The Canadian SADS Foundation

Supporting Families.
Saving Lives.

Cardiac Channelopathies

An Information Booklet for Patients and Their Families

Dear Patient/Family Member,

When you hear a diagnosis of a cardiac channelopathy, you might feel confused and helpless. Most people have never heard of these conditions, and finding information can be difficult.

The purpose of this booklet is to explain the diagnosis and treatment of channelopathies. The booklet will also prepare you for the practical and emotional issues that can arise as you learn to live with one of these conditions. When reading this booklet, it is important to realize that each person with a channelopathy is different, and your cardiologist will recommend the best course of treatment or lifestyle changes for you.

We hope that this publication will help you to realize that you are not alone. In the "References" section there are many websites that provide useful information and links to groups that offer emotional support.

We sincerely hope that this will be useful to you and your family.

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What are Cardiac Channelopathies?

Cardiac channelopathies are a group of **syndromes** that affect the electrical system of the heart. In the majority of patients with channelopathies, the heart's structure is normal (Figure 1). In a normal, healthy heart, the heart chambers contract rhythmically, pumping blood throughout the body. Electrical signals that originate in the electrical system cause the heart's muscle cells (**myocytes**) to contract. These electrical signals depend on the flow of sodium, potassium, and calcium **ions** into and out of the cells. The ions pass through **ion channels** (sometimes called "protein channels") in the cell membranes to create the electrical signal that generates each heart beat. If the ion channels do not work properly, the electrical stability of the heart and heart rhythm may become abnormal. Abnormal heart rhythms (**arrhythmias**) can lead to fainting or **cardiac arrest** and sudden death.

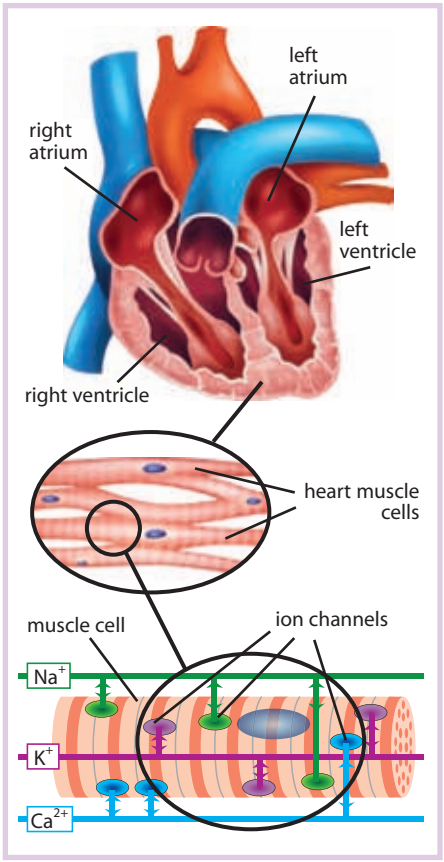

Figure 1: The heart and its muscle cells

A cardiac arrest is not the same as a heart attack, although both can be fatal. A cardiac arrest is a sudden stoppage of the heart for electrical reasons. A heart attack is a lack of blood flow to the heart muscle, usually because of disease in the arteries.

The cause of the abnormal function of the ion channels is usually inherited. This means that one of the genes that encode the proteins is not normal. If this gene is passed from parent to child, an abnormal ion

channel forms. Rarely, a **mutation** can occur spontaneously. In these cases, there will be no family history of the condition.

Most deaths due to inherited channelopathies are preventable if the conditions are diagnosed. The diagnosis of a cardiac channelopathy should be suspected in any young person with unexplained **syncope** (sudden loss of consciousness or fainting), cardiac arrest, or sudden death. Syncope is the primary symptom and may be the only warning sign of heritable cardiac channelopathies. Not all affected people will exhibit symptoms. The lack of prior symptoms, therefore, does not necessarily mean that a patient or family does not have a heritable channelopathy. However, many people who die suddenly from channelopathies had previous symptoms that were ignored, misdiagnosed, or considered insignificant. If you have any of the symptoms, it is very important that you report them to your family doctor.

A diagnosis of cardiac channelopathy should be considered in any of the following situations:

- A young person with unexplained syncope or seizures, especially in the setting of exercise or emotional stress
- A survivor of unexplained cardiac arrest
- A family history of unexplained syncope, seizures, or sudden death. A death is considered to be "unexplained" when an autopsy fails to find any obvious cause of death.

