

# Inherited Arrhythmias and Genetic Testing



Working together to improve the diagnosis, treatment and quality of life for all those affected by arrhythmias

# Glossary

**Inherited Cardiac Conditions (ICCs)** Conditions that can be passed down in families via your genes

**Electrocardiogram (ECG)** is a simple test that records the heart's rhythm and rate

**Long QT Syndrome (LQTS)** An inherited condition where there are problems with the electrical activity of the heart

**Brugada Syndrome** A rare inherited heart rhythm disorder, in which the patient is at a risk of developing a fast heart rhythm

**Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)** A rare inherited arrhythmia caused by an abnormality of ion channels, leading to abnormal heartbeats

**Wolff-Parkinson-White Syndrome (WPW)** An inherited condition caused by the presence of an extra electrical pathway

## Important Information

This information leaflet has been produced for people who have been diagnosed with an Inherited Cardiac Condition and their families. Its aim is to outline the process and implications of genetic testing and to give you an idea of what to expect from your genetic counselling appointment.

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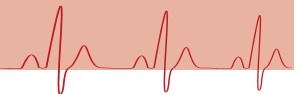
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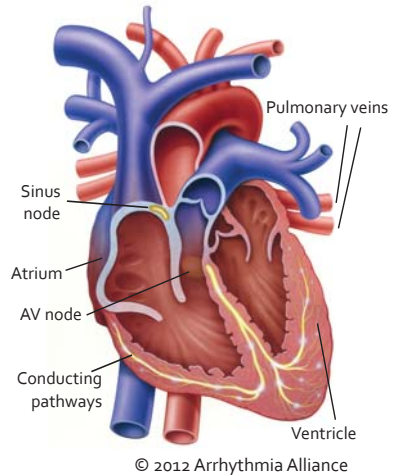


# What is an inherited cardiac condition (ICC)?

Inherited Cardiac Conditions (ICCs), for example Long QT Syndrome (LQTS) and Brugada Syndrome, are conditions that can be passed down in families via your genes. Genes act as instructions telling our bodies how to grow, develop and function. There are many genes known to be involved with ICCs. A change, or 'spelling mistake', in one of these genes can cause someone to be affected by, or be at risk of, an ICC.

The way these 'spelling mistakes' are inherited can vary, but there is most commonly a 50:50 or one in two chance that a close blood relative (child, parent, sister or brother) also has the 'spelling mistake' themselves, and therefore has the risk of developing the ICC too. Most people with ICCs do not suffer troublesome ongoing symptoms. However a small number of people are at risk of dangerous heart problems and it is important to have regular heart checks, as many of these problems are treatable or preventable.

## The heart and normal conduction



# What is genetic testing?

A genetic test is a type of medical test that is used to try and identify the gene change, or 'spelling mistake' that can cause an ICC. For example:

A correctly 'spelt' gene:

THE CAT SAT MAT

A gene with a 'spelling mistake':

THE CATTSA TMA T

Within the genes that are associated with causing ICCs, there are a large number of different gene changes that can cause the condition. It is not always possible, using current technology, to find the 'spelling mistake'. If this is the case, your sample may be kept and stored as it may be possible to do additional testing in the future. Even if the gene change causing the condition in your family is found, your sample may still be stored for further testing if and when medical knowledge increases and new technologies become available.

You may be asked for your written consent before you give a sample, and your permission will also be gained before any results are shared for the purposes of helping other family members who are also considering genetic testing.

## Are there different types of genetic tests?

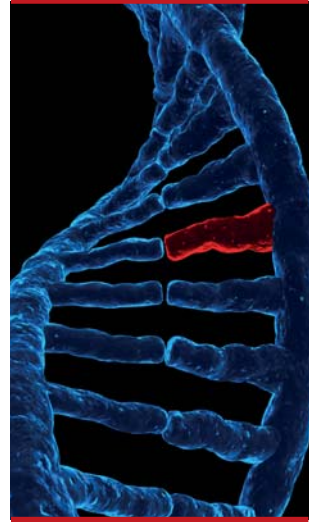
There are two different types of genetic tests: 'genetic screening' and 'pre-symptomatic' or 'familial' testing. The first affected person in the family to come forward for testing will be offered a genetic screen; if a causative spelling mistake is found, then their relatives may be offered a familial test. The process involved and implications to consider are quite different for each type of test.

