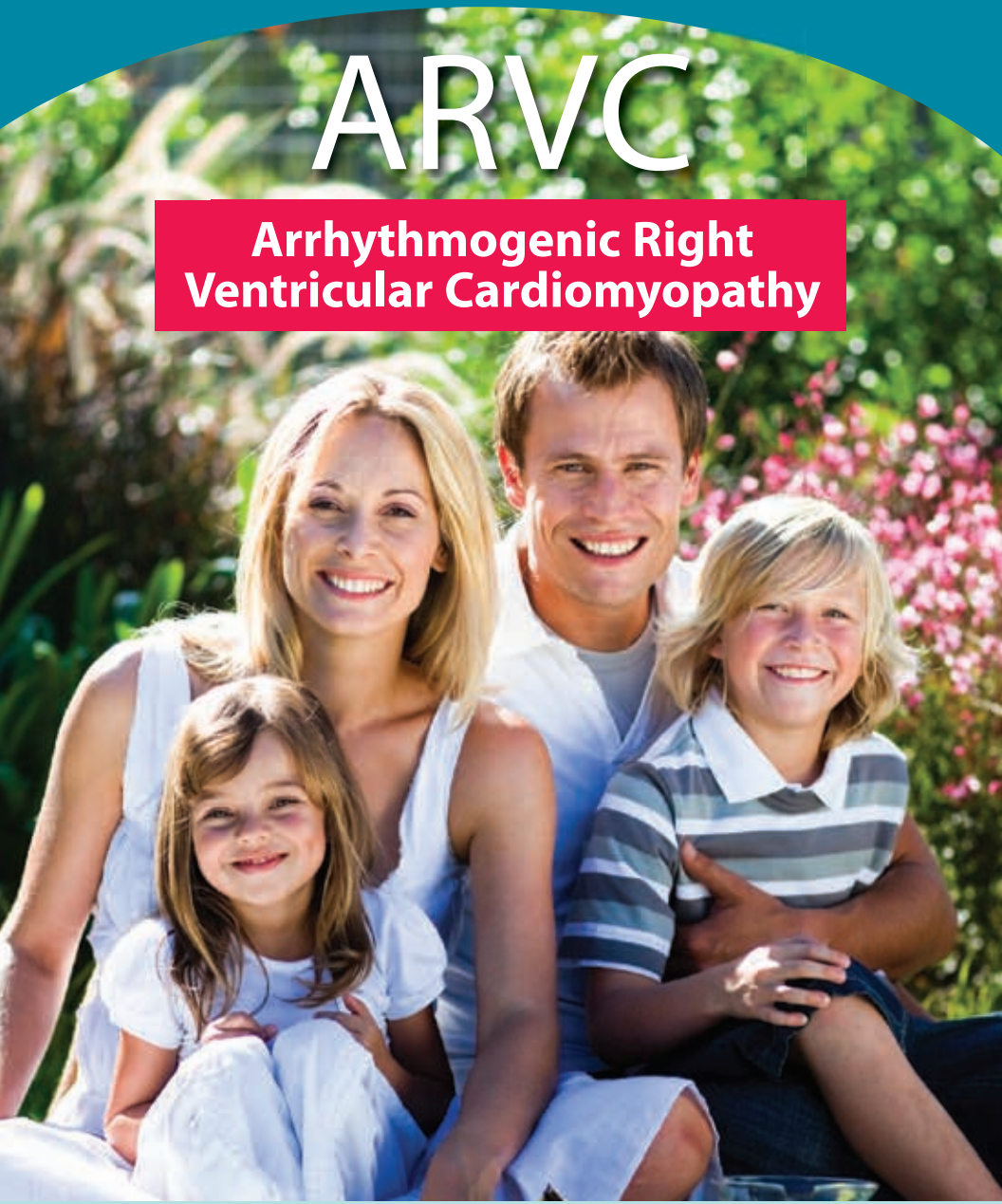
The Canadian SADS Foundation

Supporting Families.
Saving Lives.

ARVC

Arrhythmogenic Right Ventricular Cardiomyopathy



An Information Booklet for Patients and Their Families

Dear Patient/Family Member,

A diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) can trigger feelings of confusion and helplessness. Since most people have not heard of this condition, accessing information can be difficult. Parents often have many questions but aren't sure exactly what to ask their child's doctor. While a diagnosis of ARVC can be initially devastating news, with time parents and families can develop coping strategies. The purpose of this booklet is to explain some of the aspects of diagnosis and treatment of ARVC and to prepare you for the practical and emotional issues that can arise when learning to live with this condition. It is important to realize that each person with ARVC is different, and your cardiologist will recommend the best course of treatment.

