

Drug provocation challenge

A patient's guide

Why have I been asked to have a drug provocation challenge? This is a specialised test carried out to look for rare heart rhythm disturbances which can be inherited. Some heart rhythm disturbances can cause dangerous heart rhythms and in rare cases cause sudden death.

Your doctor may suspect that you have an inherited heart rhythm disturbance if:

- A member of your family is known to have a heart rhythm disturbance.
- You have been found to have some changes on your resting ECG (heart tracing) You have suffered unexplained blackouts, dizzy spells or heart rhythm disturbances.
- A member of your family suffered an unexplained sudden death.

If you are unsure of the reason you have been asked to have this test, please ask your cardiologist or call the arrhythmia specialist nurse helpline (the number is on page 5 of this leaflet)

What is an inherited heart rhythm disturbance?

An inherited heart rhythm disturbance is a condition which affects the electrical activity in the heart muscle cells, without affecting the hearts structure.

Sometimes this is called an ion channelopathy. An ion is a chemical substance such as sodium or potassium which forms the basis of the movement of the electricity through the heart muscle

An ion channel is the route that the ions take in and out of the heart muscle, they regulate the flow of electrical charge.

There are several different types of channelopathies including:

- Long QT syndrome
- Brugada syndrome
- Catecholaminergic polymorphic ventricular tachycardia (CPVT)
- Short QT syndrome

They are usually inherited from your parents, although sometimes can occur in one family member.

How is the condition inherited?

An inherited heart rhythm disturbance occurs when there is a fault in one of the genes which determines how the heart muscle cells work, this can cause the electrical function of the heart to become abnormal.

The gene fault is passed from one generation to the next by what is known as an autosomal dominant inheritance pattern.

This means that the children of someone with an inherited heart rhythm disturbance each have up to a 50% (or one in two) chance of inheriting the faulty gene.

The gene fault responsible is inherited by both men and women and doesn't skip a generation.

Most people who have inherited the gene fault do not have any symptoms so it can sometimes be difficult to spot the inheritance pattern in a family.

These inherited heart rhythm conditions are not fully understood and not all the genes responsible have been identified, therefore it is not always possible to offer genetic testing.

What problems can an inherited heart rhythm condition cause?

The majority of people who have an inherited heart rhythm condition do not experience any heart rhythm problems and feel perfectly well. However, they may be at risk of developing fast heart rhythms without any warning. The fast heart rhythms can cause the heart muscle cells to contract so quickly and chaotically that the heart is no longer able to pump blood around the body effectively. This can result in a temporary blackout or, very rarely, sudden death.

What are the benefits of having a drug provocation challenge?

A drug provocation challenge can help to determine if you have an inherited heart rhythm disturbance. This will help your cardiologist to provide advice about your risk of developing heart rhythm disturbances. You will then be able to discuss the treatment options for you and possibly other family members. The purpose of any treatment is to prevent sudden death from a heart rhythm disturbance.

What are the risks of having drug provocation challenge?

Complications from a correctly performed challenge are extremely rare. Drug provocation challenges are usually performed in a specialist cardiac centre where doctors and nurses experienced in performing the test and correctly interpreting the ECGs are available.

These rare complications include:

- A heart rhythm disturbance which may need to be

treated urgently with a defibrillator that shocks the heart back into the correct rhythm.

- An allergic reaction to the drug but this is rare.
- If you are found to have an inherited heart rhythm disturbance, this can have implications for life and health insurance and employment.

Is there any special preparation?

- The risks will be discussed with you before you decide to have the drug provocation challenge. You will have the opportunity to ask questions and when you feel you understand the risks and benefits, you will be asked to sign a consent form stating that you understand the procedure. We recommend that you read this leaflet thoroughly and make a note of anything you would like to ask.
- You must not have anything to eat or drink after 6am on the day of the test.

What happens on the day of the drug provocation challenge?

- The test is done on the day ward at Papworth Hospital and you may need to be there for up to eight hours.
- The drugs we use are ajmaline, flecainide or adrenaline. This will depend on which particular condition we are looking for. This will be explained on the day of the test.
- On arrival the test will be explained by a doctor or specialist nurse and you will be asked to sign a consent form to confirm that you understand the benefits and risks of the test and the possible implications of the result.
- You will be weighed, your blood pressure and an ECG will be recorded and a blood sample taken to test for potassium levels. A small tube called a cannula will be inserted into a vein in your arm which is used to deliver the drug solution.
- Once we know the blood test result, we will connect you to a continuous heart monitor and to the ECG machine to allow ECGs to be recorded at intervals during the test. These monitor the electrical activity in your heart during and after the test.
- The drug is delivered into your vein sometimes via a pump and this takes approximately 15 minutes. Some people experience a metallic taste or a warm flush during the test.
- You need to stay in the hospital for a minimum of 30 minutes and sometimes up to four hours after the drug challenge has finished or until any changes on your ECG return to normal.

How do I get the results of my drug provocation challenge?

The person performing the test will discuss the result with your cardiologist and you too will be able to discuss the result with a cardiologist or specialist nurse before you leave hospital.

They will also discuss screening of other family members if required. You will be given a letter summarising your test results and any future treatment. This will also be copied to your GP.

Who can I speak to if I would like more information or have any questions?

The Arrhythmia Specialist Nurses helpline at Papworth Hospital on 01480 364947.

or

The Cardiac Genetic Specialist Nurse at Papworth Hospital on 01480 364361.

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For more information about Papworth Hospital please visit our website www.papworthhospital.nhs.uk

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