# Genetic screening

Genetic screening can be offered to someone who has been diagnosed with an ICC by their specialist heart doctor (cardiologist) based on the results of clinical tests (like ECG and echocardiography) and examination.

The aim of genetic screening is to try and identify the genetic cause of your ICC. The results of a genetic screen may not change anything for you in terms of your diagnosis and management - these decisions are usually based on the results of clinical tests and examination.

However, if the causative spelling mistake is found, it can be a very useful and efficient tool for finding out who else in your family may be at risk of developing the same ICC as you.



There are several possible results from 'genetic screening':

- 1) A genetic 'spelling mistake' is found which is believed to be responsible for causing your ICC. This allows for pre-symptomatic genetic testing of family members to be offered.
- 2) The responsible genetic 'spelling mistake' is not found. This does not mean you do not have the ICC you were clinically diagnosed with, or that it is not an inherited condition, just that the specific responsible genetic 'spelling mistake' in you has not been identified.

This may be because the testing procedure was unable to identify your particular gene alteration or because the particular gene alteration is in genes that the laboratory does not test, or have not been discovered yet. In this situation, pre-symptomatic genetic testing cannot be offered to other family members. However, usual practice is to store your sample, in case additional testing becomes possible in the future.

- 3) A genetic 'spelling mistake' is found, but it is not yet certain whether this is what is causing your ICC. In this situation, further genetic tests and clinical investigations of your family may be needed to learn more about this gene change.

# Pre-symptomatic genetic testing

If an ICC-causing gene change is found in a family member who has already been diagnosed with an ICC (see genetic screening section), pre-symptomatic genetic testing can be offered to blood relatives who are not already known to have the ICC. In the first instance this would usually mean offering testing to close relatives such as parents, brothers, sisters and children, depending on their age.

If a relative chooses to have pre-symptomatic testing, there are two possible outcomes:

- 1) The causative gene change is found. That person is at increased risk of developing the ICC and should have their heart checked by a cardiologist. If they have children, they too would be at risk of the familial ICC and genetic testing and/or screening would be recommended, depending on their age.
- 2) The causative gene change is not found. That person is not at increased risk of developing the ICC and does not need heart checks. If they have children, then they do not need genetic testing or heart checks either.

This approach of finding out who else in the family may have inherited the ICC is sometimes called 'cascade screening'.

The aim of pre-symptomatic testing is to predict someone's future risk of developing the ICC that is known to run in the family. By identifying family members who also have the genetic 'spelling mistake', it is hoped that their risk of ill-health and dangerous heart problems can be reduced through screening, surveillance and appropriate medical and/or surgical therapy.

If you would like to find out more about genetic testing, you should ask your GP or cardiologist to refer you to a specialist inherited cardiac conditions clinic or your local clinical genetics department. You should be offered a genetic counselling appointment with a specialist health professional (usually a genetic counsellor or doctor) to discuss genetic testing in greater depth.

If, for any reason, you do not want to have pre-symptomatic genetic testing you can still have clinical tests such as ECG and echocardiography. Your GP should refer you to a specialist inherited cardiac conditions clinic for this.





# What is genetic counselling?

Your genetic counselling appointment is a chance to learn more about the genetic basis of your ICC and to discuss what having the condition may mean for you and your family. You will be able to discuss the pros and cons of genetic testing in reference to your own personal situation, so you can work out whether having a genetic test really is the right choice for you.

Genetic counselling has an especially important role when you are considering pre-symptomatic genetic testing. Discovering you are at increased risk of an ICC when you feel well can have a significant impact on your life, and people can often react to this in different ways. It is worth thinking about your motivation for having genetic testing and finding out what genetic testing can actually tell you. For example, pre-symptomatic genetic testing will only be able to tell you whether you are at an increased risk of developing the ICC. It will not be able to tell you if you will ever develop symptoms, when you will develop them or how severe they may be.

If your pre-symptomatic genetic test shows that you do carry the 'spelling mistake', an outpatient appointment with a heart specialist (cardiologist or specialist nurse) is recommended and this can have implications for insurance, exercise, lifestyle and career choice. So it is worth thinking about how it may affect you and your lifestyle, before and during your appointment.

