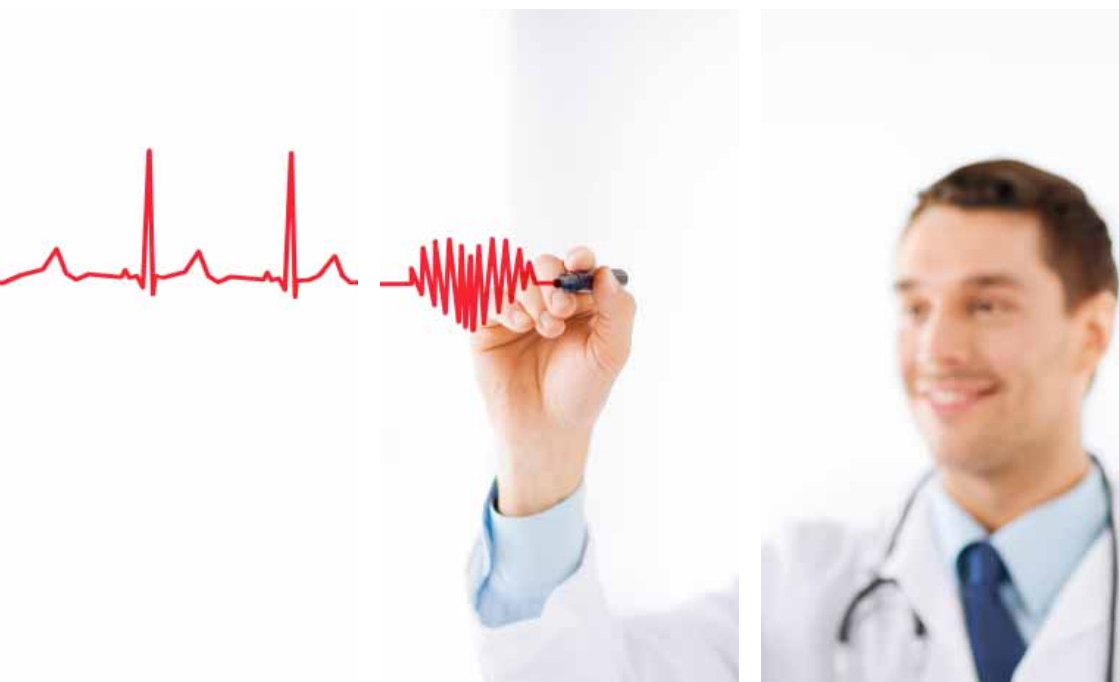




# Arrhythmias: Understanding your condition



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

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

Registered Charity No. 1107496

# Glossary

**Arrhythmia** An abnormal heart rhythm

**Atrium** Top chambers of the heart that receive blood from the body and from the lungs. The right atrium is where the heart's natural pacemaker (sino atrial node) can be found

**Asystole** Cessation of heartbeat

**Bradycardia** A slow heart rate, normally less than 60 beats per minute

**Cardiac arrest** A sudden cessation of the heart to contract effectively, or at all

**Cardiologist** A doctor who has specialised in the diagnosis and treatment of patients with a heart condition

**Cardioversion** A procedure by which an abnormally fast heart rate (tachycardia) or other cardiac arrhythmia is converted to a normal rhythm using electricity or drugs

**Defibrillation** Treatment for life threatening cardiac dysrhythmias whereby a high energy shock is given to the heart in an attempt to return it to normal rhythm

**Electrocardiogram (ECG)** An ECG is a simple, non-invasive test that records the electrical activity of the heart

**Heart attack** A medical emergency in which the blood supply to the heart is blocked, causing serious damage or even death of heart muscle

**Heart block** Electrical impulses are slowed or blocked as they travel from the top to the bottom chambers of the heart

## Contents

The heart and arrhythmias

Atrial Fibrillation (AF)

Atrial Flutter

Bradycardia

Brugada

BBB

CPVT

Ectopic Beats

Heart Failure

IST

JET

Long QT S

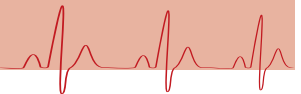
SVT

Syncope

VF

VT

WPW Syndrome



### **Implantable cardioverter defibrillator (ICD)**

A small device connected to your heart to help treat irregular heart rhythms by using electrical shocks, helping to control life-threatening arrhythmias, especially those that can cause sudden cardiac arrest (SCA)

**Implantable loop recorder (ILR)** Monitor implanted for a period of time to continually record your heart rhythm

**Pacemaker** A small device implanted under the skin, which produces electrical impulses to treat an abnormal heart rhythm

**Syncope** The medical term for fainting or unexplained loss of consciousness caused by a sudden lack of blood supply to the brain

**Tachycardia** An abnormally fast heart rate over 100 beats per minute

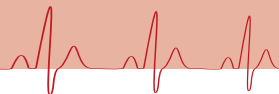
**Ventricles** The two lower chambers of the heart. The right ventricle pumps blood into the lungs and the left ventricle pumps blood around the body

**Ventricular fibrillation (VF)** A fast, life threatening heart rhythm which causes the heart to stop pumping effectively. This rhythm needs an electrical shock to stop it and return the heart back to a normal rhythm

**Ventricular tachycardia (VT)** A dangerously fast heart rhythm which causes the heart to pump less efficiently, and can lead to dizziness, fainting and unconsciousness. If not treated with medication or an electric shock, the rhythm can lead to VF

## **IMPORTANT INFORMATION**

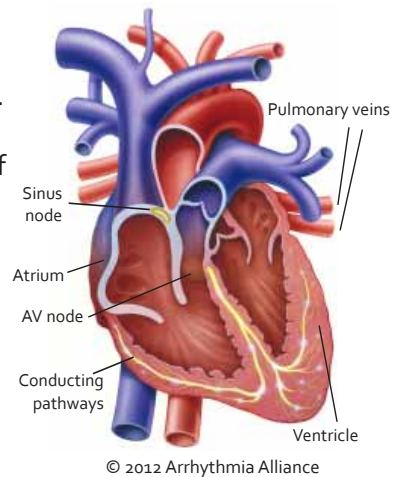
This booklet is intended for use by people who wish to understand more about arrhythmias, SVT, bradycardia, WPW syndrome, inappropriate sinus tachycardia, Brugada, long QT, CPVT, AVNRT, atrial fibrillation, atrial flutter, ventricular tachycardia and ventricular fibrillation. The information within this booklet comes from research and previous patient's experiences.