If you have any suspicious symptoms, it is always best to seek medical attention for basic screening. This is especially true when a faint is suspected to be secondary to an arrhythmia. These "warning" faints are usually sudden and unexpected, occurring during physical exertion or emotional stress. In contrast, a common faint usually has a fairly obvious cause, such as pain, injury, or an unpleasant experience, and is preceded by warning symptoms such as dizziness, blurring of vision, tingling, nausea or sweating.

When an individual has been diagnosed with a heritable arrhythmia, all immediate family members (parents, siblings, children) should also be evaluated. They should undergo routine testing such as ECG, Holter, and treadmill testing. If the affected family member has been identified with a specific mutation, also consider genetic testing.

Once identified, these diseases can often be very well managed. For the most part, sudden death can be prevented and you can continue living a full life.

There are many different types of genetic mutations that ultimately lead to a variety of cardiac channelopathies. These channelopathies may result in a number of syndromes, including

- Brugada Syndrome (BrS)
- Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)
- Long QT Syndrome (LQTS)
- Short QT Syndrome (SQTS)

In most cases of these conditions, simple preventative drug treatments control the majority of the symptoms. However, some individuals or families benefit from keeping an **automated external defibrillator (AED)** in the home. Discuss this suggestion with your physician. The following sections provide more detailed information about each of the four most common channelopathies.

Brugada Syndrome (BrS)

What is BrS?

Brugada Syndrome (BrS) is an inherited channelopathy that results in abnormalities in the electrical state of the heart muscle. The hallmark of this condition is the characteristic ECG finding that reflects the electrical abnormality. Individuals with BrS are at risk of arrhythmias that may result in syncope or sudden death. BrS affects more men than women. The average age at which symptoms appear is 40 years; although symptoms have been reported in patients aged from newborn to 84 years.

What causes BrS?

BrS is an inherited disease resulting from mutations in one or more of several different ion channel genes. Genes carrying instructions for the sodium and calcium channels in the heart muscles are most often affected. Although some specific genetic defects leading to BrS have been identified, it is likely that there are other mutations that have not yet been discovered.

What are the symptoms of BrS?

Syncope and **cardiac arrest** are the most common indications leading to a diagnosis of BrS. Cardiac arrest occurs most often during sleep or rest, rather than during physical activity. There may be a history of trouble breathing at night. Fever and medications appear to trigger or worsen the symptoms of Brugada Syndrome in many. Some patients, however, show no obvious symptoms. A family history of **sudden cardiac death** may be present, though not universal, as the **syndrome** can occur in an individual with no known family history of the disease.

How is BrS diagnosed?

Your physician may suspect that you have BrS based on your symptoms and/or family history. Characteristic patterns on an **electrocadiogram (ECG)** can also suggest the diagnosis, or be used to confirm it (Figure 2). These patterns may be present all the time, may appear and then normalize spontaneously, or may be brought on by fever or the administration of particular drugs. BrS may be discovered only when a routine electrocadiogram (ECG) shows an abnormal pattern known as

ST segment elevation. Many individuals have other ECG abnormalities such as heart block or **atrial fibrillation**. (Figure 3).

BrS is sometimes difficult to diagnose. Many people who have a BrS gene may never experience any warning symptoms. The electrical malfunction may or may not show up on an ECG.

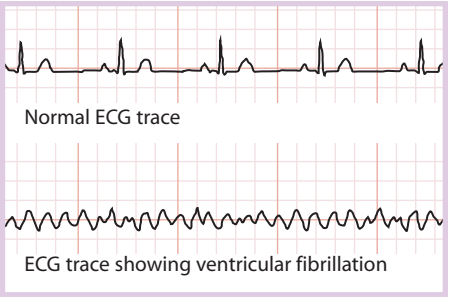

Figure 2: Electrocardiograph comparing a normal reading and a reading indicating ventricular fibrillation

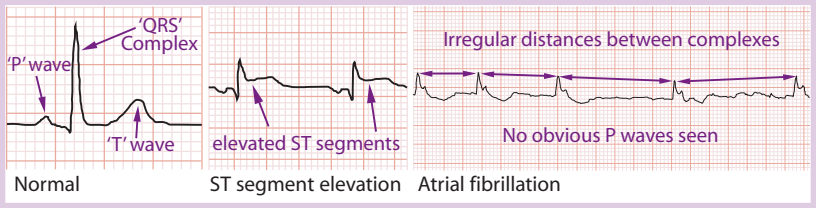

Figure 3: ECG traces showing a normal heartbeat, ST segment elevation, and atrial fibrillation

Genetic testing for BrS

The yield of **genetic testing** remains relatively low: only 11–28% of those with a diagnosis of BrS are found to have an identified BrS gene. Genetic testing may help in the early identification of family members at potential risk. If you have had a positive clinical diagnosis of BrS, your children, siblings, and parents should be evaluated.