A genetic counselling appointment will typically last 30-45 minutes and can be used to discuss a variety of issues, as mentioned above, as well as any other questions you may have. The genetic counsellor or doctor will draw a family tree and discuss any health issues in the family. You should therefore come prepared with as much relevant personal and family information as possible (including documents where available).

# How is the genetic test done?

The genetic test itself involves you giving a blood sample or cheek swab. This will then be sent away to a specialist genetics laboratory.

If it is a genetic screening test, scientists there will look at the genes in your sample to see if they can find a gene change that might be responsible for causing your ICC. They will only look in the genes they know are involved in causing your ICC, not in any other genes, so it will not usually be possible to tell whether you are also at increased risk of other inherited conditions. The results of genetic screening can take several months (sometimes longer) to come through as this is a complex process.

If it is a pre-symptomatic genetic test, the scientists in the lab simply look to see whether you carry the same gene change as your relative(s) or not. The results for pre-symptomatic genetic tests therefore take less time to come back – usually about a month.

Results will be sent back to the genetic counsellor or doctor you saw originally and they will then let you know by whatever means you arranged e.g. another appointment, telephone, letter or email. When your genetic counsellor or doctor informs you of your results, you can have another discussion as to what are the appropriate next steps to take and ask any more questions that you may have.

## Where can I get more information?

If you would like more information about testing then please ask your GP to refer you to either a specialist inherited cardiac conditions clinic, a specialist cardiologist or your local clinical genetics department.

# Long QT Syndrome

Your genetics consultant/counsellor will be able to give more extensive advice. Long QT Syndrome is a rare condition. Experts would suggest that approximately 1 in 7000 people are affected but, as it is often never diagnosed, this is only an estimate. Long QT is a syndrome which can cause a disturbance in the electrical system of the heart, whilst the mechanical function of the heart can remain completely normal. It is often the result of inheriting an abnormal gene which causes an imbalance in molecules that control the electrical impulses of the heart. Long QT may result in a very fast abnormal heart rhythm (known as an arrhythmia). This particular arrhythmia is technically known as 'Torsade de Pointes'. When this abnormal rhythm occurs the heart loses its normal action and the blood is pumped out. The brain quickly becomes deprived of oxygen, resulting in a loss of consciousness (syncope) and, very occasionally death.

Arrhythmia in patients with LQTS may be triggered by exercise or stressful situations. Not everyone who has LQTS will have an arrhythmia, but if it does occur it can be fatal. LQTS is an electrical problem so it does not change the shape and function of the heart muscle.

## Symptoms

It is estimated that up to 50% of people with LQTS do not have symptoms. They may be aware of their condition only from results of an ECG performed for an unrelated reason, because they have a family history of LQTS or because of genetic testing results.

People with LQTS may start to experience symptoms in childhood, although this is not always the case. Symptoms can include unexplained seizures with no warning.

Sudden, unexplained fainting, particularly when in response to a stressful situation. This can often be misdiagnosed as having a 'hysterical reaction'.

Unexplained seizures. A sudden loss of consciousness may be mistaken or misdiagnosed as an epileptic seizure. Sudden cardiac arrest or death in the absence of any structural heart disease or other cardiac problems. Approximately 1 in 10 sudden cardiac arrests or death are the first sign of LQTS.

Diving into water could potentially be very dangerous for a person with Long QT, as a momentary loss of consciousness in water could result in drowning, due to a diving reflex.

Another sign or symptom of LQTS, is sudden loss of consciousness following a startle or a sudden loud noise.

## Causes

LQTS can be inherited or acquired. Acquired LQTS is usually due to the administration of certain medications. Many groups of common medications can cause a prolonged QT interval including certain antibiotics, antihistamines, antidepressants, antipsychotics and heart medications. A comprehensive list of medications can be obtained from your physician. Inherited LQTS is caused by mutations of certain genes and can be passed onto family members. The frequency of inherited LQTS is not known. There are several different types of inherited LQTS and your cardiologist may be able to tell you which type you have. The three most common types of inherited LQTS are called LQTS 1, LQTS 2 and LQTS 3.