# The heart and normal conduction

## The heart and normal conduction

The heart has its own electrical conduction system. The conduction system sends signals throughout the upper (atria) and lower (ventricles) chambers of the heart to make it beat in a regular, coordinated rhythm. The conduction system consists of two areas called nodes that contain conduction cells and special pathways that transmit the impulse. The normal heartbeat begins when an electrical impulse is fired from the sino-atrial node (SA node/ SAN), in the right atrium. The sinus node is responsible for setting the rate and rhythm of the heart and is therefore referred to as the heart's 'pacemaker'.



The electrical impulse fired from the SAN spreads throughout the atria, causing them to contract and squeeze blood into the ventricles. The electrical impulse then reaches the atrioventricular node (AV node/ AVN), which acts as a gateway, slowing and regulating the impulses travelling between the atria and the ventricles. As the impulse travels down the pathways into the ventricles the heart contracts and pumps blood around the body. The cycle then begins all over again. The normal adult heart beats in a regular pattern 60-100 times a minute; this is called sinus rhythm.

# What are arrhythmias?

Arrhythmias are disorders of your heart's electrical system whereby there is a change in the regular beat of your heart. Sometimes if the conduction pathway is damaged or becomes blocked, or if an extra pathway exists, the heart's rhythm changes. The heart may beat too quickly (tachycardia), too slowly (bradycardia) or irregularly which may affect the heart's ability to effectively pump blood around the body. These abnormal heart rhythms are known as arrhythmias. Arrhythmias can occur in the atria, the ventricles, or both. Arrhythmias may occur at any age, and are most often a nuisance rather than a serious problem.

Most arrhythmias arising from the top of the heart (supraventricular) are troublesome, but not life threatening, although there are exceptions. Many arrhythmias arising from the bottom chambers (ventricles) can be life threatening. Many arrhythmias can be completely cured by keyhole techniques (catheter ablation). Ventricular arrhythmias are often much harder to treat, and often require powerful drugs and implantable life saving devices called defibrillators.

## What happens in the heart to cause an arrhythmia?

Any interruption in the heart's electrical system can cause an arrhythmia. For example, an irregular heartbeat may begin with an abnormal impulse in a part of the heart other than the normal pacemaker (the sinus node); or the sinus node may develop an abnormal rate or rhythm.

## What can trigger an arrhythmia?

Common causes of arrhythmias include electrical variations that people are born with, which may only become a problem in adult life. Certain triggers can include stress, caffeine, tobacco, alcohol, diet pills, and cough or cold medicines, but there is usually an underlying physical reason for it. If your heart tissue is damaged as a result of acquired heart disease such as myocardial infarction (heart attack) or congenital heart disease, you may also be at risk of developing an arrhythmia. In rare cases, it may be that doctors cannot identify a cause of their arrhythmias.

# How do I know what kind of arrhythmia I have?

You will need to visit your doctor and have an electrocardiogram (ECG). If the ECG does not detect any abnormality, it may be necessary to arrange for further monitoring of your heart. This may involve having a continuous ECG for a period of time, usually 24-72 hours. This is done via a small recording device which can easily be carried around with you. You do not have to stay in hospital for this test. Once the recording device is fitted, which involves attaching some small stickers to your chest and connecting the leads of the device to these, you can go home and return the recorder at the end of the specified period. There are also other ways of monitoring your heart over a period of time; your nurse, physiologist or doctor will discuss these with you if required.



## Symptoms of arrhythmias

Patients with arrhythmias often complain of awareness of the heart beating or palpitations. Doctors will usually ask if the palpitations are rapid, if they are regular, or irregular. Tests will usually be done to determine if the arrhythmia is associated with a normal or damaged heart function and these are used to assess the risk to the patient. A key to successful treatment of an arrhythmia is to record the attacks on an ECG whilst they are happening. This gives further information about the nature of the arrhythmia, any risk, and the possibilities of successful treatment.

# Diagnosing an arrhythmia

If your doctor suspects that you may have an arrhythmia, one or more of the following tests may be performed to determine the cause of your symptoms.

## Electrocardiogram (ECG)

An electrocardiogram is a simple, non-invasive recording of the electrical activity of your heart. Electrode stickers are placed on your chest and connected by wires to a recording machine. Your heart's electrical signals produce a pattern on graph paper which is the ECG recording. The ECG may be digitally recorded on a screen and then uploaded to your electronic patient record. By analysing the pattern of these waves, your doctor can often determine what type of arrhythmia you have. ECG testing can be done at rest, or while you are exercising on a treadmill. The ECG is critical in assessing a heart rhythm disorder, the presence of any underlying heart problems, whether an arrhythmia is present, and whether it is an arrhythmia that requires treatment.

## Holter monitor

A holter monitor shows changes in your heart rhythm over the course of 24 hours, 48 hours or a longer period, that may not be detected during a resting or exercise ECG. If your doctor wants you to have this test, you will be asked to go about your daily activities as usual (except for showering or bathing) while you wear a small, portable recorder that connects to electrode stickers on your chest. You will be asked to come back to the hospital the next day so that the information can be retrieved and analysed.

## Tilt test

A tilt test is sometimes used to find the cause of blackouts. In some cases, a blackout may be precipitated and then the doctors will have a much clearer idea of the cause. The patient is secured to a tilting table, and monitors are attached, such as an ECG and blood pressure cuff. The table tilts up to a near standing position where the patient remains for about one hour. During this period heart rate and blood pressure are monitored and any changes recorded. Sometimes you may be given some medication towards the end of the test to see if a reaction can be provoked. This will all be discussed with you before the test.

