

THE HEART INSTITUTE

Fontan Circulation Patient Handbook



Table of Contents

Introduction

The Importance of Heart Care as an Adult	
When You Had a Fontan Operation as a Child	1
Your Fontan Care Team	1
Important Terms	1
In Case of Emergency	1

Chapter 1 | The Heart

Understanding the Heart	5
The Single Ventricle Heart	5

Chapter 2 | The Fontan Operation

History of the Fontan	9
Types of Fontan Operations	9

Chapter 3 | Living with a Fontan Circulation

Related Conditions	
<i>The Liver</i>	13
<i>The Kidneys</i>	13
<i>The Lungs</i>	13
<i>The Digestive System, Including Small & Large Bowels</i>	14
<i>Coagulation, Blood Clots & Stroke</i>	14
<i>The Heart</i>	15
<i>Sleep Apnea</i>	16

Additional Surgeries & Transplantation	16
Sexual Activity, Pregnancy & Contraception	
<i>Sexual Activity</i>	16
<i>Pregnancy</i>	17
<i>Birth Control</i>	17
Genetics & Heredity	18
Life Expectancy	18
Living Will & Healthcare Proxy	18

Chapter 4 | Being Heart Healthy

Physical Health	
<i>Healthy Weight</i>	21
<i>Diet</i>	21
<i>Sleep</i>	22
<i>Exercise</i>	22
Mental Health & Coping	23
Managing Stress	23
Alcohol and Drugs	24
Medications	24
Work	25
Travel	25

Appendix

Glossary	29
Support and Resources	29



Introduction

Introduction

THE IMPORTANCE OF HEART CARE AS AN ADULT WHEN YOU HAD A FONTAN OPERATION AS A CHILD

As someone who was born with a single ventricle heart, you have special health concerns now as an adult. This book was written for our patients who had Fontan heart surgery and are now getting care from us as adults.

Throughout your childhood, you may not have known what heart procedure you had or how it would impact you throughout your life.

Now that you're an adult, it's important that you take ownership of your health and healthcare. We hope this book helps to answer questions you may have about your condition, how it will affect your health moving forward, and what you can do to help prevent problems. Since technology has advanced, more patients are living to adulthood, but resources on this topic are limited. We hope this serves as a helpful guide for you and your heart health as you continue living with a Fontan.

YOUR FONTAN CARE TEAM

A heart affected by congenital heart disease (CHD) is unique. For that reason, the gold standard of care today for adults with CHD is to have routine check-ups and treatment by experts trained in adult congenital heart disease (ACHD). In our Adult Congenital Heart Disease Program at Cincinnati Children's, we offer that gold standard of care.

The Fontan Management Clinic at Cincinnati Children's is part of our ACHD Program. It is one of the few programs in the country to provide comprehensive and specialized care to people who have had a Fontan procedure.

Our program includes a team of internationally known faculty. They are experts in recognizing and treating issues linked to CHD. We treat teens and adults with all forms of CHD. We also treat those with problems linked to prior surgery for CHD.



IMPORTANT TERMS

You have likely been seeing a cardiologist for many years and may know these terms. If not, take a few minutes to get to know them, as they will be used throughout this book.

ACHD: adult congenital heart disease

CHD: congenital heart disease

Cardiologist: heart specialist

Exercise physiologist: a healthcare professional who is able to do exercise tests to check heart and lung fitness. They can also work with patients on improving their health or maintaining good health through fitness

Fontan: operation performed to treat patients with a single ventricle heart

Fontan circulation: the reconfigured blood flow that occurs as the result of having a Fontan operation