At least 12 genes associated with LQTS have been discovered so far, and hundreds of mutations within these genes have been identified. Mutations in three of these genes (LQTS 1, 2 and 3) account for about 70 to 75 percent of LQTS cases. The type of LQTS may be identified by genetic testing. In types 1 and 2, the potassium channels within the heart cause the problem. In these types, arrhythmia may be triggered by exercise or by emotional stress. In type 3 it is the sodium channel that is affected and a low heart rate during sleep or rest may be the trigger for arrhythmia.

## Diagnosis

### ECG

An ECG is a simple tracing of the heart's electrical activity. It involves attaching electrodes to the chest and limbs and a recording is made. It may reveal a long QT interval which may suggest that it is more likely that you have LQTS. Not all people with LQTS have a prolonged QT interval on their resting ECG and it may be necessary to undertake several ECGs over a period of time, or have a period of continuous monitoring using a portable heart monitor.

### Exercise tolerance test

Some people may only have a prolonged QT interval when exercising so it may be necessary to have ECG monitoring done while exercising on a treadmill. This is normally the best way for your doctor to find out if you have LQTS. It can also help your doctor find out which type of LQTS you might have.

### A nonexercise (medication) stress test

An ECG test is performed while you are given a medication that stimulates your heart in a similar way to exercise. The medication is given through a vein in your arm and may include epinephrine (adrenaline). Doctors monitor the effects of the adrenaline on the way your heart recharges. This test can unmask what is known as concealed or borderline LQTS and mimics the heart's response to a sudden burst in adrenaline.

### Neurological test

An electroencephalogram (EEG) test looks for neurological causes of fainting, such as a seizure disorder. The procedure measures the waves of electrical activity the brain produces. Small electrodes attached to your head pick up the electrical impulses from your brain and send them to the EEG machine, which records brain waves. An EEG looks for other conditions that LQTS may be misdiagnosed as, e.g. epilepsy.

## Treatment

The main aim in treatment is to prevent loss of consciousness and a life threatening arrhythmia from occurring. There is no cure for LQTS but treatment options include: medications, medical devices, surgery or lifestyle changes. Treatment will be dictated by what type of long QT you have and what is most suitable for you.

It is wise to inform other people if you have LQTS so that they know to call for urgent medical help if you were to faint. Identity bracelets are available from certain charities which carry medical information about you. Your local arrhythmia nurse or your cardiologist may be able to give you more information about this. Alternatively, ask Arrhythmia Alliance for further details.

There are many medications which might affect the heart rhythm in patients with LQTS. These include some over-the-counter cough or cold remedies (decongestants) and some antibiotics. Other drugs that might affect the QT interval include some antidepressants, some treatments for fungal infections, and drugs for heart rhythm disorders.

If you are prescribed any medicines, always check with your doctor and pharmacist that it is safe for a patient with LQTS to take these medicines. Some herbal remedies are also to be avoided (e.g. St John's Wort). Please take care with herbal remedies and ask your doctor for specific advice.



A list of drugs currently known to affect long QT are available now on [www.crediblemeds.org](http://www.crediblemeds.org). This list will not be exhaustive as newer drugs are becoming more available. Always inform anyone who is prescribing you medication that you have LQTS as there may be newer drugs on the market which may have not have been added to the website.



## Medications

**Beta blockers:** These drugs slow the heart rate and make the dangerous rhythm associated with LQTS less likely. They work by blunting the way a LQTS-affected heart reacts to adrenaline in times of stress, fear or exertion.

**Mexiletine:** In people with a form of LQTS called LQT<sub>3</sub>, taking this antiarrhythmic drug in combination with a beta blocker may help shorten the QT interval.

**Potassium:** Potassium is a mineral in your body that is important for your heart's electrical system. Potassium supplements may improve the heart's recharging system and may be helpful for people with certain forms of LQTS.

## Medical devices and surgery

**A pacemaker or implantable cardioverter defibrillator (ICD):** These devices are implanted under the skin of your chest and stop a potentially fatal arrhythmia. A pacemaker produces electrical impulses to treat an abnormal heart rhythm. An ICD continuously monitors your heartbeat and will deliver electrical shocks to restore a normal heart rhythm when necessary.

**Left cardiac sympathetic denervation surgery:** Specific nerves in your chest controlling your heart rhythm are surgically removed which significantly reduces the risk of sudden death. This surgery is generally reserved for people considered at high risk of sudden death, people who do not tolerate the medications or have a fainting spell despite their medications.