# Types of arrhythmia

Arrhythmias that occur in the atria (the top chambers of the heart) are either atrial or supraventricular (above the ventricles) in origin whereas ventricular arrhythmias start in the ventricles (the lower chambers of the heart). While some arrhythmias are merely a nuisance, others can be life-threatening. A doctor will determine which type of arrhythmia you may have and treat your symptoms accordingly. In general, ventricular arrhythmias caused by heart disease are the most serious kind, and require prompt medical attention.

## Atrial fibrillation (AF)

Atrial fibrillation (AF) is one of the most common types of arrhythmia, and affects approximately one million people in the UK alone. It occurs in the atria, in the upper chambers of the heart. The electrical impulse normally originates at the SA node. However, in AF, many electrical impulses are fired rapidly and at random throughout the atria down to the ventricles. The resulting heartbeat is irregular and usually fast.

### Symptoms of AF

Symptoms of palpitations which may be rapid, shortness of breath, dizziness, chest discomfort, light headedness, fainting or fatigue are common in AF. Some people with AF do not have any symptoms, and it may only be discovered at a routine medical examination or following an admission to A&E with another condition. The easiest way to detect AF is to feel your pulse.

### Causes of AF

AF is related to age, the older you get, the more likely you are to develop it. AF is frequently noted after an 'open heart' operation. Other conditions or diseases can also increase your risk of getting AF. This does not mean that AF always develops, but the risk does increase.

Other conditions associated with AF are high blood pressure, coronary heart disease, mitral valve disease, congenital heart disease, pneumonia, lung cancer, pulmonary embolism, overactive thyroid and carbon monoxide poisoning.



In addition, alcohol and drug abuse or misuse may predispose you to AF. While your risk of AF goes up with the problems mentioned, many people develop AF for no explainable reason.

### **Treatment of AF**

Currently, drugs are the most common treatment for AF, and have the aim of restoring the normal sinus rhythm, alleviating symptoms and reducing the likelihood of an AF-related stroke.

Some drugs are used to restore the normal heart rhythm, these are known as antiarrhythmic drugs. They work by blocking specific channels in the cardiac cells.

Beta blockers are commonly used to slow the heart rate and are effective in active patients with better exercise capacity.

Because the atria are beating rapidly and irregularly (fibrillating), they are unable to completely empty all the blood they receive into the ventricles and this can cause blood clots to form. Therefore, if you are at an increased risk of stroke you may be treated with an anticoagulant. Anticoagulants can reduce the risk of AF-related stroke by at least 65%.

There have been many medical advances and innovations in the treatment of AF. The outlook for people with AF is excellent and there is every reason to believe that quality of life will be as good as anyone else's.

For more extensive information about atrial fibrillation, please contact our sister charity AF Association on 01789 867 502 or email [info@afa.org.uk](mailto:info@afa.org.uk)



## Atrial flutter

Atrial flutter is an abnormality in the rhythm of the heart (arrhythmia). In this arrhythmia, the atria beat very rapidly. Unlike AF, atrial flutter is a more organised electrical disturbance which originates in the right atrium in the majority of patients. The atria beat very quickly and regularly, at around 300 beats per minute, and hence take over from the sinus node in controlling the heart rate. The AV node will not conduct all of these atrial beats to the ventricles but tends, instead, to only allow every second, third or fourth beat through, creating an often-regular heart rate of around 150, 100 or 75 beats per minute.

### Symptoms of atrial flutter

Some people with atrial flutter experience no symptoms, and the arrhythmia can be a chance finding on a routine ECG. Common symptoms are palpitations, fatigue or poor exercise tolerance, mild shortness of breath, dizziness and less commonly chest pain and fainting.

### Causes of atrial flutter

As atrial flutter and AF share many similarities and can occur in the same patient, the two arrhythmias share many causes. Atrial flutter is more likely to occur as one gets older, and is more common in patients who have a history of previous heart disease. Men are more than twice as likely to get atrial flutter as women. Often there is no single factor that causes atrial flutter, rather there are a number of factors and conditions that increase the likelihood of atrial flutter.

Some of the risk factors for developing atrial flutter are high blood pressure, ischaemic heart disease, disease of the heart valves, cardiomyopathy, pneumonia, cardiac surgery, congenital heart disease, pericarditis, over active thyroid, COPD (chronic obstructive pulmonary disease), excess alcohol and pulmonary embolism.

However, these are not the only causes for developing atrial flutter and for some there may appear to be no obvious reason.

## Treatment of atrial flutter

The treatment of atrial flutter follows similar lines to the treatment of AF. Treatment is centred around reducing symptoms and reducing the risk of stroke, so the treatment for individuals may vary depending upon their symptoms and their stroke risk.

There are a variety of drugs that can be used in the treatment of atrial flutter. Different drugs are used to achieve different treatment goals, and often two or more drugs are used in combination.

Drugs such as flecainide, amiodarone, dronedarone, sotalol or propafenone may be prescribed to restore and maintain a normal heart rhythm and are referred to as antiarrhythmic drugs.

Drugs such as beta blockers, calcium channel blockers or digoxin are used in atrial flutter in order to slow the heart rate by reducing the number of atrial flutter beats that are conducted via the AV node from the atria to the ventricles.

The risk of stroke in atrial flutter is thought to be similar to that for AF and is five times greater than in the normal sinus rhythm. Anticoagulants are drugs that are used in the treatment of atrial flutter, and are ones that 'thin' the blood and reduce the risk of stroke.



## **AV nodal re-entry tachycardia (AVNRT)**

This type of arrhythmia occurs when a problem arises in the way the electrical impulses pass through the AV node. Normally, the AV node acts as a gateway slowing and regulating the impulses as they travel between the atria and the ventricles. In AVNRT there are two pathways, known as dual conduction pathways, that can pass impulses to and from the AV node. This type of arrhythmia usually starts following an early beat (ectopic beat). An electrical short circuit then occurs whereby the electrical impulse rotates around the circuit and with each cycle passes to the ventricles resulting in a very fast heartbeat.