Hepatologist: liver specialist

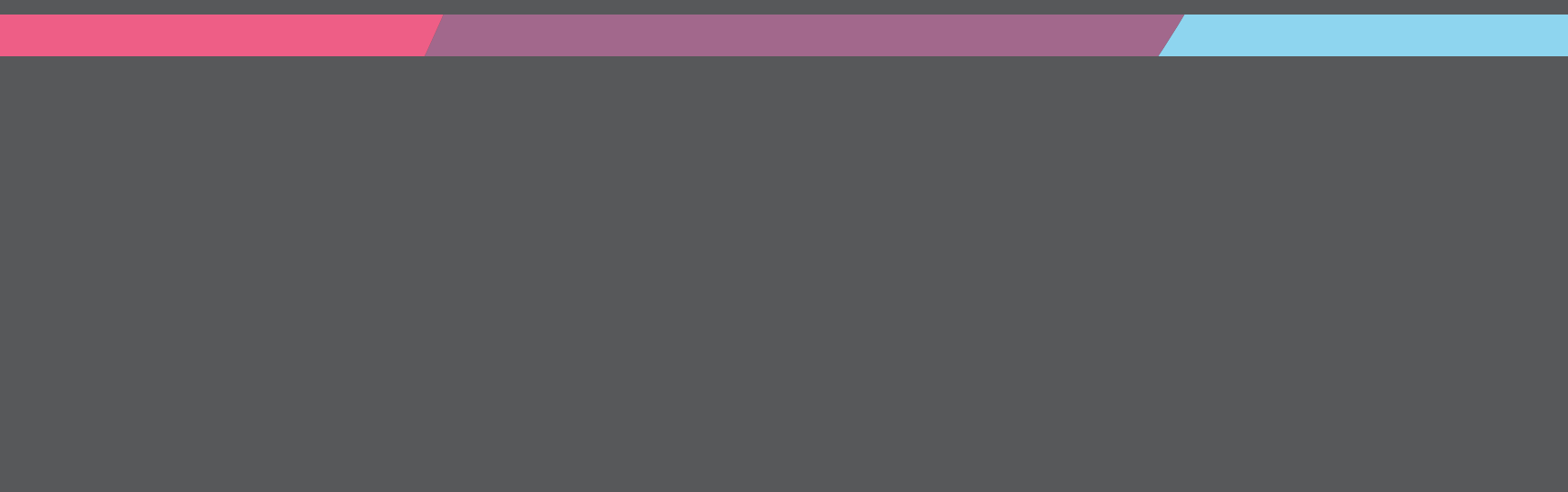
In Case of Emergency



Talk with your cardiologist about what is best for you to do in an emergency. You should also know which hospital to go to for an emergency. It is best to talk about this before an emergency occurs. Discuss this when you are well. You will want to have time to make plans in advance.

When you have an emergency with either your heart or other body organs, you should go to your closest Emergency Department. Bring a copy of the paperwork from your most recent cardiology visit. Also bring the phone number of your cardiologist so the Emergency Department doctors can reach them. Your cardiologist will be able to help manage any medical care linked to your Fontan.

In the case of a more direct emergency, call 911.



CHAPTER 1

The Heart

The Heart

UNDERSTANDING THE HEART

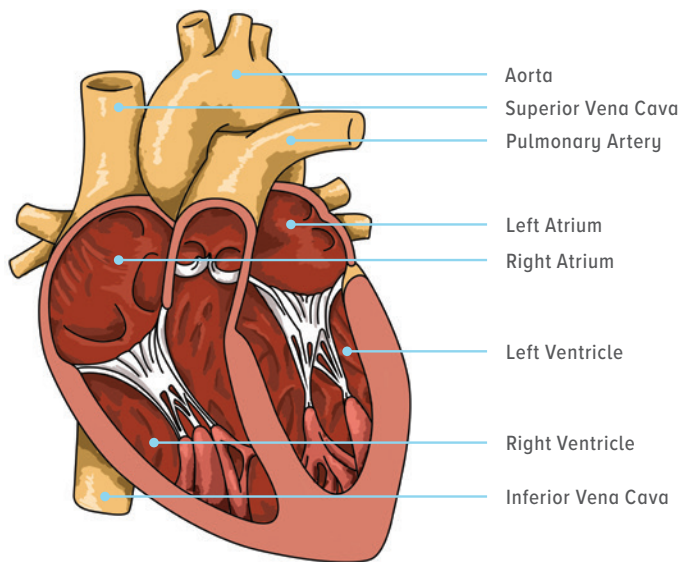
In order to understand what a Fontan is, we have to understand how the normal heart works, and also how a heart with only one ventricle works.

