline testing are the recommended means of diagnosis.

Further information and support

If you have any questions or would like some more information, please contact the Clinical Nurse Specialists at Great Ormond Street Hospital on 020 7405 9200 ext.5124, 5139 or 5305. You can also email the service at icvd@gosh.nhs.uk

- **SADS UK** offers support and advice about heart conditions that can lead to sudden unexpected death. Telephone them on 01277 811 215 or visit their website at www.sadsuk.org
- **CRY – Cardiac Risk in the Young** offer support and advice to families affected by young sudden cardiac death. Telephone them on 01737 636 222 or visit their website at www.c-r-y.org.uk

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