## **Bradycardia**

Bradycardia is a term that describes a number of different conditions in which the heart beats at an unusually slow rate. If impulses are sent from the sinoatrial node at a slow rate, or if the impulses are delayed as they travel through the conduction system, the heartbeat will be slow. Sinus bradycardia is an unusually slow heartbeat due to normal causes and commonly occurs in athletes or during a state of deep relaxation. This is perfectly normal and should not usually cause any concerns. The severity of and treatment required for the bradycardia depends on the area of the heart affected. Bradycardia may also be caused by age-related degeneration of the heart's electrical conduction system, coronary heart disease or by medications prescribed to treat arrhythmias or high blood pressure. The benefits of the medication may outweigh the risk of Bradycardia however, so it is best to discuss your options with your physician. Another cause is conditions that can slow electrical impulses through the heart. Examples include having a low thyroid level (hypothyroidism) or an electrolyte imbalance, such as too much potassium in the blood (hyperkalaemia). Other causes which can lead to bradycardia such as heart disorders present at birth (congenital heart disease), heart surgery, heart attacks and heart infections.

### **Symptoms of bradycardia**

Some types of bradycardia produce no symptoms, and others may cause fatigue, dizziness, breathlessness on exertion, chest pain or fainting (syncope).

## Syncope

There are several causes of syncope, some more common than others. Most cases of syncope are due to the 'common' faint. However, other important causes include defects of the 'wiring' of the heart. Syncope can occur when the heart slows or momentarily stops (asystole). Therefore, oxygenated blood is not pumped to the brain, causing light-headedness, dizziness, fading of vision, buzzing in the ears and loss of consciousness. Often patients will recognise these symptoms and be able to sit or lie down before losing consciousness. However, for many there are no symptoms, just an abrupt loss of consciousness. People of all ages experience syncope, including children (reflex anoxic seizures/ reflex asystolic syncope due to an unexpected stimulus such as a bump or fright). Syncope involving bradycardia can be diagnosed by taking a detailed history, having an ECG (electrocardiogram), and using a special event monitor, of which there are various types, based upon the frequency of the symptoms.

## Investigations for bradycardia

There is a range of different investigations that commonly look for the causes as well as the nature of the different bradycardias. It is important to note that these will be tailored to the individual patient and are performed as part of a complete assessment done by your doctor, which also includes a careful review of your medical history, as well as a physical examination. Common investigations include blood tests, especially the ones that assess the levels of your blood electrolytes, and an ECG which gives a snapshot of your rhythm at the time of recording, as well as more prolonged monitoring by means of a heart monitor. The later investigations include portable ECG devices called holter monitors or event recorders. In some cases, especially if the symptoms are occurring very infrequently such as once or twice every few months, and there is a suspicion that the heart monitors may miss detecting the arrhythmia, your doctor may recommend the implantations of a small device underneath the skin called an insertable/ implantable loop recorder (ILR).

In some cases, your doctor may recommend an exercise tolerance test to look at the heart rate in response to physical exercise, or a tilt table test, which involves tilting you gently on a flat table and assessing if changes in your posture lead to abnormal heart rhythms.



### **Treatment for bradycardia**

The goal of treatment is to keep your heart rate at a level high enough to allow sufficient blood flow to the body. If severe bradycardia is left untreated, it can cause serious problems such as fainting or even death.

### **What can I do to help my condition?**

Try to be active on a daily basis if possible. Your doctor will be able to advise on safe exercise for your individual circumstances. Eat a healthy and balanced diet. Try to maintain a healthy weight. Ask your doctor for help if you need to lose weight. If you smoke, your doctor will provide support to help you quit. Ensure that any other health problems such as high cholesterol or high blood pressure are well managed.

## 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 of Brugada syndrome

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

### Causes of Brugada syndrome

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.

### Diagnosis of Brugada syndrome

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

Ajmaline and 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 ajmaline or flecainide allowing the doctor to collect detailed information about the cause of your potential arrhythmia.

### **What treatment options are available?**

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. 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.

For more information on Brugada syndrome, including more on the flecainide challenge and genetic screening, please contact Arrhythmia Alliance.





## Bundle branch block (BBB)

Bundle branch block (BBB) is a condition whereby there is a delay or blockage along the conduction pathways within your heart that control your heart beating/contracting. It makes it harder for your heart to effectively pump blood to the rest of your body. The delay or blockage can either affect the left or right conduction pathways, resulting in left or right bundle branch blocks.

### Symptoms

Many people with BBB won't have any symptoms and therefore won't be aware that they have the condition. However others may experience fainting (syncope) or feeling as if they may faint (presyncope)

### Causes of left bundle branch block (LBBB)

- Myocardial infarction (MI) (heart attack)
- Cardiomyopathy (thickened, stiffened or weakened heart muscle)
- Myocarditis (viral or bacterial infection of the heart)
- Hypertension (high blood pressure)

### Causes of right bundle branch block (RBBB)

- May be a normal finding
- Increasing age
- Underlying health problems such as heart disease and hypertension

### Investigations

- Electrocardiogram (ECG) - This records the electrical rate and rhythm of the heart and can detect left and right BBB amongst other abnormalities.
- Echocardiogram - This test uses ultrasound waves to create detailed images of the heart's structure including the thickness of the heart muscle, and whether the heart's valves are moving normally. It may diagnose a condition which has caused the BBB.

## Complications

The main complication of bundle branch block is if it progresses to a complete heart block whereby the electrical conduction from the upper atria chambers of the heart down to the lower ventricular chambers is blocked. This results in bradycardia (a slow heart rate) leading to fainting and potentially serious arrhythmias (abnormal heart rhythms).

People who have a heart attack and subsequently develop LBBB have a higher chance of complications including sudden cardiac death.

## Treatment

Many people with BBB will be asymptomatic and will not require any treatment. However if you are symptomatic and have a condition that is causing the BBB, treating that condition may involve taking medication to reduce high blood pressure or lessen the effects of heart failure.

Sometimes individuals are offered a pacemaker or cardiac resynchronisation therapy (CRT). A pacemaker provides electrical impulses to the heart to keep it beating regularly. Therefore the BBB is overcome by having a pacemaker implanted. CRT, also known as biventricular pacing, is a procedure similar to having a pacemaker implanted. It keeps both the right and left sides of the heart beating in a normal rhythm, with both ventricles contracting at the same time. It can be beneficial to people with heart failure and a BBB.