What is a normal heart and what does it look like?

If you do not know the anatomy of the heart, take some time to study the figure here. Many of these terms will be used throughout this book.

A normal heart is made up of four chambers: two atria and two ventricles. The left and right atria sit above the left and right ventricles.

Normal Heart



The ventricles serve as the main pump for the heart. The first part of the pump (the right ventricle) squeezes blood into the lungs. The second part of the pump (the left ventricle) squeezes blood into the body. These two muscle pumps can only work if they are filled the right way with blood all the time. This is done by the other two chambers (the atria), which feed the pumps with blood in a timed and orderly way. The atria receive the blood and then pump the blood into the bottom chambers (ventricles).

How the Heart Works

- The right-sided top chamber (right atrium) receives the used (oxygen-poor) blood from the superior vena cava (SVC) and inferior vena cava (IVC). The right atrium then feeds the blood into the right-sided pumping chamber (right ventricle).

- The right ventricle then pumps this used blood to the lungs by using the pulmonary artery.
- On the left side, the top chamber (left atrium) receives new (oxygen-rich) blood from the lung (pulmonary) veins. Then it feeds this blood into the left-sided pumping chamber (left ventricle).
- The left ventricle then pumps this new blood to the body by using the aorta.

THE SINGLE VENTRICLE HEART

What is a single ventricle heart?

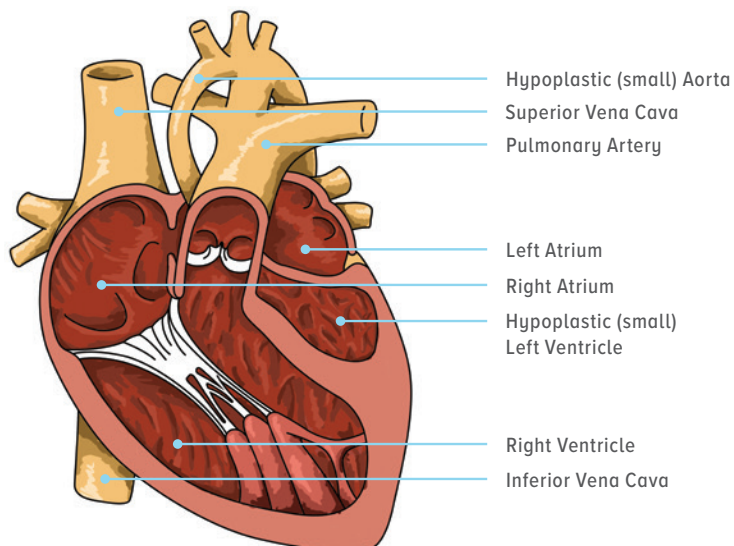
Some people are born with a heart that has only one ventricle (or pump). It has to work harder to do the job of both pumps. This is known as a univentricular, meaning “one ventricle,” heart. You may also hear it called a “single ventricle heart.”

When there is only one ventricle pumping blood, the blood that is oxygen-rich mixes with the blood that is oxygen-poor. This causes the person to have cyanosis—a bluish look of the skin, nails, and tongue. The pump also has to enlarge as it tries to pump blood both into the body and into the lungs, which can overwork the ventricle.

The Fontan operation was developed to avoid overworking the single ventricle, to address the complications faced by people with single ventricle hearts, and to give them the chance to live longer, and more healthy lives.

Single Ventricle Heart

Example Before Surgery (Hypoplastic Left Heart Syndrome)





CHAPTER 2

The Fontan Operation

The Fontan Operation

HISTORY OF THE FONTAN

What exactly is a Fontan?