What treatments are available for BrS?

There is no cure for BrS, and there are no treatments. However, there are ways of reducing the risk of arrhythmia and sudden death. For example, many prescription drugs, over-the-counter medications, and drugs used in dental procedures can increase your risk of arrhythmia and should be avoided. Discuss the use of these potentially dangerous medications with your cardiologist. In addition, fever can provoke electrical changes in the heart in BrS, and fever must be aggressively managed with antipyretics (acetaminophen or ibuprofen). If the fever does not respond to antipyretic medications, cardiac monitoring is

important until the fever resolves. Since the Brugada ECG can be easily misinterpreted as a "heart attack", it is important that you carry a copy of your ECG whenever you seek medical attention, and inform health professionals of your condition.

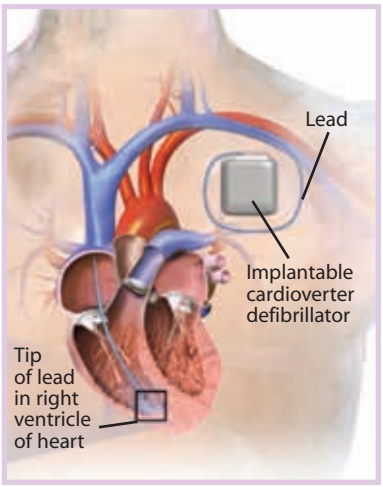

Figure 4: An ICD placed in the upper chest, with wires running into the heart

If you have symptoms such as syncope, seizures, trouble breathing at night, or cardiac arrest, your physician may recommend a surgically placed device called an **implantable cardioverter defibrillator (ICD)** (Figure 4). An ICD monitors the heart's activity and, when arrhythmia occurs, this device is able to restore normal rhythm with pacing or an electric shock. There is not universal agreement about how to predict risk in Brugada syndrome. In some specific cases, your doctor may recommend a procedure called an **electrophysiology study (EPS)**.

What lifestyle changes should I make after being diagnosed?

Not all medical professionals agree on whether patients with BrS should participate in high intensity physical activities. You should discuss this issue with your cardiologist.

Figure 4: Image is from *Blausen.com staff. "Blausen gallery 2014". Wikiversity Journal of Medicine. DOI:10.15347/wjm/2014.010. ISSN 20018762. - Own work* (Image was modified by removal of call-out indicator lines and addition of new ones)

Catecholaminergic polymorphic ventricular tachycardia (CPVT)

What is CPVT?

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is characterized by abnormal heart rhythms (arrhythmias), which can lead to syncope or cardiac arrest. These events usually occur during physical exercise or with emotional stress. The average age at which symptoms of CPVT appear is between seven and nine years, although symptoms may appear as late as the fourth decade of life. If untreated, this condition can be lethal. Up to 80% of diagnosed individuals experience one or more syncopal spells and approximately 30% experience cardiac arrest. CPVT is thought to be far less common than other channelopathies, but does affect otherwise healthy infants, children, adolescents, and adults. The true prevalence of CPVT remains unknown but is estimated to be about 1 in 10,000. With increased awareness, genetic testing, and effective treatment options, doctors can now diagnose CPVT early and provide treatment to prevent sudden death.

What causes CPVT?

CPVT is an inherited channelopathy resulting from mutations in one or more of the genes that handle calcium inside the heart muscle cells. Arrhythmias can be triggered by conditions involving emotional or physical stress that leads to a release of **adrenaline**.

What are the symptoms of CPVT?

Patients with CPVT may experience **palpitations**, seizures, or syncope when the body is producing high levels of adrenaline, such as during exercise or in times of stress. CPVT is suspected when an individual has an episode of syncope, seizures, or cardiac arrest associated with exercise or emotional stress. CPVT is also suspected when the autopsy is normal following the sudden and unexpected death of a young person.