We hope this publication will help you to realize that you are not alone. In addition to factual information, we have included perspectives from other people who are coping with a diagnosis of ARVC. On page 22, there is a list of several websites that provide constructive information. We sincerely hope that this publication will be useful to you and your family.

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What is Arrhythmogenic Right Ventricular Cardiomyopathy?

Arrhythmogenic right ventricular cardiomyopathy, ARVC, (formerly known as arrhythmogenic right ventricular dysplasia, or ARVD) is an increasingly recognized genetic disease of the heart muscle. Although most people with ARVC do not show any symptoms, the condition causes 15%-25% of heart-related deaths in people under 35 years of age. The exact prevalence of ARVC is unknown but is estimated to be between 1 in 5000 to as high as 1 in 1000 people. The condition is slightly more common in males than in females although the known genes are inherited equally between males and females. The symptoms of the disease mostly occur between the second and fourth decades of life.

In order to understand ARVC, it is important to first understand how the heart works. The heart is made up of four chambers: the left atrium, the right atrium, the left ventricle, and the right ventricle. These chambers are made of muscle cells that contract at the same time to pump blood through the body. Low-oxygen blood from the body enters the heart in the right atrium. From there it moves into the right ventricle, where it is pumped to the blood vessels of the lungs in order to collect oxygen to deliver to the body. High-oxygen blood from the lungs returns to the left atrium of the heart. It then moves to the left ventricle, where it is pumped to the rest of the body. The simultaneous contraction of the two heart ventricles creates the pressure necessary to move blood through the entire body. This contraction is controlled by an electrical current that is transmitted through pathways in the heart to all of the heart muscle cells.

The word cardiomyopathy is loosely translated as “heart-muscle-disease”, and arrhythmogenic means “causing the heart to beat irregularly”. Research conducted by physicians and scientists have discovered that ARVC is caused by a defect in the tiny proteins that hold the heart muscle cells together. These complex proteins or “desmosomes” can be disrupted in ARVC, leading to areas

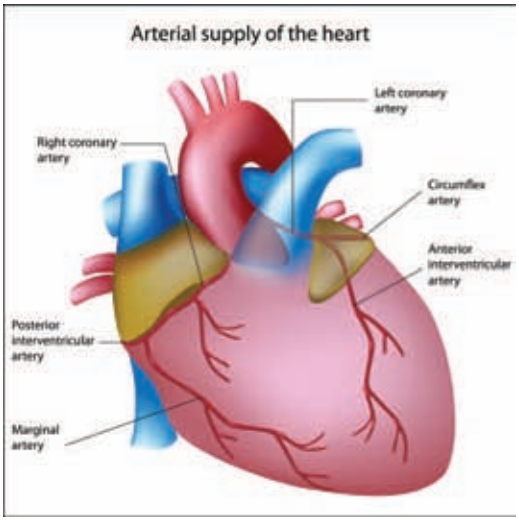
of scarring and fatty deposits. The right ventricle is more susceptible to this damage, though the left ventricle can also be involved. These areas of scarring give rise to abnormal heart rhythms which can lead to symptoms and sudden death.

One type of rapid heartbeat is called ventricular tachycardia. When ventricular tachycardia occurs, the heart is beating too fast to pump blood out of the heart effectively so the brain does not receive enough blood and the person may lose consciousness within seconds. This sudden loss of consciousness is called syncope (SIN-co-pee) and it may look like a simple faint or like a seizure. ARVC patients who collapse for this reason may recover on their own or they may require CPR (cardiopulmonary resuscitation) or defibrillation (an electric shock to the heart delivered by paddles) to start the heart beating normally again. Tragically, some patients never wake up because the abnormal beating of the heart cannot deliver enough blood to the brain and vital organs.

"I had been having "fainting" episodes since I was a teenager. I had never heard of ARVC or anyone else having a similar problem with fainting. One day at a track meet I collapsed during my race and my heart stopped. The first aid people at the track did CPR and I received an implantable defibrillator a week later."

– Erin

The first signs of ARVC may be subtle and are quite easily missed. This is why family members of people with ARVC must undergo extensive testing to check if they are affected by the condition. ARVC is thought to progress in several stages. The first stage is a "quiescent phase" where the patient has no symptoms, but still may have a risk of sudden death, particularly during heavy exercise. Patients experience symptoms in the second, or



"overt" phase. In this phase, the ventricles may have episodes of arrhythmia (irregular beating), which are perceived as dizziness, heart palpitations, and fainting. Other more nonspecific symptoms like fatigue, confusion, and abdominal pain may be present as well. More advanced ARVC causes generalized heart disease. In this later phase, symptoms of heart failure, such as swelling of the legs, fluid in the lungs, and shortness of breath, may develop. Because tests may be normal in patients in the first phase of ARVC, repeat testing of people at risk for ARVC is necessary. Early diagnosis is important because in some people the first symptom of ARVC is sudden cardiac arrest. Sudden cardiac arrest (SCA) is a condition in which the heart suddenly and unexpectedly stops beating. If this happens, blood stops flowing to the brain and other vital organs. SCA usually causes death if it's not treated within minutes. Treatment can reduce the risk of sudden death considerably.