The Fontan operation is named after Dr. Francis Fontan, who performed the first Fontan procedure in 1971. The operation is designed to help a single ventricle heart work better. It also takes away cyanosis (the bluish look of the skin, nails, and tongue). The operation is often referred to today as simply “a Fontan.”

TYPES OF FONTAN OPERATIONS

The Fontan operation has changed dramatically over the past 40-50 years. With all Fontans, the goal is to change the circulation to make the single ventricle function the best it can without overworking it. This means the single ventricle will end up doing the harder work of the heart, pumping blood to the body. The job of getting blood to the lungs must be done without a pump.

The “Fontan circulation” refers to this configuration where the single ventricle pumps blood returning from the lungs to the body, and the blood returning from the body travels to the lungs by direct blood vessel connections without a pumping chamber.

The Classical Fontan

Dr. Fontan and Dr. Guillermo Kreuzer performed the first Fontan operation at similar times, each in two different countries. They built the procedure because, until then, patients with a single ventricle heart did not have a good option to treat their condition. They often did not survive beyond childhood.

This form of the Fontan became known as the classical Fontan or atriopulmonary Fontan. In this procedure, the surgeon makes a new pathway for used (oxygen-poor) blood to pass from the large veins directly to the lung’s main artery (pulmonary artery) without going through the heart itself.

Since the first Fontan, the procedure has changed. Today, surgeons do these two types of Fontan operations.

The Lateral Tunnel Fontan

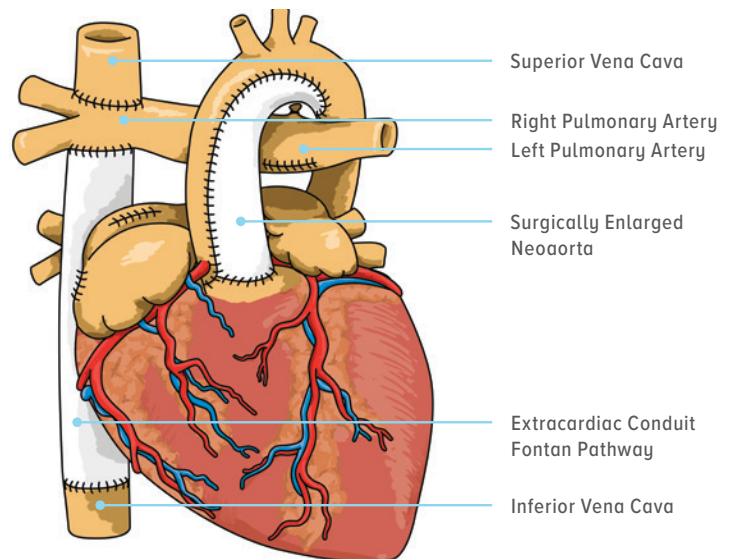
In the lateral tunnel Fontan, the surgeon makes a “tunnel” inside the heart that carries oxygen-poor blood from the inferior vena cava to the pulmonary arteries.