### WARNING

Recreational drugs such as ecstasy and cocaine are particularly dangerous in patients with Long QT syndrome and CAN BE FATAL. Patients with even mild Long QT syndrome should NEVER experiment with these drugs.



# 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).

## Symptoms

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

## Diagnosis

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 he will advise you to have a simple test known as the flecainide (or Ajmaline) challenge to confirm your diagnosis. Flecainide 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 flecainide allowing the doctor to collect detailed information about the cause of your potential arrhythmia.

## Treatments

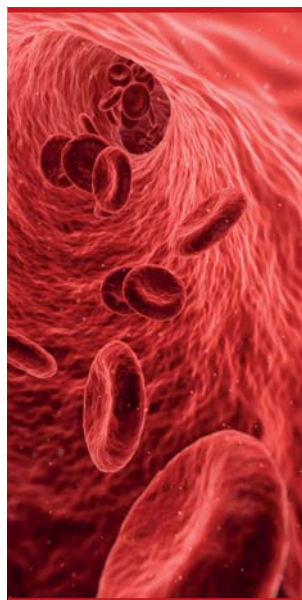
If the test result is negative, your doctor will consider your individual risk, and advise you if further tests are necessary. It is likely that you will be able to go home the same day. However, it is important to ask a friend or family member to collect you and drive you home. It is also recommended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

## Testing for 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.



# Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) is a rare inherited arrhythmia (heart rhythm disorder). CPVT is sometimes known as a channelopathy. This is because CPVT is caused by an abnormality of ion channels which allow calcium to enter the heart muscle cell. This can lead to an abnormally fast and irregular arrhythmia, ventricular tachycardia – usually during exercise or when adrenaline is released in stressful situations.

These extra beats most frequently arise from the bottom pumping chambers of the heart (ventricular ectopic beats). If there are runs of fast beats from the atria or ventricles these are known as atrial ventricular tachycardia.

In CPVT the heart usually has a normal structure. It is thought that about 1 in 10,000 people have CPVT, although the exact numbers are unknown.



## Symptoms of CPVT

Episodes of ventricular tachycardia (VT) can cause light-headedness, dizziness and loss of consciousness (syncope). Symptoms usually start in childhood but can appear in young adults for the first time. Most syncopal episodes in childhood are benign, however those who suffer syncope during exercise or in response to an adrenergic stimulus should be further investigated.

Sometimes these episodes can be mistaken for a fit/epilepsy as they can look very similar and as a consequence many children are treated with antiepileptic drugs. A child not responding to antiepileptic drugs should be referred to a cardiologist for further assessment. Unfortunately, sometimes the first presentation can be sudden cardiac arrest – an episode of VT cannot be sustained for a long period of time and may result in the heart completely stopping (cardiac arrest).



## Diagnosis of CPVT

In patients presenting with sudden cardiac arrest in the absence of structural cardiac disease, CPVT should be considered in the differential diagnosis. Clinical diagnosis is made based on family history, exercise or emotional stress induced symptoms and, significantly, response to exercise or catecholamine (adrenaline) infusion, however not all episodes are triggered by adrenaline. The cardiac ultrasound test (echocardiogram) and resting ECG are usually normal.

## Genetics

CPVT can result from one of two mutations in the cardiac ryanodine receptor gene – RYR2 and CASQ2, which are responsible for about 50% and 1-2% of cases respectively. These gene codes are responsible for making proteins that handle calcium, which help maintains a regular heartbeat.

Normally, heart muscle cells (myocytes) contract and relax in a coordinated way, but mutations in either RYR2 or CASQ2 impairs calcium handling within these myocytes and so during exercise or emotional stress, ventricular tachycardia may result. Familial occurrence has been seen in about a third of cases of CPVT. RYR2 causes autosomal dominant inheritance meaning if you inherit the abnormal gene from only one parent, you can get the disease. CASQ2 is inherited in an autosomal recessive manner meaning two copies of an abnormal gene must be present in order for CPVT to develop.