## Catecholaminergic polymorphic ventricular tachycardia (CPVT)

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an inherited arrhythmia (heart rhythm disorder). CPVT is part of a group of conditions called channelopathies. This is because CPVT is caused by an abnormality of ion channels which allow excess calcium to enter the heart muscle cell. This can lead to rapid arrhythmias, which can be dangerous, and usually occur during exercise or when adrenaline is released in stressful situations. Extra beats can arise from the top collecting chambers of the heart (atrial ectopic beats) or, more frequently, 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 or tachycardia. Atrial tachycardia can cause unpleasant symptoms but is usually not dangerous, whereas ventricular tachycardia is often dangerous. 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. Syncopal episodes in most children are benign, however those who suffer syncope during exercise or in response to an adrenergic stimulus should be further investigated as they might have CPVT. Sometimes these episodes can be mistaken for a fit/epilepsy as they can look very similar and, as a consequence, many children with channelopathies are mistakenly 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

CPVT should be considered in all patients presenting with sudden cardiac arrest or unheralded blackouts in the absence of structural heart disease, particularly in younger patients (< 40 years) and especially in those where exercise or

emotional stress is a significant factor. The diagnosis is supported if there is a family history of CPVT or unexplained sudden death. The cardiac ultrasound test (echocardiogram) and resting ECG are normal so the main diagnostic test is an exercise tolerance test on a treadmill or bicycle, which usually shows worsening ventricular arrhythmia on increasing levels of exercise. Testing with catecholamine (adrenaline) infusion can be useful, particularly in those who are unable to exercise.

## Genetics

CPVT can result from one of two mutations in the cardiac ryanodine receptor gene – RYR2 and CASQ2, which are responsible for about 65% and 1-2% of cases respectively. These gene codes are responsible for making proteins that handle calcium, which help maintain 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.

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.

First degree relatives of patients diagnosed with CPVT are also at risk of the condition and should be assessed. Genetic testing can be very useful in CPVT. If a pathogenic (disease-causing) mutation is found in the person affected by CPVT, then relatives can be tested to see whether they carry this mutation and are therefore at risk of having CPVT. 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. If no mutation is found then first-degree relatives should be evaluated with ECG, Holter monitoring and exercise stress testing (this is known as clinical testing). Sometimes, a genetic change is discovered and it is unclear if this is the definite cause. This is known as a “variant of uncertain significance” or VUS. A VUS cannot be used for family screening but occasionally further information arises which allows the VUS to be upgraded or downgraded in future.

## 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, participation in sports is likely to be restricted, e.g. swimming, and certain medications that increase the heart rate may need to be avoided.

## Ectopic beats

An ectopic beat is an additional beat of the heart which can come from either the upper chamber of the heart (the atrium) or the lower chamber (the ventricle). They are therefore described as premature atrial contractions (PACs) or premature ventricular contractions (PVCs). The beat occurs just before the normal heartbeat. Ectopics tend to occur when the heart rate is slower, such as when we are relaxing in the evening or sleeping at night.

Ectopics can either be felt as an extra beat in the rhythm of the heart or as a thud following a short pause in the rhythm of the heart. In this second example the additional beat of the heart has occurred in the pause, and the following thud is the heart catching up.

Ectopic beats are common, and in most people with no other known heart condition they are harmless. They carry no increased stroke risk even in patients with a damaged heart, such as those with heart failure or runs of SVT.

Although the symptoms can be unpleasant or cause anxiety, they do not indicate any problems with the heart, and the extra beats will not usually cause any damage.

Normally a clinician will diagnose an ectopic beat from what you have told them. A heart tracing electrocardiogram (ECG) will confirm this diagnosis. Any ectopic beats will appear as a different pattern on the visual trace. If the ectopics are less frequent, it may not be possible to catch them on an ECG, so a portable monitor may be more likely to capture them.

In patients with frequent symptoms, a 24-hour ECG will sometimes be undertaken to clarify the pattern and frequency of the ectopic beats and their relation to symptoms. Although ectopic beats are not a cause for concern in most individuals, in those with structural heart disease they may be of greater significance, and further cardiological assessment may be advised.

If a person suffering with ectopic beats is otherwise fit and healthy, all that is usually needed is reassurance and advice, such as cutting down on anything that may be acting as a stimulant, for example alcohol or caffeine. Stress can also trigger ectopic beats. Off the shelf cold and flu medicines sometimes contain decongestants and other drugs that can stimulate the heart, and these are probably best avoided.



If symptoms are persistent and uncomfortable, medical therapies may be appropriate. Generally, your doctor would initially use a medication such as a beta blocker (e.g. bisoprolol). In patients with asthma or bronchitis, such medicines cannot be used, and an alternative such as a calcium channel blocker (e.g. diltiazem) may be considered. In some otherwise healthy individual, other drugs may reduce the symptoms effectively, but their risks may outweigh the benefits.

In summary, ectopic beats are nearly always harmless and do not indicate any problems with the heart. However, it is always best to see your doctor for evaluation and investigation.

## Heart failure

Heart failure is a condition in which your heart does not pump blood as efficiently around your body as it should, which makes it difficult for your body to get as much oxygen and blood as it needs. When the organs do not get enough oxygen to work effectively, this can then cause a number of symptoms, particularly when being active.

### Symptoms of heart failure

One of the most common symptoms of heart failure is breathlessness, which worsens with physical activity or exercise. Fluid can also build up in the lungs which can make it difficult to breathe. This can happen when you are exercising or have been active, or in more severe cases can also happen at rest. This build-up of fluid can also cause swollen ankles, legs or abdomen, as the heart is not pumping effectively enough to remove this fluid. A persistent cough may also develop, due to the excess fluid sitting in the lungs.

Other symptoms can include tiredness, palpitations, a rapid or irregular heartbeat, weakness, dizziness or fainting, rapid weight gain from fluid retention or decreased alertness.