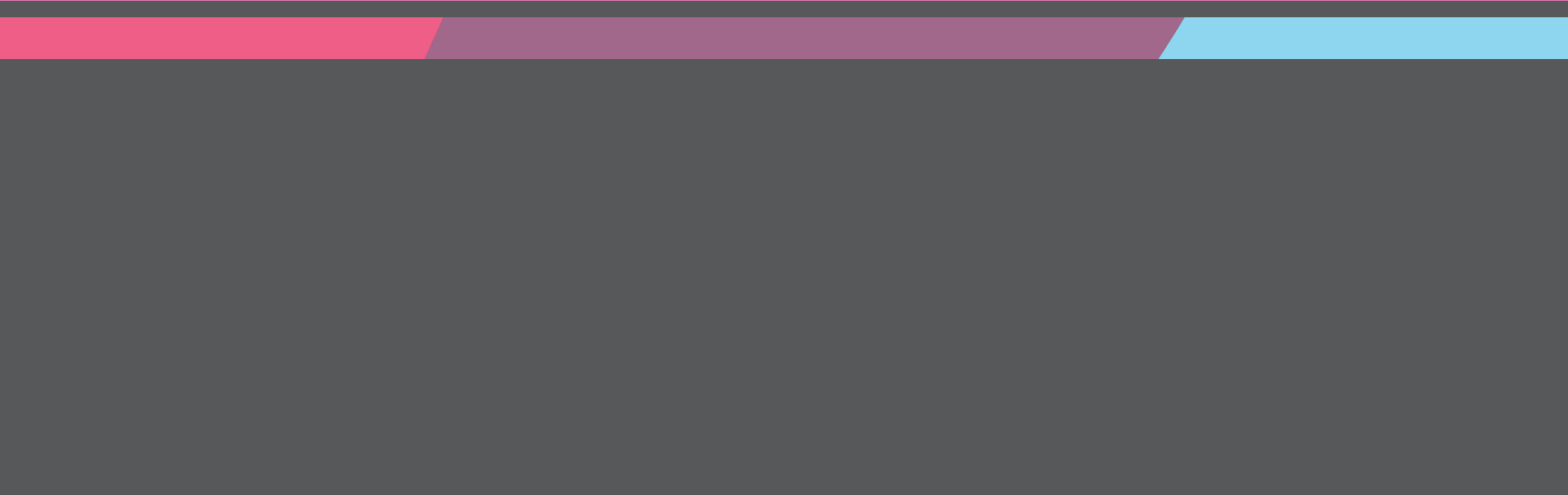
The Extracardiac Fontan

In the extracardiac Fontan, the surgeon stitches a tube on the outside of the heart. This tube is stitched so that it takes oxygen-poor blood from the inferior vena cava and carries it separately from the heart right into the pulmonary arteries.

Sometimes when a Fontan operation is done, the surgeon makes a small hole in the Fontan lateral tunnel or extracardiac tube, so that the tube connects to the rest of the heart. This is called a fenestration.

Extracardiac Fontan





CHAPTER 3

Living with a Fontan Circulation



Living With a Fontan Circulation

RELATED CONDITIONS

How will the Fontan affect other parts of my body?

Over the past decade, we found out that as result of its unique circulation, the Fontan operation affects not only the heart, but also other parts of the body. More than half of Fontan patients will have some degree of organ damage or strain within 10 years of having the Fontan. It is important to be aware of this and the side effects that could occur.

This section reviews how other organs can be affected by a Fontan operation, and conditions that may happen in people with a Fontan circulation. It also reviews your risks of getting certain related health issues as you age.

THE LIVER

The most common organ affected by the Fontan operation is the liver. By the time patients become adults, most will show signs that their liver has been affected. These early changes are not harmful, but over time, they may change into different degrees of liver damage.

Fortunately, severe liver damage is less common in patients with a Fontan. It is important, though, for your doctor to do routine checks for signs of liver disease and also for signs of changes. In patients with severe liver disease, it is important to have routine check-ups with a congenital heart disease expert and also a liver expert.

Protecting the Liver

How can I best protect my liver when I have a Fontan?

When the Fontan circulation affects the liver, it is often very mild and will not change your day-to-day life. The Fontan puts higher back pressure on your liver, and it may become damaged. This happens more as you age. This damage shows up as scars in your liver known as fibrosis, or when severe, cirrhosis.

In most cases, though, cirrhosis does not have any distinct symptoms. You may feel some fullness or discomfort in the right upper stomach. You may feel sick and tired. Sometimes, you may have belly bloating or jaundice (yellow skin). It is important to talk about your liver issues with your cardiologist and hepatologist (liver specialist).

Rarely, liver cancer may happen in patients with Fontan liver disease. The best plans for watching for this are still being worked out. At this time, we recommend yearly screening.