Currently, genetic testing is not widely available. However first-degree relatives should be evaluated with ECG, Holter monitoring and exercise stress testing. Genetic testing can sometimes be helpful. Identification of a genetic abnormality that is the definite cause of the condition (a “pathogenic” mutation) can allow other family members to be tested. Genetic analysis might identify silent carriers of CPVT– related mutations, and it may be recommended that even symptom-free carriers are treated with medication such as beta blockers. Sometimes, a genetic change is discovered, and it is unclear if this is the definite cause. This is known as a “variant of unknown significance” or VUS. A VUS cannot be used for family screening.

## Treatment

Once diagnosed, treatment is usually with a beta blocker. Beta blockers decrease the activity of the heart by blocking the action of hormones such as adrenaline, which would normally increase in times of exercise or emotional stress. Subsequently, the number of episodes of VT is reduced. A high dose is often required. Flecainide may also be used in addition to beta blockers if the response is inadequate. Flecainide inhibits cardiac ryanodine receptor-mediated calcium release.

Missing even a single dose of beta blocker can be potentially dangerous. Internal cardiac defibrillators (ICD) are sometimes fitted in addition to medication to 'shock' the heart back into normal rhythm if VT occurs. Survivors of cardiac arrest or high-risk patients with a strong family history of sudden death are likely to receive an ICD. ICD treatment without use of beta blockers is not advised as a shock from the defibrillator can lead to an adrenaline surge and multiple runs of ventricular tachycardia known as an 'electrical storm'.

A left cervical sympathectomy may be offered to some patients, e.g. those in whom beta blockers are contraindicated, when an ICD cannot be fitted, or where there is recurrent VT in patients with an ICD despite maximal medical treatment. A cervical sympathectomy is an operation carried out through a small incision under the arm. This blocks a group of nerves that produce and deliver adrenaline to the heart. These nerves are not essential to normal heart function but sympathectomy can be very helpful in preventing serious arrhythmias.

Additionally, some sports may be restricted, e.g. swimming, and certain medications that increase the heart rate may need to be avoided.

# Wolff-Parkinson-White Syndrome (WPW)

Wolff -Parkinson-White Syndrome (WPW) is a combination of symptoms of palpitations, with the presence of an extra electrical pathway, resulting in an ECG abnormality. The extra pathway can result in episodes of fast heart rhythm. This is a problem present from birth but may not present itself until adulthood. It affects between one and three in every 1,000 people. In most cases, the heart is structurally normal.

The extra electrical pathway (known as an accessory pathway) directly connects the atria (the top chambers of the heart) to the ventricles (the bottom chambers of the heart). If the electrical impulses travel up the accessory pathway, and down the AV node, a fast heart rhythm called supraventricular tachycardia (SVT) may occur. In rare cases, a different, irregular rhythm, called atrial fibrillation (AF), may travel down the accessory pathway and cause a very fast unstable heart rhythm disorder. This is known as pre-excited AF and usually requires emergency medical treatment.

## Testing for family and relatives

There is very little evidence of any genetic or familial form of WPW. It is thought to be caused by a small heart muscle fibre which becomes stranded slightly out of place during development in the womb. If there is any major concern about a relative, then a simple 12-Lead ECG test can be used to screen them for WPW or pre-excitation.

## Symptoms

Some people have no symptoms and just have the ECG abnormality due to the accessory pathway. This is not strictly speaking WPW, but called pre-excitation. The condition is usually then only discovered on routine ECG recording.

People may report the following symptoms varying from mild to severe: a fast and racing heartbeat (supraventricular tachycardia), feeling lightheaded or dizzy, shortness of breath, chest pain, sweating, feeling anxious, syncope (fainting).



Symptoms can last for seconds, minutes or hours, and vary in frequency from daily occurrence to only a few times a year.

## Causes

Symptoms due to WPW usually occurs randomly without any identifiable triggers.

## Diagnosis

Initially you will have an electrocardiogram (ECG) performed which may show evidence of an accessory pathway responsible for WPW. You will then be referred to a specialist heart doctor; a cardiologist or electrophysiologist.

If your doctor suspects you have WPW, but it is not completely clear on the routine ECG, he/she will advise you to have an adenosine challenge to confirm your diagnosis. This is known as latent pre-excitation. Some patients have intermittent pre-excitation, which means it is not seen on every ECG.



## Adenosine challenge

Adenosine is a naturally occurring substance found in all of us. Adenosine briefly blocks normal conduction through the AV node, which slows your heart rate and unmasks ECG changes in patients who have a latent WPW, since the accessory pathway (or bypass tract) is not blocked, and so a fast heart rate still occurs.