"I have been surprised at the relief I feel when I tell other people about our son's heart problem. Everyone has been very helpful and supportive, and it has really helped us to cope with the diagnosis."

– Sarah

Symptoms

Patients may present with any one or more of the following symptoms:

- Palpitations (abnormality of heartbeat that causes a conscious awareness of its beating)
- Syncope (SIN-co-pee: loss of consciousness)
- Light-headedness
- Chest pain
- Cardiac arrest

Who should be assessed for ARVC?

- People with known relatives affected by ARVC
- People who had relatives who died suddenly at a young age (under the age of 40)
- People who have fainted or had seizures during exercise or emotional excitement

How it is diagnosed?

ARVC is a difficult condition to diagnose, and this leads to frustration on the part of patients, families, and their doctors. Even after many tests, your doctor may not be able to say for sure whether or not you or your child have ARVC. In order to make a diagnosis the cardiologist uses a list of criteria or scoring system (which was modified in 2010) to determine the likelihood of having ARVC or to make a clinical diagnosis. An important criterion in the formula is your family history, so it is helpful to examine your family tree closely to determine if there were any relatives who died suddenly at a young age, as well as relatives with unexplained heart arrhythmias. Testing for ARVC involves a combination of tests, some of which are painless and non invasive, and some that are more invasive. The number of tests you will have to undergo depends on how definitive the results of earlier tests were, and whether genetic testing is an option for your family. People at high risk will often be tested every year or two years to determine if signs of ARVC have developed over time. Many of these tests are not available at most hospitals, so you may have to travel to a larger centre to have them performed.

Electrocardiogram (ECG), SAECG, Holter Monitoring and Stress Testing:

An ECG machine measures the electric pattern of the heart through electrodes placed on the skin. The detailed ECGs used for the diagnosis of ARVC involve ten or more electrodes on the chest, arms, and legs.

SAECG is a special type of ECG used for detecting subtle abnormalities in the electrocardiogram (ECG) that are not visible to the naked eye. The SAECG is derived by computing the average of multiple ECGs. Some people with ARVC have abnormal ECGs when they are at rest, and sometimes the irregularity is only apparent when the person is feeling palpitations or lightheadedness. Since these episodes don't always happen at the doctor's office, you may be given a device to take home. Holter monitors are small ECG machines that you wear for 24 or 48 hours. They are useful because they can detect irregular heartbeats that may be occurring during your daily activities.

An exercise stress test will be part of diagnostic testing for ARVC. In this test, a person is hooked up to an ECG machine while they run on a treadmill or ride an exercise bicycle. This test can be useful in diagnosis because the heart is more likely to have an unusual rhythm when it is beating quickly.

Transtelephonic Monitors and Implantable Loop Recorders:

Unfortunately, stress and Holter tests may be normal in patients with ARVC. They are useful in ruling out other diseases that are similar to ARVC but cannot be used to arrive at a definite diagnosis. Cardiologist may choose to give a transtelephonic monitor or an implantable loop recorder to better assess the heart's rhythm over several weeks. Some transtelephonic monitors work like Holter monitors and use electrodes that are attached to the skin each day. Some of the newer models are simply pressed to the chest when a symptom occurs. The type of transtelephonic monitor you receive will depend on what is available in your hospital and the frequency and durations of your symptoms. ECG information collected by these devices can be transmitted over the telephone so you do not have to visit your cardiologist each time a recording is made.

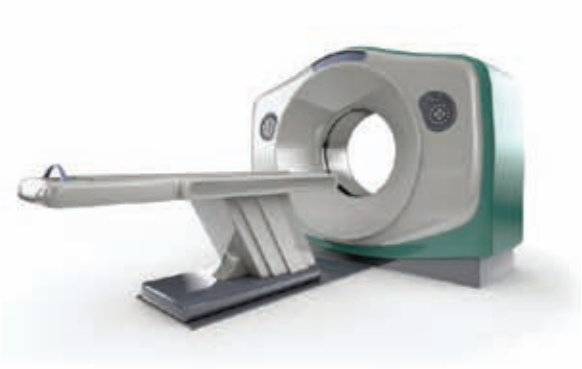
Implantable loop recorders are used in patients who are having concerning symptoms of arrhythmias without clear evidence of arrhythmia being found on other tests. The recorder is a small device about the size of an adult's little finger. It is surgically implanted under the skin through a one-inch cut on the chest. The recorder monitors the heart rate continuously and stores information according to the programmed heart rate levels or when activated by the patient while he is experiencing symptoms. The physician is able to analyze the recording to determine what the heart was doing at the time the symptoms occurred. Although they are more invasive than other monitors, patients report very few complications with implantable loop recorders. Sometimes the skin over the device becomes a little tender. It is, however, safe for the patient to get wet since the device is protected by the skin. The implantable recorder is removed once the cause of the symptoms has been discovered, but they may remain in the chest for more than 12 months.