How is CPVT diagnosed?

Testing for CPVT includes a resting ECG (which is usually normal) and an exercise stress test (walking on a treadmill or riding a bicycle while your heart rhythm is recorded with an ECG) to try to provoke the rhythm abnormalities. Children that are too young for a treadmill or

bicycle test can be monitored during routine activities by wearing a **Holter monitor** (Figure 5). An alternative to treadmill **stress testing** in someone who cannot exercise is **catecholamine provocation testing**, in which adrenaline is administered intravenously while the heart rhythm is monitored. A diagnosis of CPVT is made when the characteristic arrhythmias are observed. These include frequent premature ventricular beats, and ventricular arrhythmias such as bidirectional **ventricular tachycardia**, which may lead to **ventricular fibrillation**.

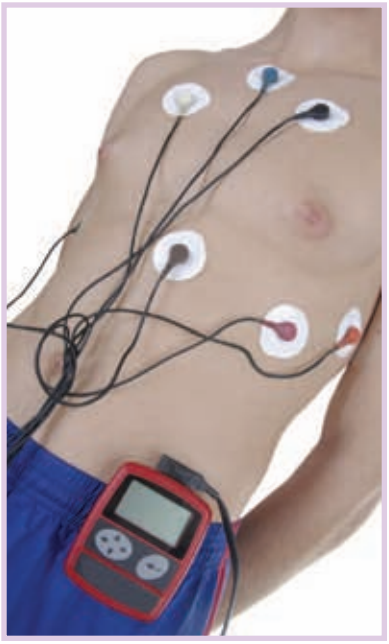

Figure 5: A Holter monitor

Genetic Testing for CPVT

There are two known genes that cause CPVT. Both affect males and females equally. The most common CPVT gene is dominant while a rare form is recessive (Figure 6). Once a family member is identified with CPVT, other immediate family members should seek testing for the syndrome.

What treatments are available for CPVT?

There is no cure for CPVT, but there are effective treatments. All patients who have CPVT symptoms should receive treatment. All children and young adults with CPVT should be treated even if they do not have symptoms. This is because symptoms might occur at any time, with sudden death sometimes being the first symptom.

The usual treatment involves taking a medication called a **beta-blocker** every day. Beta-blockers prevent the response of the heart to adrenaline, reducing the likelihood of arrhythmia. Repeated treadmill stress tests may help to determine the correct dose. If the beta-blocker does not adequately control arrhythmia, an additional medication, Flecainide, has also been found to be useful. These medications are

usually very successful at controlling arrhythmias if they are taken regularly. You will require regular follow-up with your physician to be sure that your arrhythmia is well controlled. Growing children need their medication dose adjusted regularly. You must report any recurring symptoms.

If you continue to have symptoms in spite of appropriate doses of medication, there are other treatments. One treatment is left cardiac sympathetic denervation (LCSD). This is a surgery in which the nerves that are responsible for adrenaline delivery to the heart are removed. Rarely, an ICD is needed.

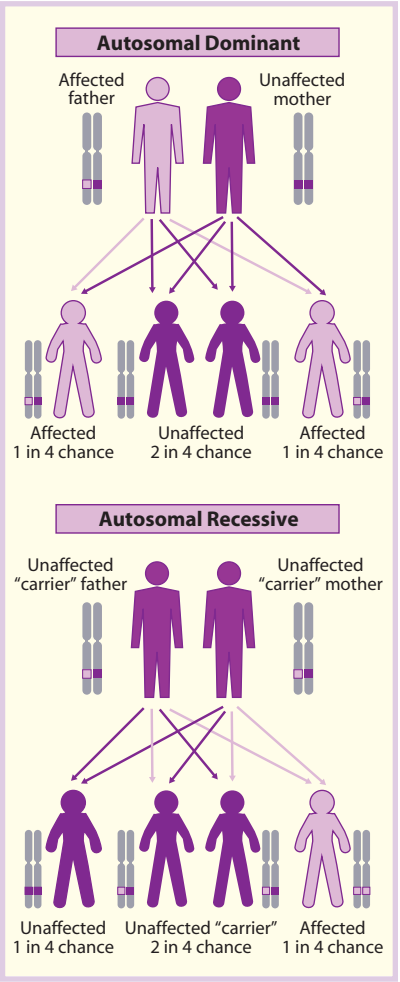

Figure 6: Inheritance pattern of the two CPVT genes

What lifestyle changes should I make after being diagnosed?