Here is how you can best protect your liver when you have a Fontan:

- Follow the directions/advice given by your cardiologists. When your heart works well, it causes much less damage to your liver.
- Make sure you are up to date with your vaccinations (shots). This includes those to protect you against hepatitis A and hepatitis B.
- Aim to keep a good body weight. Eat a balanced diet and exercise regularly. When you are very overweight, the liver stores a large amount of fat. This bothers the liver and damages it. (See chapter 4.)
- Avoid too much added sugar in your diet. Be mindful of drinks that are sweetened with sugar. (See chapter 4.)
- Avoid alcohol. It is very important not to take in large amounts of alcohol. (See chapter 4.)
- Be sure to ask about your liver when you see your cardiologist.
- Avoid medications that damage your liver. This includes high doses of Tylenol [more than 4 grams per day].
- If you get jaundice (yellow discoloration of your eyes and skin), wide-spread itching, or easy bruising, please call your hepatologist or doctor. These may be signs of liver disease.

THE KIDNEYS

As many as 15 percent of people with a Fontan will have a mild degree of kidney damage as a result of their Fontan. Most often this mild form of kidney damage does not need treatment, apart from making sure that the damage does not become worse over time.

THE LUNGS

Restrictive Lung Disease

In patients with a Fontan, the muscles that sit around the chest cage are often weakened. The rib cage itself may not be able to move as much as normally because of previous operations. Sometimes the diaphragm may also be weakened. The lungs may be smaller than in people with a normal heart. This is called restrictive lung disease. This may affect how much exercise you can do. It is important for you and your doctor to carefully monitor your lungs.

In rare cases, a Fontan patient may get a serious lung condition called plastic bronchitis, which requires prompt treatment.

Plastic Bronchitis

Plastic bronchitis is a serious condition where protein leaks into airways. Symptoms include coughing, wheezing, and shortness of breath. These proteins can sometimes clog the airways. When the protein is coughed out, it can take the shape of the airway. This gives it a streaky branching look.

Patients who get plastic bronchitis will need repeated blood tests, imaging scans, and very likely a cardiac catheterization procedure. This will help to find out how severe it is and in order to start treatment. Most often treatment will involve using inhalers and other medicines.

Patients who get plastic bronchitis, must be treated by someone who has training and skill in CHD and in Fontan operations.

THE DIGESTIVE SYSTEM, INCLUDING SMALL & LARGE BOWELS

Your digestive system may be affected with a Fontan. This does not happen very often. Roughly up to 1 in 10 may be affected in this way.

The Fontan circulation puts pressure on the bowels, causing irritation of the bowel lining. In some people it will cause protein to leak into the bowel. This is called protein losing enteropathy, or PLE.

Protein Losing Enteropathy (PLE)

PLE is a serious condition that affects a small number of people with a Fontan. People who get PLE lose proteins in their stool (poop) because of a leak of protein from the blood vessels into the bowels. When the body cannot keep up with this leak of protein, the body's protein levels become very low and the patient starts having problems.

PLE causes fluid/water to build up all over the body.

Symptoms include:

- Swelling in the legs, belly, face, back and lungs
- Loose stools
- Cramping and belly pain

It is not always clear why people get this condition. In some, a cause can be found and then the condition may be reversed. Therefore, it is very important to be followed by a CHD cardiology expert who understands the Fontan condition. They are better able to assess and treat you as soon as you have any of these symptoms.

The condition needs early treatment. Your congenital heart cardiologist will work hard at finding any likely causes that are treatable. This often means having blood, stool, and ultrasound tests and sometimes a cardiac catheterization test on your heart. Treatment includes treating any known cause, and then giving water pills (diuretics), proteins, immunoglobulins, steroid medicines, and other medicines that may help, such as sildenafil. There are also newer surgical procedures that help ease the pressure on the bowels. These procedures have been successful in treating some patients with PLE. There are also procedures that block of the leaking points in the bowel and these procedures look very promising. However there is too little experience with the procedure to know what happens months to years after the procedure.

