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Brugada syndrome

The heart has its own electrical conduction system. The conduction system sends signals through the heart to make it beat in a regular rhythm. Sometimes a conduction pathway can be damaged or blocked, which can cause the heart rhythm to change. The heart may beat too quickly (tachycardia), too slowly (bradycardia) or irregularly which may affect the heart's ability to pump blood around the body.

What is Brugada syndrome?

Brugada syndrome is a rare inherited heart rhythm disorder in which the heart is structurally normal, but patients may be at risk of developing a fast heart rhythm due to changes within the ion channels of the heart. Brugada syndrome restricts the flow of sodium ions into the cells of the heart. These ion channels alter the chemical balance of cardiac cells, by adjusting the amount of electrical charge to them. Therefore, if the electrical properties of a cell are faulty this can result in a disturbance of the heart rhythm (arrhythmia).

Signs and symptoms that could mean you have Brugada syndrome include fainting (syncope), irregular heartbeats, fast and chaotic heartbeats and rarely, sudden cardiac arrest.

Some genes for Brugada syndrome have been identified but the list is not complete. It is therefore impossible to be sure that a patient does not have Brugada syndrome even if a genetic screening, with a blood test or mouth swab, is negative.

If your doctor suspects that you may have Brugada syndrome they will advise you to have a simple test known as the ajmaline (or flecainide) challenge to confirm your diagnosis.

What is a Ajmaline challenge?

Ajmaline is a drug which blocks sodium channels.

As it blocks the faulty sodium channels it unmasks ECG changes in patients with Brugada syndrome.

Your doctor will administer the drug through a vein in your hand and record your ECG. The ECG will record how your heart reacts to the ajmaline allowing the doctor to collect detailed information about the cause of your potential arrhythmia.

Risks of the procedure

The Ajmaline challenge is safe, but as with any procedure there are potential risks that are very rare, can be treated and are rarely life threatening.

It is common to experience a metallic taste in your mouth during the procedure, and visual disturbance such as double vision may also occur. These side effects usually resolve once the procedure is complete. Very rarely the ajmaline can cause your heart to go into a very fast rhythm; a cardioversion is required to correct this. A cardioversion is a well-established and effective treatment for fast heart rhythms. Before you are cardioverted you will be given a short-acting sedative. Once you are asleep a special machine called a defibrillator is used to restore the normal rhythm and rate.

There is a very low risk that following the procedure you will develop an arrhythmia. In this instance you will be required to remain in hospital overnight to monitor your heart rhythm. It is important that for the duration of the procedure, if you feel any palpitations, dizziness, or uncomfortable symptoms you should inform your nurse or doctor.

Before the procedure

You may be asked to have nothing to eat or drink for a period of time before the test. On arrival, you will be introduced to the nurse and the doctor who will be looking after you. The procedure will be explained to you, and if you have any worries or



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questions please do not be afraid to ask. Before the procedure, you will have blood taken and an ECG recorded. It is important for you to tell your nurse or doctor if you have any allergies or have had a previous reaction to drugs or other tests.

A small needle will be inserted into a vein in your hand to allow the doctor to give you the ajmaline during the procedure. You will also wear a hospital gown to make it easier to record the ECG.

During the procedure

During the procedure, you will be awake and able to talk. Your doctor and nurse will stay with you throughout the test and there will be equipment by your bedside which is used to monitor your heart rhythm and record your blood pressure.

For the duration of the challenge you will be connected to the ECG machine. The nurse or doctor will connect you to the ajmaline infusion which may sting a little. Once the infusion is running, your doctor will record the ECG at three minute intervals. After the test, you will be kept in the monitored bed for approximately one hour.

After the procedure

Following the procedure your heart rhythm will be monitored for several hours. Your blood pressure and pulse will be checked and the small needle in your hand will be removed. Your doctor will discuss the results with your consultant.

What treatment options are available to me?

If the test result is negative, your doctor will consider your individual risk, and advise you if further tests are necessary. It is likely that you will be able to go home the same day. However, it is important to ask a friend or family member to collect you and drive you home. It is also recommended that you have someone with you for the rest of the day after the test.

If the test is positive, and you are at risk of a

fast heart rhythm developing, your doctor may suggest an electrophysiology study and ultimately you may be advised to have an implantable cardioverter defibrillator (ICD) fitted. An ICD will not prevent the arrhythmia but can treat it when one happens. If the test result is positive, it is likely that you will be advised to remain in hospital until after these further tests.

Following your discharge from hospital you will be able to resume your normal daily activities, including returning to work.

What about my family and relatives?

As noted above, genetic testing is at a very early stage for the diagnosis of inherited heart rhythm disorders. However, you and your family may be offered genetic testing if Brugada syndrome is diagnosed in a relative. If the faulty gene is discovered in the suspected case, and then also found in the relative, there is a risk that the relative could suffer symptoms because of Brugada syndrome. If the genetic test is negative, it may not rule out a genetic heart rhythm disorder. Your genetics consultant/counsellor will be able to give more extensive advice.

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