Echocardiogram:

An echocardiogram is an ultrasound test that determines the size and shape of the heart. This is a non-invasive and painless test in which a wand covered in gel is passed over the chest to obtain images. In ARVC the heart may appear larger than normal on an echocardiogram. This finding is not enough to make a diagnosis of ARVC, but it is a piece of the diagnostic puzzle. Newer techniques can use echocardiography to measure the speed of movement of the wall of the right ventricle.

Cardiac MRI:

Recent studies have shown that a cardiac MRI (magnetic resonance imaging) is a good way to visualize the changes in the heart that can occur with ARVC. Like echocardiography, an MRI is a non-invasive test that does not involve radiation. It is more precise than echocardiography and it requires more time to assess the heart. During an MRI the doctor can see the size, shape, and tissue composition of the heart, as well as how well the heart is moving as it contracts and how the valves in the heart are working. To have an MRI, you must lie still in a chamber for about one hour. Being in an MRI machine is not painful, but some children may find it upsetting since it is a small, enclosed space and it makes a loud noise.The child must be very still during the examination, so sedatives or a general anaesthetic are sometimes used in younger children before an MRI. It is important to reassure children of all ages before the test that they will not be hurt. Sometimes a special dye called gadolinium will be injected into your vein before the test so that parts of the heart show up more clearly on the image. Because the machine uses large magnets to capture images, people with metal in their bodies (including pacemakers and implantable defibrillators) are not allowed to have an MRI. Unfortunately a normal MRI does not completely exclude the possibility of ARVC, and occasionally ARVC is suspected on an MRI in patients who do not have the disease. Because an MRI does not use



radiation, it is a test that can be done many times to follow changes in the heart and for this reason it is used for screening of family members of confirmed ARVC patients. Since MRI machines are very expensive, they are not available in every hospital. You may have to travel to a larger city to have a cardiac MRI performed.

Electrophysiological Testing:

Electrophysiology testing is one of the invasive tests that is sometimes performed both to diagnose and treat ARVC. This is done in some ARVC patients to identify the part of the heart that is causing the irregular beat, to assess the risk of future episodes, and to determine if the patient is a candidate for an implantable cardioverter defibrillator or radio-frequency catheter ablation. This test takes place in the Catheter Laboratory of the hospital. With adults, it is usually done under local anaesthetic meaning that patients are awake but are not able to feel the procedure. General anaesthesia may be necessary for children. A cardiologist inserts long catheters (thin tubes) into a large vein called the femoral vein in the top of the leg. The catheters are then advanced slowly through the patient's veins until they reach the right side of the heart. The doctor can then record electrical signals (like mini-ECGs) from different parts of the heart at the same time and this helps to produce a "voltage map" of the heart. Once the heart is "mapped", the cardiologist may decide to proceed with a treatment called radio-frequency catheter ablation (please refer to page 16).

Angiography and Endomyocardial Biopsy:

Like electrophysiology testing, angiography is an invasive test used in the diagnosis of ARVC. The setup is very similar to electrophysiology testing and the two are often performed during the same procedure. Once again, a catheter is

inserted into a vein in the leg and gently passed up into the heart. A special dye that shows up on X-rays is injected into the heart and X-ray pictures are taken. These pictures allow the cardiologist performing the test to see the size and shape of the right ventricle very clearly. In ARVC the right ventricle of the heart will appear larger than usual and parts of the wall of the ventricle may bulge out. During angiography, your cardiologist may decide to remove a small piece of heart tissue from the middle wall of the heart. This sample of tissue (biopsy) can be examined under a microscope to determine if the characteristic features of ARVC are present. Even biopsies are not a perfect test, however, and may not yield a definitive diagnosis. Biopsies increase the diagnostic rate for ARVC by about 33%, but are associated with a 1-2% risk of complications which will be explained to you by your doctor.

Genetic Testing:

With recent modification to the scoring system, genetic testing is now a part of diagnostic criteria and can help in making a diagnosis. However, unlike in some other conditions, there is a lot of uncertainty about what some of the genetic test results mean.

Most cases of ARVC are inherited in what scientists call an "autosomal dominant" pattern. In genetics, the word "dominant" means that a child can inherit a condition from only one parent. Dominant traits are different from recessive traits, which only occur when the gene comes from both parents. Different members of a family may share the same genetic mutation, but one person can have severe ARVC and the other may have no symptoms at all. This is called "variable penetrance" and scientists are not exactly sure how or why it occurs. Possible explanations include the presence of other "modifier genes", differences in lifestyle, or certain types of infections.