If PLE cannot be controlled/cured, the only other choice may be a heart transplant.

COAGULATION, BLOOD CLOTS & STROKE

Am I more likely to get blood clots because I have a Fontan?

Am I more likely to suffer a stroke as compared to other people?

Coagulation refers to the clotting of blood. Having a Fontan will put you at higher risk for getting blood clots. These can include:

- Clots near or inside the heart
- Clots in the large veins of the legs or arms (deep venous thrombosis)
- Clots that travel to lungs (pulmonary embolism)
- Strokes

Blood clots can raise the risk of having a stroke. Studies have shown that the chances of having a stroke in Fontan patients are often quite small; but, the risk is still higher than if you did not have a Fontan.

When you have a Fontan, your blood may be more sticky than normal. This can make it easier to get a blood clot. This may cause a stroke or mini-stroke. Other things that can cause a stroke are:

- History of having an arrhythmia
- An older-style Fontan operation
- A prior blood clot/clots
- Having had stents put inside your heart
- Having any blind ending pouches inside your heart

While patients with a Fontan have a higher risk of getting blood clots than other people, medication will be prescribed to help with this. This medication will lower your risk of getting blood clots. Your doctor's will assess your overall risk of getting blood clots to figure out which type of medicine is best for you.

Medications (see also chapter 4)

You may be prescribed daily aspirin and/or a blood thinner such as warfarin (Coumadin®) to help prevent clotting. These medicines lengthen the time it takes for your blood to clot. They also lower the chances of a dangerous blood clot forming.

Anticoagulants are medicines that prevent blood from clotting. Warfarin is the most often used anticoagulation medicine. When on warfarin, patients need to have their blood drawn regularly to look at how anticoagulated the blood is (to make sure the blood doesn't clot too much or too little). There are also many new drugs that can do this, but do not need regular blood draws to track their effect. These drugs are known as novel anticoagulants, "NOACs," or "DOACs." The safety data for these medications in patients with a Fontan is slowly coming out. The decision to use these medicines needs to be made with caution.

Whichever medicine your doctor orders, it is key that you take the medicine each day as prescribed. Be careful to not miss doses. Missing doses can increase your chances of having a dangerous blood clot. This includes blood clots that could result in death.

Because anticoagulants lengthen the time it takes blood to clot, they can slightly increase your chances of bleeding. However, serious bleeding events are rare when patients take anticoagulants as ordered. Your doctor will talk to you about any safety measures, such as avoiding high-risk activities like contact sports.

It is very important to follow safety measures related to blood clots when you have a Fontan. Ask your cardiologist about your specific risks, as they differ for each patient with a Fontan.

Bleeding Risk

In addition to having a risk of forming blood clots, some patients with a Fontan also have a risk of bleeding. Bleeding may occur into the airways, and you might get blood in your spit. If this happens call your cardiologist. If there are more than a few drops, make sure that you are seen at the closest emergency department. You may also have bleeding in your bowels. This may show up as black tarry stools, or blood mixed into your stools. Your cardiologist will carefully weigh the risk of bleeding vs. blood clotting in making the decision whether to put you on aspirin or warfarin or a NOAC.

THE HEART

Am I more likely to suffer a heart attack with a Fontan?

Your chances of having a heart attack with a Fontan are low. Risk factors for heart attacks are the same as those for all people, and include:

- High blood pressure
- High cholesterol
- Smoking
- Family history of coronary artery disease
- Sedentary (not active) lifestyle and lack of exercise
- Older age
- Male gender
- Obesity and extra weight around your belly

Arrhythmias

Arrhythmias are when your heart rate is not normal. It could be fast or slow or otherwise not normal. These happen often in people with a Fontan operation. If you had an older style Fontan operation, it is very likely to happen during adult life. If you have one of the newer Fontan operations, such as the lateral tunnel, it is less likely to happen but can still happen in as many as 1 in 10 people with a Fontan.