It is very important that you take your medications every day. Parents should teach their children about the importance of daily medication, and should make sure that each daily dose is taken. Preteens and teenagers should also be supervised when taking their medication. Because strenuous physical exercise increases the risk of arrhythmia, you should avoid competitive or high-intensity activities. Your cardiologist will discuss those activities with a lower risk of arrhythmia.

You will also be asked to avoid using medications that may stimulate the heart, such as over-the-counter cold medications or some asthma inhalers.

Long QT Syndrome (LQTS)

What is LQTS?

Long QT Syndrome (LQTS) is a heart condition that affects about 1 in 2500 people. In this condition, the electrical recovery of the heart muscle after each heart beat can take longer than normal. This is evident on the ECG as prolongation of the QT interval. (Figure 7).

What causes LQTS?

LQTS is an inherited channelopathy resulting from mutations in one or more of the genes that handle the movement of ions in and out of the heart muscle cells. This abnormality can be amplified by certain medications.

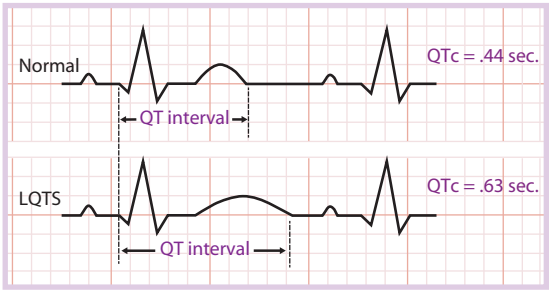

Figure 7: ECG comparing a normal reading and a reading indicating LQTS

What are the symptoms of LQTS?

Syncope is the most common symptom of LQTS. Other symptoms can be palpitations, seizures or even sudden death. Patients with LQTS may experience symptoms when the body is producing high levels of adrenaline, such as during exercise or during times of stress. Symptoms can also be triggered by a sudden surprise or a loud noise, or even occur during rest or sleep. Most people who experience symptoms of LQTS have their first episode by the age of 40, but symptoms may be seen in infancy or later in life. Many people with LQTS become aware of their condition only from the results of an ECG performed for an unrelated reason, because they have a family history of LQTS, or because of genetic test results.

How is LQTS diagnosed?

Testing for LQTS includes a resting ECG (which may be normal) and an exercise stress test: walking on a treadmill or riding a bicycle while your heart rhythm is recorded with an ECG (Figure 8). These tests allow your cardiologist to observe the electrical behavior of the heart at different heart rates. An alternative to treadmill stress testing for

someone who cannot exercise is catecholamine provocation testing, in which adrenaline is administered intravenously while the heart rhythm is monitored. A diagnosis of LQTS is made when the characteristic QT prolongation is observed.

Figure 8: Patient undergoing a stress test

Genetic testing for LQTS

At least 12 genes are associated with LQTS, and hundreds of mutations have been identified within these genes. A gene mutation can be identified in most people with a clinical diagnosis of LQTS. Hereditary LQTS is most commonly inherited as an autosomal dominant condition (Figure 6). In cases where the genetic test results for known mutations are negative, family screening can only be carried out with ECG and stress testing.

What treatments are available for LQTS?

There is no cure for LQTS, but there are effective treatments. Discuss treatment options with your cardiologist. All patients who have symptoms should receive treatment. In most cases, physicians recommend that all children and young adults with LQTS – even those without symptoms – should be treated. Symptoms might occur at any time, with sudden death sometimes being the first symptom.

The usual treatment involves taking a medication called a **beta-blocker** every day. Beta-blockers prevent the response of the heart to adrenaline, reducing the likelihood of arrhythmia. Repeated treadmill stress tests may help to determine the correct dose. Beta-blockers are usually very successful at controlling arrhythmias if they are taken regularly. You will have regular follow up with your physician to be sure that your arrhythmia is well controlled. Growing children need their medication dose adjusted regularly. You must report any recurring symptoms.

If you continue to have symptoms in spite of appropriate doses of medication, there are other treatments. One treatment is left cardiac sympathetic denervation (LCSD). This is a surgery in which the nerves that are responsible for adrenaline delivery to the heart are removed. Rarely, an ICD is needed.