A number of ARVC mutations are not inherited from parents but appear spontaneously in an individual, but they can still be passed on to the next generation. Therefore, gene mutations can be identified in some, but not all, cases of ARVC. Genetic testing in individual patients can be used to

"Our family had access to genetic testing through a research study. Even though the news wasn't good – two of my three children had ARVC – it was still better to know we were giving them medication and imposing restrictions on them for a good reason."

– Zoe

rule in the possibility of developing ARVC if the mutation is proven to cause ARVC. Unfortunately, many mutations seen in these genes are of unknown importance and they may not be the cause of the ARVC. As well, having a gene mutation does not mean someone will definitely get ARVC. Many times a negative test does not rule out ARVC.

Should other family members be tested for ARVC?

When a patient is clinically identified as having ARVC (meaning that the numerous tests described in the previous section indicate a diagnosis of ARVC), genetic testing is often recommended. If a causative mutation is detected in one of the patient's genes, the rest of the patient's family can have the same test done to determine if they have the same ARVC gene. While a negative result on the test may be a huge relief and may prevent years of medical tests, a positive result can be extremely upsetting to a person who thought they were healthy, and can cause a great deal of uncertainty about the future. As well, some positive results may be "false positives" because the significance of the results is not really known at this time and may not be causing the ARVC.

Genetics counsellors can help families cope with these difficult decisions and interpret the meaning of test results if you decide to have the testing done.

As well, it is recommended that close relatives (children and siblings) of people with known ARVC should be screened by non-invasive testing methods once a year during puberty and every two years after puberty. Many of the

"My immediate family has had genetic testing, but several members of our extended family will not agree to participate in testing. I know I have to respect their decision, but it is very frustrating, since they may be at risk."
– Parker

uncertainties about ARVC diagnosis and genetic testing need to be discussed with the cardiologists, geneticists and genetic counsellors who are looking after people with ARVC.

If you or your partner have ARVC and are considering having a baby, a discussion with a genetics or cardiac expert will help clarify all the important issues that you will face and that you need to consider.

What are the available treatments for ARVC?

Although ARVC is a chronic and progressive condition, there are many treatments that can slow the progression of symptoms and arrhythmias and improve quality of life for patients. There are many different treatments available, and which one is appropriate in a given situation depends a great deal on the particular patient. For example, patients who have had episodes of syncope in the past or who show signs of heart failure are treated more aggressively.

The standard first-line treatment for ARVC is a class of medication called beta-blockers. Some people continue to have symptoms even when they are taking the right dose of beta-blockers and the medicine may have to be changed or another medicine added on. In other circumstances, patients may have an implantable cardioverter defibrillator (ICD) installed in their chest. In those with an ICD and frequent tachycardia despite medications, radiofrequency catheter ablation may be considered.

"I was shuttled between eight doctors before someone figured out what was wrong with me – at first I was angry that it took them so long to make this diagnosis, but now I'm glad that I have the treatment I need to be healthy."
– Michael

Beta-Blockers:

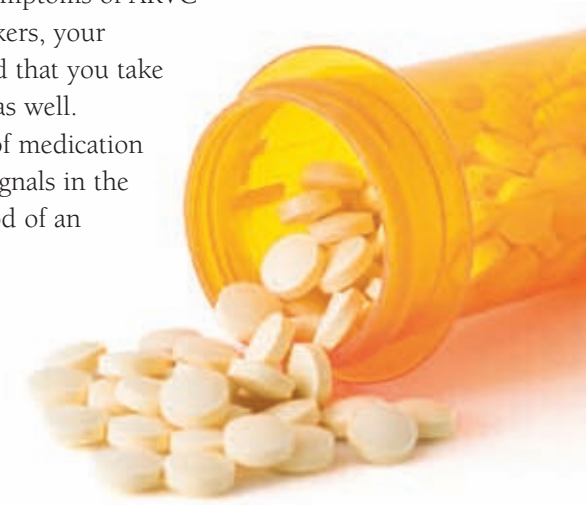
In ARVC, medication cannot cure the underlying disease, but may relieve symptoms like palpitations and reduce the chance of a fatal episode. Beta-blockers, which have names like atenolol, metoprolol, and nadolol, are frequently recommended. Adrenaline is the hormone that our body produces when we are excited, exercising, or frightened. By blocking this molecule, the drug makes the body less reactive to adrenaline. Some of the side effects of beta-blockers are slow heart rate, fatigue, dizziness, drowsiness, cold feet, sleep problems, and weight gain. Sometimes it takes a while for the body to adjust to beta-blockers, and your doctor may have to adjust your dose or change

medications to deal with side-effects. Also, as a child grows, his or her beta-blocker dose will usually have to be increased to adjust to his increase in weight and size.