### Treatment options for heart failure

As such, there is not usually a 'cure' for heart failure. There are however many treatment options that control symptoms, ease discomfort, and allow people with heart failure to lead a happy and healthy life. Lifestyle changes, medications, medical devices and surgery, are different treatment options for people with heart failure. The options will be discussed with you, and will depend greatly on your symptoms, the severity of your condition and what is causing your heart failure.

For more information about heart failure, please contact Arrhythmia Alliance on 01789 867501 or by emailing [info@heartrhythmalliance.org](mailto:info@heartrhythmalliance.org) for our living with heart failure booklet.



## Inappropriate sinus tachycardia

Inappropriate sinus tachycardia (IST) is a condition in which an individual's resting heart rate is abnormally high – greater than 100 beats per minute or rapidly accelerating to over 100 beats per minute without an identifiable cause for the tachycardia, although small amounts of exercise, emotional or physical stress are triggering factors. An ECG will not show any abnormalities (other than an abnormally fast heart rate), as IST arises within the sinus node where normal sinus rhythm is generated i.e. the heart rhythm is arising from the normal location but at an inappropriately high rate. It is a relatively new disorder (first recognised in the late 1970's) that is underappreciated by many in the medical profession and with many doctors regarding it as a psychological condition. Individuals with this condition can find themselves increasingly disabled and experiencing high levels of anxiety.

### Symptoms of IST

IST is primarily experienced by young women in their thirties, who may have been suffering symptoms for a few years. The main symptoms of IST include palpitations, shortness of breath, exercise intolerance, fatigue, resting heart rate of greater than 100bpm, sleeping heart rate of 70 – 90bpm, heart rate will rapidly reach 150bpm upon minimal exertion. Potentially related symptoms include a drop in blood pressure upon standing, blurred vision, dizziness, syncope, pre-syncope and sweating.

### What causes IST?

Unfortunately, to date nobody knows. There is a belief that IST is a result of the sinus node having an abnormal structure. There is another view that individuals with IST might be supersensitive to adrenaline as the smallest amount of exertion can cause a pronounced rise in the heart rate. However, a number of informed medical professionals believe there are a number of factors and disorders which point to disturbance within the autonomic nervous system. It is for this reason that it is a condition that can be mistaken for, or overlap with, postural tachycardia syndrome (PoTS). Please contact Arrhythmia Alliance for further information.

## Diagnosis of IST

The following guidelines may help towards securing the correct diagnosis for an individual presenting with IST symptoms.

1. ECG for resting heart rate. A resting heart rate will generally exceed 100bpm to be considered IST. A 24hr ECG/Holter monitor will determine if the average heart rate during a 24 hr period is under 95bpm. When an individual is lying down or sleeping there should be a little reduction in their heart rate.
2. Exercise/stress test - demonstrate an inappropriate heart rate response to exercise
3. Symptoms documented should indicate tachycardia
4. Other known causes of sinus tachycardia must be excluded - these include anemia, hyper thyroidism, pheochromocytoma, diabetes induced autonomic dysfunction, fever, and dehydration
5. Sometimes an EP Study will be performed to exclude atrial tachycardia before an IST diagnosis is finally made
6. Echocardiogram – to rule out any structural abnormality of the heart

## Treatment options

IST doesn't shorten life expectancy, therefore treatment is aimed at alleviating symptoms. Patients can choose to do nothing, and live with the symptoms if they are not that severe or have little impact on their life. They can take medications to try and slow the heart rate, or have an ablation procedure to destroy the heart tissue responsible for the tachycardia.

## Medication

It is a matter of trial and error to see if a helpful medication can be found for each individual. Beta blockers are often tried first; calcium channel blockers (diltiazem or verapamil) are an alternative. Where there is an overlap with autonomic dysfunction, fludrocortisone, midodrine, and serotonin-reuptake inhibitors have all been used. More recently drugs such as ivabradine have been tried, but with varying results. It is important not to make things worse with side effects from the drugs, which may be worse than the symptoms of IST itself.

## **Ablation**

Ablation of the sinus node (either to modify the node or destroy it completely) has been used with some success, but this is very variable and may not be long-lasting, and ablation carries its own risks. Ablation is a surgical procedure that uses either heat or freezing methods to destroy areas of the heart tissue via a catheter (thin, flexible tubes passed through blood vessels to the heart). Scar tissue is deliberately created in an attempt to prevent the tachycardia. It is best to discuss this with an electrophysiologist who specialises in this form of treatment. For further information, please see our booklet catheter ablation for SVT.

## **Self help**

Cognitive behavioural therapy (CBT) has had some success in helping patients come to terms with IST, and help manage their lives. It is very important to recognise that, however disabling symptoms may be, this is not a life-threatening disorder and lifespan is normal; with no increased risk of strokes or heart attacks. The number of people who develop any problems with their heart function as a result of a long-term fast heart rate is also very small. So, in some people, the best thing is to wait and see how things develop – it may improve on its own, particularly once a person has been reassured that there is no other sinister problem. Where there is an overlap with autonomic dysfunction, increasing salt and fluid intake on medical advice may help.

## **Important information**

Where sinus tachycardia is identified it is important to rule out other treatable conditions before making the diagnosis of IST – it may be that there is a curable cause. IST is not a life-threatening condition but symptoms can be very disabling. A variety of therapies exist, but it is important not to make things worse with any treatment. The opinion of a specialist, usually an electrophysiologist, can be helpful.

## Junctional ectopic tachycardia (JET)

Junctional ectopic tachycardia (JET) is a rare heart rhythm abnormality which occurs more in childhood than adult life. The most frequent time that it happens is immediately after surgery for congenital heart disease. However, it can occur as an isolated congenital phenomenon or after ablation for atrioventricular nodal re-entry tachycardia.

JET is characterised by a very fast heart rate (tachycardia) that originates within or very close to the atrioventricular (AV) node. The AV node is part of the heart's conduction system that usually slows down the naturally faster atrial (top chamber) rate as the impulse travels down to the ventricles. The result is the ventricles contracting at a much higher rate than normal.