What lifestyle changes should I make after being diagnosed?

It is very important that you take your beta-blocker every day. Parents should teach their children about the importance of daily medication, and should make sure that each daily dose is taken. Preteens and teenagers should also be supervised when taking their medication. Because strenuous physical exercise increases the risk of arrhythmia, you should avoid competitive or high-intensity activities. Your cardiologist will discuss those activities with a lower risk of arrhythmia.

Many prescription drugs, over-the-counter medications, and drugs used in dental procedures can affect the QT interval and may increase your risk of arrhythmia. These drugs should be avoided. Discuss the use of these potentially dangerous medications with your cardiologist.

Depending on your specific ECG changes or genetic mutation, you may be advised to avoid loud, surprising noises such as alarm clocks or telephones in the bedroom.

The Canadian SADS Foundation has produced two comprehensive publications about Long QT Syndrome. If you would like to receive a complimentary copy of *LQTS: An Information Booklet for Patients and Their Families* or *Long QT Syndrome in Women*, please contact us.

Short QT Syndrome (SQTS)

What is SQTS?

Short QT Syndrome (LQTS) is a rare heart condition that carries a risk of arrhythmias that may result in syncope or sudden death. In this condition, the electrical recovery of the heart muscle after each heart beat is abnormal. This is evident on the ECG as shortening of the QT interval (Figure 9). SQTS appears to be rare but the condition may be underdiagnosed because some affected individuals never experience symptoms.

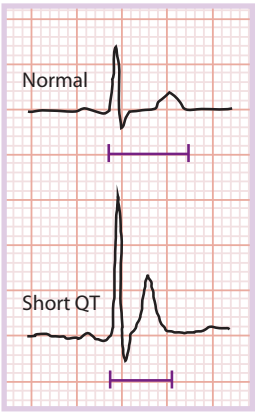

Figure 9: ECG showing normal and SQTS traces

What causes SQTS?

SQTS is an inherited channelopathy resulting from mutations in one or more of the genes that handle the movement of ions in and out of the heart muscle cells.

What are the symptoms of SQTS?

Syncope is the most common symptom of SQTS. Other symptoms can be seizures or even sudden death. Many individuals also have other ECG abnormalities such as heart block or atrial fibrillation. Many people with SQTS become aware of their condition only from the results of an ECG performed for an unrelated reason, because they have a family history of SQTS, or because of genetic test results.

How is SQTS diagnosed?

Testing for SQTS includes a resting ECG (which may be normal) and an exercise stress test: walking on a treadmill or riding a bicycle while your heart rhythm is recorded with an ECG. These test allow your cardiologist to observe the electrical behavior of the heart at different heart rates.

Genetic testing for SQTS

Six genes are associated with SQTS. A genetic mutation can be identified in about 25% of people with a clinical diagnosis of SQTS. In cases where the genetic test results for known mutations are negative, family screening can only be carried out with ECG and stress testing.

What treatments are available for SQTS?

There is no cure for SQTS, but there are effective treatments. Discuss treatment options with your cardiologist. All patients who have symptoms should receive treatment. In most cases, physicians recommend that all children and young adults with SQTS – even those without symptoms – should be treated. Symptoms might occur at any time, with sudden death sometimes being the first symptom.

The placement of an ICD is an effective first-line treatment to prevent sudden cardiac death. For arrhythmia management there are medications that can be used to prolong the QT interval, effectively reducing the risk of sudden death. These medications include quinidine, which is available in Canada if your physician makes a special request to use it in your case.

What lifestyle changes should I make after being diagnosed?

Not all medical professionals agree on whether patients with SQTS should participate in high intensity physical activities. You should discuss this issue with your cardiologist.