Beta-blockers are also frequently prescribed for patients with ICDs, as they reduce the chance of both appropriate and inappropriate shocks.

Once you are taking beta-blockers it is very important that you take the prescribed pills every day, since stopping the medication suddenly can put you at an even higher risk for an arrhythmia. There are different types of beta-blockers, so other ones can be tried if the first one prescribed doesn't appear to work.

Anti-Arrhythmic Medications:



If you continue to have symptoms of ARVC even while taking beta-blockers, your cardiologist may recommend that you take another type of medication as well. Antiarrhythmics are a type of medication that changes the electrical signals in the heart to reduce the likelihood of an arrhythmia. Some types of antiarrhythmic medications that are used in ARVC are amiodarone (brand name Cordarone), sotalol ("Betapace"), flecainide ("Tambocor"), propafenone ("Rhythmol"), and mexilitene ("Mexilitil"). These medications can cause serious side effects, so it is important that you understand how to take them properly and you discuss all your concerns with your cardiologist before starting them. Your doctor may ask you to take precautions that seem strange to you; for example, patients using amiodarone should not drink grapefruit juice because it will interfere with the medication. Patients taking some of these drugs will have to have blood taken regularly to make sure the concentration of medication in the blood is not too high or too low. Your response to these medications may be monitored with Holter monitoring or stress testing.

Implantable Cardioverter Defibrillators (ICDs):

In some people with ARVC, the heart may suddenly start beating too rapidly to properly pump blood to the body. When this fast rhythm is present, a strong electric shock to the heart is the only way to "reset" it so it can start beating in a normal rhythm again. The process of shocking the heart back into a normal rhythm is called defibrillation, and many people are familiar with the electric paddles used for this purpose from medical TV shows like "ER". Implantable cardioverter defibrillators (ICDs) are devices that sit in the chest and work like tiny defibrillators inside the body. If it detects a heart rate above a programmed threshold, an ICD delivers a shock to the heart to fix the abnormal rhythm. ICDs are recommended in ARVC patients who still have an irregular heartbeat despite medical therapy, or who have a worrisome family history with relatives who died suddenly of ARVC. They are also used as first-line treatment in people who experienced a cardiac arrest and had to be resuscitated.

An ICD is made up of three main parts: a generator box which contains the computer and battery, the wire (or "lead") which connects the generator to the heart, and a programmer which is separate from the rest of the device and outside the body. The external programmer is what your doctor will use to adjust the settings of the device. Some patients need their ICD to work as a pacemaker, which means that the device tells the heart when to beat all the time by delivering low-energy pulses that are not felt by the patient. Other patients only need the defibrillation function of the ICD. In this case, the heart beats on its own with the device monitoring the heartbeat. The device will deliver a high-energy shock if it senses a dangerous type of beating. Patients usually remain in hospital for one day after their surgery to implant the ICD.

Patients with ICDs must be monitored every 6 months to check that the device is working properly and to determine if any dangerous heart rhythms have been detected. Sometimes your doctor can check the ICD over the telephone using a transtelephonic monitor or other remote interrogation device. When the doctor checks the ICD, he or she can tell how much energy is left in the battery and predict when a new battery will have to be installed.

"I am so glad I chose to get an ICD. I used to worry about my heart all the time and it seriously affected my life. I had some problems at first and had to have an operation to fix a lead, but ultimately I trust my ICD and I feel much more free."

– Maxwell

Usually ICD batteries last 3 to 6 years, but the timing depends on the number of shocks the device has given and whether constant heart pacing is required. ICDs are very useful in the management of ARVC, but they are associated with certain complications. These are uncommon but may include infections, inappropriate shocks (giving a shock when it was not needed), generator problems, and breaks in the wire. The implantation of an ICD can affect your emotional state as well. Many people have difficulty adjusting to the idea of an object inside their body. For these reasons, it is important to address all of your concerns, including emotional concerns, with your doctor.

There are also special precautions for patients with ICDs. Currently, these people should not have MRIs (X-rays are still safe), and they should try to avoid standing near the doorways of shops with theft-detection devices and airport security chambers (it is all right to pass through these, however). Patients are also advised not to hold cellphones directly over the device and not to stand close to a microwave oven when it is on. Patients with ICDs are also given special security cards when they travel, since they may require different inspection procedures than other passengers. Most people with ICDs remain on medication to help reduce the chances of requiring a shock.