### Treatment options for JET

The rate of this rhythm is temperature sensitive and will slow down with cooling and speed up if a child is hot. Therefore, one option is to use cooling blankets to prevent fever. A target body temperature of 35 degrees celsius is often helpful. It is also important to maximise pain relief and sedation keeping the child as calm and comfortable as possible. The rhythm can be resistant to medicines, but intravenous amiodarone is often used to slow the heart rate.

The rhythm results in dissociation of the top and bottom chambers of the heart and pacing techniques can be used to restore synchrony again. The temporary pacing wires applied routinely by the surgeon can be used for this. Alternatively surgical ablation of the responsible heart tissue can terminate the arrhythmia. A child can be very critically unwell whilst this rhythm continues, and supportive measures may be required to maintain adequate pumping action and output of blood from the heart.

### Prognosis

The rhythm usually exists for 3-5 days following surgery but then spontaneously resolves. It has been known to continue for up to 10 days. Once it has resolved it will not usually return and long-term medication is not required.

## Long QT Syndrome

Long QT syndrome (LQTS) is a heart rhythm disorder that can cause a disturbance in the electrical system of the heart. It is often the result of inheriting an abnormal gene which causes an imbalance in molecules that control the electrical impulses of the heart. LQTS can also be caused by medications for other conditions.

Electrically-charged molecules (ions) such as potassium, sodium and calcium, cross heart cell membranes through specialised ion channels. This generates the electrical activity (depolarization and repolarization) that initiates the heart pumping blood to the rest of the body. The abnormal function of one or more ion channels in LQTS prolongs the repolarization process which predisposes patients to cardiac arrhythmias.

LQTS may result in a very fast abnormal heart rhythm (arrhythmia), known as 'torsades de pointes'. When these rapid heartbeats occur, no blood is pumped out from the heart and the brain quickly becomes deprived of oxygen, potentially resulting in a loss of consciousness (syncope) and rarely, sudden 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 of long QT syndrome

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 include:

- 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.

## Causes of long QT syndrome

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 twelve 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.

### **What are the risks?**

LQTS is a rare condition, experts suggest that approximately 1 in 2,000 people are affected but this is not certain as it may never be diagnosed. You may be at risk if anyone in your family has had unexplained fainting episodes or seizures, or an unexplained sudden cardiac death. You are at increased risk if you have a first-degree relative with known LQTS. You are also at risk if you are taking any medications that prolong the QT interval. Your doctor can inform you whether any of your medications can do this. Certain metabolic conditions can also cause prolonged QT interval. People with low potassium, magnesium or calcium blood levels; such as those with the eating disorder anorexia nervosa, or have had diarrhoea or vomiting, are also at increased risk of prolonged QT interval.

### **Diagnosis of long QT syndrome**

An ECG may reveal a long QT interval which may suggest 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.

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, an exercise tolerance test. 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 non-exercise (medication) stress test is 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.

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 for long QT syndrome**

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). 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.

A more extensive publication on LQTS is available from Arrhythmia Alliance.



## Supraventricular Tachycardia (SVT)

Supraventricular tachycardia (SVT) is a rapid abnormal heart rhythm that begins in the upper chambers of the heart. The atria are above the ventricles, hence the term supraventricular. The term tachycardia refers to a rapid heartbeat of over 100 beats per minute. Typically, patients have varying degrees of symptoms, but occasionally they may have no symptoms. Palpitations are a common symptom during SVT or a sensation that the heart is beating rapidly, fluttering, or racing. This may last for a few seconds or several hours. Occasionally, you may feel short of breath or feel a pressure or pain in your chest. Sometimes patients will feel light headed or dizzy, and may lose consciousness (also known as syncope) but this is a rare occurrence. Although such symptoms may raise concern, in general, the majority of SVTs are not life threatening. Nonetheless, if any of these symptoms develop, immediate medical attention should be sought.

### Symptoms you may experience during SVT

Palpitations, chest pain, light headedness, shortness of breath, dizziness and rarely a loss of consciousness.

### How is SVT diagnosed?

An electrocardiogram (ECG) provides a tracing of the heart rhythm and is recorded by placing stickers and leads on the chest and limbs. If the patient is experiencing SVT during the ECG, a clear diagnosis can be made. A 24-hour ECG (heart monitor) may be used to record the heart rhythm continuously.

### What causes SVT?

SVT results from a problem within the electrical system of the heart, causing a very fast heartbeat. It may be caused by an extra electrical connection present from birth (wolff -parkinson-white syndrome), or it may develop in an otherwise normal heart. It can be triggered by extra (ectopic) heart beats, alcohol, caffeine, stress, or cigarette smoking.

## What treatment options are available for SVT?

Once SVT is diagnosed your doctor or nurse specialist will discuss your treatment options. If your symptoms are very infrequent, you may decide to have no treatment for your SVT, and your doctor and nurse specialist will advise you if this is an appropriate course of action. Medications may be used to treat patients with SVT. Your doctor will discuss with you the most commonly used medications, the benefits and side effects. A procedure called an electrophysiology study allows the doctor to detect any abnormalities in the electrical system within your heart that have been causing your symptoms. Once the abnormality is found the doctor may then perform a catheter ablation as an alternative to medications for some patients with SVT. This aims to cure the abnormal heart rhythm by destroying the area of extra cells which is causing the palpitations.



## What can I do if I develop SVT?

Fast heart rhythms that come on suddenly can often be stopped by performing some simple tricks called physiological manoeuvres. These are easy and safe to perform in any setting and may stop the fast rhythm and return the heart to normal. This helps avoid having to go to a hospital or call an ambulance. If you feel unwell when SVT begins, for example you have (bad) chest pain, feel very faint or find breathing difficult, call for an ambulance without delay. It may be useful to keep a record of how frequent your SVT occurs. If you find that your symptoms worsen or the method you use to stop them no longer works, do not worry. Talk to your GP who should be able to recommend a heart rhythm specialist that you can be referred to, to discuss further treatment options.

## Syncope

Syncope is the medical term for a blackout caused by a sudden lack of blood supply to the brain. There are many causes of syncope, some common and some rare. Reflex syncope is the most common form of syncope, which is sometimes called vasovagal syncope. In reflex syncope, blood that should be going to the brain is diverted elsewhere in the body, mostly to muscles. In an upright person, the brain quickly shuts down and consciousness is lost.