Glossary

adrenaline – a hormone, produced by the body, that has the effect of speeding up the heart; also called epinephrine

arrhythmia – a potentially lethal irregular heart rhythm

atrial fibrillation – irregular contraction of the atria of the heart

automated external defibrillator (AED) – a device that detects an abnormal heart rhythm (fibrillation) and sends and electric shock to stop the fibrillation and allow the heart to resume a normal rhythm

beta-blocker – a medication that treats abnormal heart rhythms by blocking the heart's response to adrenaline

cardiac arrest – a potentially fatal sudden stoppage of the heart due to a malfunction of the heart's electrical system

catecholamine provocation testing – a diagnostic technique to provoke bad heart rhythms that may not otherwise be expressed; IV administration of epinephrine (adrenaline) while a patient undergoes an ECG to monitor the heart's response to the drug

electrocadiogram (ECG) – a graph showing the electrical activity of the heart over time

electrophysiology study (EPS) – a medical test to investigate abnormalities of the heart's electrical system; involves the placement of electrodes into the heart

genetic testing – an analysis of an individual's genetic material with the purpose of identifying certain genes

Holter monitor – a diagnostic technique in which the electrical activity of a patient's heart is measured using a portable device for an extended period

implantable cardioverter defibrillator (ICD) – a device, implanted in the chest, that detects any arrythmia in the heart and administers an electric shock to stop the fibrillation and permit normal heart function to resume

ion – a charged particle of an element, e.g., sodium, Na^+; calcium, Ca^{2+}

ion channel – a protein in the cell membrane that moves ions in or out of the cell, thus maintaining an electrical potential difference across the cell membrane

mutation – a change in the sequence of genetic material that often affects the structure of proteins in the body

myocytes – muscle cells within the heart

palpitations – the sensation of an unusually fast heart beat

ST segment elevation – a feature of an electrocardiogram in which a portion of the trace is unusually high

stress testing – a diagnostic test in which the patient engages in physical activity (often on a treadmill) while undergoing electrocardiography to record the electrical activity of the heart

sudden cardiac death – unexpected death resulting from loss of heart function

syncope – sudden loss of consciousness with no apparent cause

syndrome – a collection of symptoms with a related cause

ventricular fibrillation – a disorganized contraction of the heart muscles that fails to produce a proper contraction of the ventricle, so blood is no longer pumped around the body

ventricular tachycardia – rapid contraction of the ventricles of the heart

Where can I find more information about channelopathies?

www.sads.ca The Canadian SADS Foundation website is a great place to start your search for more information. This website provides you with an opportunity to read the latest newsletter, sign up to be on The Canadian SADS Foundation mailing list, or learn about plans for a patient education conference in your geographical area.

www.sads.org The American SADS: information and support

www.chrsonline.ca Canadian Heart Rhythm Society: a resource to help you find a heart rhythm doctor in your area of Canada

www.cagc-accg.ca The Canadian Association of Genetic Counsellors: a resource to help you find a genetics counsellor in your area of Canada

www.cardiomyopathy.org Cardiomyopathy Association: a helpful British site with information about the cardiac and psychological effects of cardiac channelopathies

www.hrspatients.org Heart Rhythm Society: an American site about different cardiac arrhythmias with lots of information about ICDs and procedures

www.nlm.nih.gov/medlineplus MedlinePlus: a service of the U.S. National Library of Medicine; a good general reference for information on health prolems and medications

Notes

The Canadian SADS Foundation

Supporting Families.
Saving Lives.

Preventing Sudden Cardiac Death In Children and Young Adults

The Canadian SADS Foundation , a registered Canadian charity, is the only patient advocacy group in Canada dedicated to supporting families affected by inherited cardiac rhythm disorders and committed to raising awareness about the warning signs for these sometimes devastating disorders.

The Canadian SADS Foundation is committed to promoting awareness to health care professionals, educators, sports groups, and the general public and to providing information and support to families affected by inherited cardiac rhythm disorders.

It is estimated that as many as 50% of young people who experience a sudden cardiac death (SCD) had symptoms prior to their event. These symptoms may have been either misdiagnosed or dismissed as insignificant. Recognition of the warning signs and early medical intervention are the keys to preventing a SCD in children and young adults:

- **Fainting (syncope) or seizure** during physical activity.
- **Fainting (syncope) or seizure** resulting from emotional excitement, emotional distress, or startle.
- **Family history of unexpected sudden death** during physical activity or during seizure, or any other unexplained sudden death of an otherwise healthy young person.

A young person who has experienced any one of these warning signs should be referred to a cardiologist or an electrophysiologist for a complete cardiac assessment. This assessment should include an analysis of the heart rhythm and, where indicated, cardiac imaging and exercise testing.

For further information, please contact **The Canadian SADS Foundation** at www.sads.ca or call 1-877-525-5995.

Supporting Families.
Saving Lives.

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