Radio Frequency Catheter Ablation:

This treatment is usually recommended for ARVC patients who do not respond well to antiarrhythmic medications, or patients with an ICD that is shocking too often. The abnormal heart rhythms in ARVC occur when the heart muscle is slowly replaced by scar tissue and fat, which conducts electrical signals differently than normal heart muscle does. Destroying the tiny area of tissue that is responsible for conducting this dangerous electrical signal in the heart can prevent the heart from beating abnormally. Ablation is performed during the procedure of

electrophysiological testing, which is described on page 8. During this test, which is done under anaesthetic, the cardiologist will guide a tiny tube with an electrode on the end through a blood vessel in the groin all the way up to the heart, using images from a machine similar to an X-ray called a fluoroscope. He or she will induce a fast heartbeat in order to locate the area where the signal is transmitted abnormally. Once the area is found, radiofrequency energy (heating) is used to destroy the small part of the heart that is causing the problem. Sometimes freezing is used instead of heating and it is called "cryoablation". This procedure can take many hours and you will have to lie still for up to six hours after the operation to allow the incision to heal properly. Patients may feel stiff after lying still for so long, but usually there are no other complications. Although this treatment works well for many patients, often more than one session of ablation is required as the disease progresses and other areas of the heart become affected.

"Try not to think about what life would be like without this. With each bad day there are lots of good days, so cope the best way you can."

– Henry

Treatment for Heart Failure:

Heart failure is a term used to describe the situation when the heart becomes weak and is no longer able to pump blood effectively. There are many causes of heart failure, but in patients (rarely children) with ARVC, it is due to the replacement of muscle with fat and scar tissue. Symptoms of heart failure include leg swelling, cough, chest pain, trouble exercising, and shortness of breath. If you are experiencing these symptoms, your cardiologist may prescribe medications to help strengthen the heart or to reduce its workload. These medications include diuretics ("water pills"), beta-blockers, and blood pressure medication. Sometimes blood thinners are used if there is a risk of blood clots forming. In very rare cases of severe heart failure that does not respond to treatment, heart transplantation may be considered.

Treatment of Asymptomatic Patients and Family Members:

Asymptomatic patients and family members do not require specific antiarrhythmic or other cardiac treatment. However, they should be followed by regular non-invasive cardiac investigations for early recognition of disease. According to individual conditions, your cardiologist will decide if you require any prophylactic medication and how often you need to be seen.

What lifestyle changes can I make to help my condition?

In addition to your medication, surgery, and/or ICD, there are some lifestyles changes you can make to reduce your chance of having an episode of arrhythmia. The same recommendations are not necessarily appropriate for every patient. Ask your doctor which lifestyle modifications he or she recommends for you.

Participation in Sports:

Because dangerous arrhythmias are more likely to occur when the heart is beating rapidly, strenuous and competitive sports are not recommended for people with ARVC. Ask your cardiologist which level of activity is appropriate for the severity of your particular condition. Sometimes lower energy sports such as golf, bowling, and curling are good ways for people with ARVC to stay in shape and have fun without increasing their risk. It is also very important to discuss your child's physical limitations with his or her school so that accommodations can be made in gym class and recess. Making emergency plans for the school to follow if the child faints or receives a shock from his or her ICD is crucial in making school as safe as possible for your child.

"I've been a "jock" my whole life. When I was diagnosed with ARVC, I had to stop playing competitive hockey and I thought my life was over. After the initial shock, I learned to play golf and I'm becoming quite good at that. I'm also coaching my kids' hockey team, which allows me to still be involved in the game."

– Stephen

Medications to Avoid:

Some medications cause the heart to beat more quickly as well, and these should be avoided. These medications include stimulants like pseudoephedrine, which is in non-drowsy allergy medicines and can be purchased without a prescription. Always ask your doctor before taking any medication. Being ill with a fever, vomiting, or diarrhea can cause the balance of electrolytes (little

charged particles in the body that help the heart to beat normally) to be disrupted. Drinking a sports drink like Gatorade when you are ill can help replace these electrolytes. Nicotine, caffeine, and alcohol can also trigger an arrhythmia, so these substances should be avoided or used in moderation. In addition to medications, many illegal drugs are dangerous for the heart as well. You will be at particular risk if you choose to use these substances.

Complementary Therapies:

Some patients find complementary therapies, such as reflexology, massage, homeopathy, and chiropractic treatments, helpful. Before trying any of these therapies it is important that you consult with your doctor to make sure the therapy is safe for ARVC patients and it does not interfere with your medication. Remember that naturopathic remedies are also drugs, and they can react with beta-blockers and other medicines.