Autonomic sympathetic nerves pass from the brain to blood vessels that maintain blood pressure. In reflex syncope a reflex suddenly withdraws the effect of these nerves, and blood pressure falls abruptly. Another part of this reflex involves the vagus nerve from the brain. When the vagus nerve stimulates the heart, it slows down the number of beats per minute. The activation of the vagus nerve can be very abrupt, causing the heart to slow suddenly or even stop.

A fall in blood pressure and heart rate results in a fall in brain blood supply. At very low levels this can cause dizziness, visual disturbances and blackouts. These symptoms are most likely to be a problem when a person is upright (due to gravity further lowering blood pressure), but other factors such as food, heat, exercise, sight of blood and emotional stress can bring it on.

Syncope is a symptom and not a condition in itself and is too often assumed to be epilepsy. Sometimes syncope is the only symptom of an arrhythmia or another condition. Syncope can be a symptom of various other conditions such as postural orthostatic hypotension (PoTS), reflex anoxic seizures (RAS) and many other arrhythmias. If you have any doubt, then please contact STARS on 01789 867503 or email [info@stars.org.uk](mailto:info@stars.org.uk) for the blackouts checklist and other information which has been written specifically to help doctors and patients reach a correct diagnosis for an unexplained loss of consciousness.

## **Ventricular fibrillation (VF)**

Ventricular fibrillation (VF) occurs in the ventricles, in the bottom chambers of the heart. In VF, the electrical impulses are fired from multiple sites in the ventricles in a very fast and irregular way, causing the heart to quiver rather than to beat and pump blood effectively. VF is an extremely dangerous heart rhythm and prompt emergency care must be provided to get the heart pumping again, or death can occur.

## **Ventricular tachycardia (VT)**

Ventricular tachycardia (VT) occurs when the electrical impulses arise in the ventricles, causing them to beat at an abnormally fast, regular rate. Because the ventricles are beating rapidly the heart does not work as efficiently. This can cause symptoms of weakness, dizziness, chest pain, shortness of breath or even collapse. There are several different types of VT and the seriousness of the condition can vary. However, VT can be a potentially life-threatening heart rhythm as it can progress to ventricular fibrillation and cause the heart to stop beating (cardiac arrest). There are a number of reasons why people may develop VT. For example, in people who have had a previous myocardial infarction (heart attack), the area of the heart muscle damaged by the heart attack forms scar tissue and this can make the heart susceptible to abnormal heart rhythms. Other people who may experience VT are patients with cardiomyopathy, previous corrective congenital heart surgery or inherited arrhythmias. There is also a small group of people who have VT with a structurally normal heart where the VT may be well tolerated.

## Wolff -Parkinson-White syndrome

The heart has its own electrical conduction system which sends signals throughout the heart muscle to make it beat in a regular rhythm. Sometimes an extra pathway exists, which can cause the heart rhythm to change. This extra electrical pathway may be a reason that the heart can cause the heart to beat too quickly (tachycardia), or irregularly which may, in some cases, cause a very fast unstable heart rhythm disorder.

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 tachycardia. 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 to the ventricles. If the electrical impulses travel down the accessory pathway, and bypass 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.

### Symptoms of Wolff -Parkinson-White Syndrome

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, racing heart beat (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 of Wolff -Parkinson-White syndrome

Symptoms due to WPW usually occur randomly without any identifiable triggers. Sometimes caffeine, alcohol, or strenuous exercise can bring about symptoms.

## Diagnosis of Wolff -Parkinson-White syndrome

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.

## What is an 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 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.

## Treatment options

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. See our leaflet on catheter ablation for SVT for further information.

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.

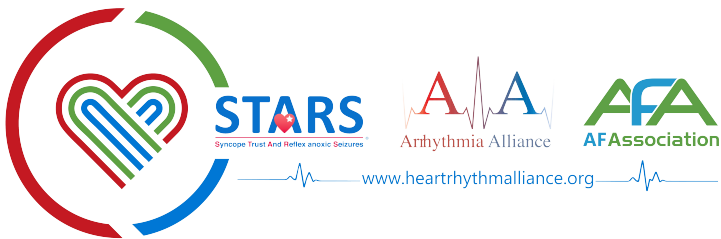
Your doctor/nurse specialist may recommend drug treatment prior to any invasive treatment. This may be an antiarrhythmic drug, such as flecainide or a beta blocker, which both help to prevent fast heart rhythms and slow down the action of the accessory pathway. With regard to lifestyle modifications,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

For more information, please see the WPW syndrome leaflet.



This booklet has been written to support patients with an arrhythmia who struggle to find information on these debilitating conditions. Without donations and fundraising, we would not be able to provide support through our award-winning resources and helpline.

Please donate to support our vital work at  
[www.hearhythmalliance.org/aa/uk/get-involved/donate](http://www.hearhythmalliance.org/aa/uk/get-involved/donate)



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



+44 (0)1789 867 501

info@hearhythmalliance.org

www.hearhythmalliance.org

Registered Charity No. 1107496

©Arrhythmia Alliance

©AF Association

©STARS

Published July 2017  
Reviewed August 2022

endorsed by  
**DH** Department  
of Health



**Finger on your Pulse:** is our new library of educational video resources. Medical Experts share their knowledge and address specific concerns and patients share their experience living with the various conditions and treatments.

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

**I had been having heart palpitations and was getting so worried - I downloaded this booklet, it has so much information - a marvelous resource!**

**Veronica in South Wales**

Please remember that this publication provides general information. You should always discuss and seek advice from your healthcare professional what is most appropriate for you.

**Acknowledgments:** we would like to thank all those who helped in the development and review of this publication. Particular thanks are given to Dr Charlotte D'Souza Angela Griffiths and Dr Jullian Ormerod.

**Founder and Trustee:**  
Trudie Lobban MBE, FRCP (Edin)

If you would like further information or would like to provide feedback please contact [info@hearhythmalliance.org](mailto:info@hearhythmalliance.org).