Your doctor will administer the drug through a vein in your arm and record your ECG. The ECG will record the effects of the adenosine on the AV node and unmask any presence of an accessory pathway.

## Treatments

If the test result is negative, your doctor will consider your individual risk, and advise you if further tests are needed to be performed. It is likely that you will be able to go home a few hours after the test. However, it is advisable that you do not drive, and that you have someone with you for the rest of the day after the test.

If the test is positive, and you may be at risk of a fast heart rhythm developing, your doctor may suggest you have an electrophysiology (EP) study and possible catheter ablation. The EP study is an invasive procedure, where catheters are placed within the heart via the vein at the top of your leg.

Various electrical measurements are made, to determine if the accessory pathway is capable of supporting SVT, or pre-excited AF. It will be possible to

tell how fast the pathway conducts electrical impulses and if it is safer, in the long term, to destroy it in the form of catheter ablation. Catheter ablation is a curative procedure that will destroy the extra pathway that is capable of causing SVT and pre-excited AF.

Your consultant/nurse specialist will discuss the risks and benefits of catheter ablation with you should you appear to need one. Following your discharge from hospital you will be able to return to your normal daily activities, including returning to work. Your doctor/nurse specialist may recommend drug treatment prior to any invasive treatment.

This may be an antiarrhythmic drug, such as flecainide or a beta blocker, which both help to prevent fast heart rhythms and slow down the action of the accessory pathway. With regard to lifestyle modifications, it is important to avoid illicit or recreational drugs, such as cocaine, ecstasy and other stimulants, as these can be dangerous in this condition.

# Donation Form



## Your details

Title \_\_\_\_\_ First Name(s) \_\_\_\_\_ Surname \_\_\_\_\_  
Address \_\_\_\_\_  
\_\_\_\_\_ Postcode \_\_\_\_\_  
Email \_\_\_\_\_ Phone number \_\_\_\_\_

## Your donation Please tick method of payment & complete relevant section:

enclose a cheque made payable to 'Arrhythmia Alliance' for £ \_\_\_\_\_

or Please debit my credit/debit card for £ \_\_\_\_\_ Card type  maestro  visa  mastercard  
Valid from   /   Card no.                  
Expiry date   /   3-digit Security no.

or I would like to set up a regular standing order for £ \_\_\_\_\_ starting on   /   /

To be paid: Monthly  Quarterly  Annually

My bank name \_\_\_\_\_ Bank address \_\_\_\_\_  
\_\_\_\_\_

Account no.                 Sort code

**Payable to:** Arrhythmia Alliance, Account no. 02685818, Sort code 30-98-26  
Lloyds TSB Plc, 22 Bridge Street, Stratford upon Avon, CV37 6AG  
Please note you can cancel your standing order at any time by contacting your bank.

## Gift Aid

If you pay UK Tax, the Government will give us 25% on top of your donation at no cost to you.  
In order to Gift Aid your donation you must tick the box below:

\* I want to Gift Aid my donation of £ \_\_\_\_\_ and any donations I make in the future  
or have made in the past 4 years to Arrhythmia Alliance

\*I am a UK taxpayer and understand that if I pay less Income Tax and/or Capital Gains Tax than the amount of Gift Aid claimed on all my donations in that tax year it is my responsibility to pay any difference. Please notify us if you want to cancel this declaration, change your name or home address or no longer pay sufficient tax on your income and/or capital gains. Your address is needed to identify you as a current UK taxpayer.

## Your Signature

Signature \_\_\_\_\_ Date   /   /

**Post me to:** Arrhythmia Alliance, Unit 6B, Essex House, Cromwell Business Park, Chipping Norton, Oxfordshire, OX7 5SR  
If you have any queries please do not hesitate to call us on 01789 867501



Arrhythmia Alliance

[www.hearhythmalliance.org](http://www.hearhythmalliance.org)

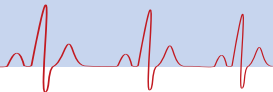
Working together  
to improve the  
diagnosis, treatment  
and quality of life for  
all those affected by  
arrhythmias

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Please remember that this publication provides general guidelines only. Individuals should always discuss their condition with a healthcare professional.

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