MedicAlert Bracelets:

MedicAlert is a company that maintains a database of people with medical problems so emergency personnel can access their medical information quickly. Members of MedicAlert wear a bracelet, dog tag, or wristband inscribed with a file number. If a person with ARVC is wearing the bracelet and has as an episode of syncope, a paramedic arriving on the scene can contact MedicAlert and reference the file number on the bracelet to find more specific information about the person's medical condition and information about who to contact in an emergency. MedicAlert bracelets are an excellent way to assure that you will get the most appropriate treatment if you have an episode. For more information about MedicAlert, go to their website www.medicalert.ca, or telephone them at 1-800-668-1507. MedicAlert charges for its services, however there is a Member Assistance Program that provides these identifying bracelets to people who may need financial assistance. Although it is possible to purchase medical bracelets from your local drug store, we strongly recommend that you buy your bracelet from MedicAlert because the other bracelet companies do not maintain a database of patient information.

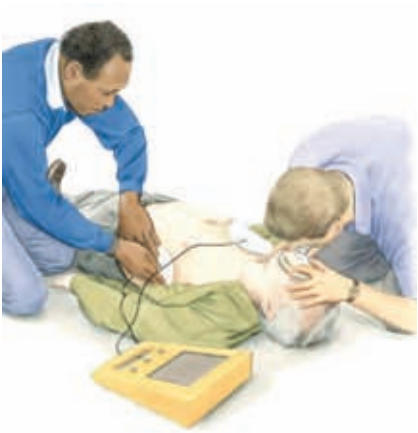
CPR Lessons:

Many family members of people with ARVC take CPR (cardiopulmonary resuscitation) courses so they are able to respond if their loved one should have an episode at home. In Canada, CPR courses are run in many locations by teaching agencies such as St. John Ambulance, Lifesaving Society, and Red Cross. There are also many private companies that provide CPR certification. Many training companies will arrange for a CPR instructor to come into your home to teach your family or a small group. For more information or to find a location in your area, use any internet search engine or the yellow pages.

Automatic External Defibrillators:

Automatic external defibrillators (AEDs) are devices that allow a non-medical person to deliver lifesaving electric shocks to a person whose heart has stopped beating. AEDs are similar to the electric paddles used in hospitals and ambulances, but many also have an automated voice telling the user what to do so they can be operated by almost anyone in the case of an emergency. If you or your child have been diagnosed with ARVC, it is reasonable to discuss having an AED available for your place of employment or for the school your child attends. Please contact The Canadian SADS Foundation for assistance in this matter.

Whether families with ARVC should purchase AEDs for the home is somewhat controversial at the moment. While many families feel that owning an AED gives them peace of mind, it also can exaggerate the perception of risk and affect the patient's quality of life. In order to own an AED, you must obtain a prescription and be trained in the use of the device. As with everything, it is best to ask your doctor if he or she thinks this is a good idea for your family.



What are some parenting challenges when raising a child with ARVC?

Raising a child is certainly one of the most difficult tasks there is, but raising a child with ARVC presents a whole new set of challenges. Children are often uncomfortable being treated differently than their peers, and this stress may manifest itself as frequent stomachaches and headaches. Diagnosis of a chronic condition in a child places stress on all of the family, and siblings may feel neglected as a result. The important thing to remember is that everybody has trouble adjusting to a diagnosis of ARVC initially, but most learn how to cope with it. There is always help available as well. Ask your child's cardiologist or genetic counsellor (if you have consulted one) about people who can help your family cope. Many parents find that learning more about the condition helps them to feel more in control. Reading this booklet is a good first step, but using some of the resources listed below can help broaden your understanding. Some people find it helpful to speak with other parents, patients, or siblings, who have had similar experiences; www.sads.ca is a great place to begin finding people to share stories and coping mechanisms. Different parents choose different ways of dealing with ARVC. What remains important is choosing the balance that is right for your family.

"My biggest challenge has been to avoid letting my son know how concerned I am about his health. It's so tough to find the right balance between informing your child about how to be safe and scaring him unnecessarily."

– Bradley

Where can I find more information about ARVC?

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.chrsonline.ca (A resource to help you find a heart rhythm doctor in your area)

www.cagc-accg.ca (A resource to help you find a genetics counsellor in your area)

www.cardiomyopathy.org (Do a search on this site for ARVC; a very helpful British website with information about the cardiac and psychological effects of ARVC and a glossary of terms.)

www.hrspatients.org (An American site about different cardiac arrhythmias with lots of information about ICDs and procedures)

www.arvd.org (If you are interested in participating in ARVC research)

www.arvd.com (The website from Johns Hopkins Medical School; this may be a little too advanced if you do not have a science background)

www.medlineplus.gov (A good general reference for information on health problems and medications)

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 an 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 more information, please contact
The Canadian SADS Foundation at
info@sads.ca.

Supporting Families.
Saving Lives.

For more information:
The Canadian Sudden Arrhythmia Death Syndromes (SADS) Foundation

www.sads.ca

info@sads.ca

This booklet was originally written in 2007 for The Canadian SADS Foundation by Dr. Kate Houston who, at that time, was a medical student at the University of Toronto.

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Quotes are a summary of patient perspectives.

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