It is important to notice when you have an arrhythmia. This will feel like fast heart beats, as if you have been running, only they will happen when you are sitting quietly. Sometimes people feel dizzy or have chest pain when they get arrhythmias. It is important to let your cardiologist know when you have any of these symptoms.

If your arrhythmias last more than a few minutes, make sure that you go to the closest emergency department. Make sure that you take your last clinical letter with you and let the emergency department doctors know where you get your cardiology care.

SLEEP APNEA

What should I know about sleep apnea?

Sleep apnea is a spontaneous stopping of breathing during sleep. This usually occurs for short periods of time. This condition often occurs in people with heart problems, people who are overweight, or people that have problems with their airways. About 1 in every 10 adults and about 1 in every 25 children have sleep apnea. It is also called obstructive sleep apnea, or OSA.

It is important to find out whether you have sleep apnea if you have a Fontan because of what sleep apnea does to the heart and lungs. When you have sleep apnea, your oxygen levels go down and carbon dioxide levels rise. These two events make it much harder for blood to flow through the lungs, even more so when you have a Fontan.

Sleep apnea is more common in men, women who have been through menopause, and those that are overweight. We do not know how common obstructive sleep apnea is for patients who have a Fontan. Our sense is that it affects many Fontan patients without them knowing about it. If not treated, sleep apnea can worsen heart and lung function. It has been linked to high blood pressure and high blood pressure in the lungs, stroke, heart attack and earlier death.

Symptoms of sleep apnea are:

- Snoring at night
- Feeling tired when you wake up in the morning
- Falling asleep a lot during the day because you feel tired
- Waking up with a headache

Be sure to talk with your doctor about sleep apnea if you have any of these symptoms, even more so if you have more than one.

ADDITIONAL SURGERIES & TRANSPLANTATION

Am I likely to have surgery again after my Fontan? Will I one day need a heart transplant?

Some people who have had a Fontan will need surgery again as they get older. The surgery may be needed to remove blockages of the veins or to help your heart work best. Even after the best medicine and surgery, some patients will still have symptoms due to ongoing abnormal vein or heart function. In this case, your cardiology team may talk to you about the chance of needing a heart transplant.

The timing to discuss heart transplantation is different for every person. The team most often begins to discuss a heart transplant when your heart problem cannot be treated well by surgery, but before your other organs show major damage. If you are referred to see if you need a heart transplant, you will get a full test of your heart and other organs by the heart transplant team. At this test, you will get information about the risks and potential benefits of a heart transplant and be asked to talk about your questions about the procedure. After the test, you, your cardiologist, and the heart transplant team will decide if listing for heart transplant is the right thing for you.

SEXUAL ACTIVITY, PREGNANCY & CONTRACEPTION

SEXUAL ACTIVITY

Can I have sex like normal when I have a Fontan circulation?

Many people feel worried about having sex when they have congenital heart disease, but sex is safe for those with CHD and those with a Fontan. The chance that you may have a heart attack during sex is most unlikely.

You should talk with your cardiologist who can outline if there are any special concerns for you during sex. Some people who are very short of breath may choose positions that prevent strain or a lot of physical effort for more comfort. Sometimes the medicines you take may change your sex drive, or may cause problems with erections. Medicines that tend to lower your blood pressure are more likely to cause problems with erections.



PREGNANCY

If I have a Fontan, can I become pregnant?

Most women with a Fontan can become pregnant and have a healthy pregnancy, with a healthy mom and baby, but there is a higher risk in women with a Fontan. For some women, the risk may be so high that it is not safe to have a baby. If this is the case for you, thinking about adoption or surrogacy may be helpful. It is very important that each pregnancy is planned.

ACHD guidelines suggest that women who are thinking about getting pregnant have a full evaluation to figure out what the actual risk might be for both the mother and her fetus. The concern with getting pregnant after having a Fontan operation is that the heart muscle has to work harder and faster. Some of the other normal changes that happen in pregnancy are:

- The amount of blood volume going through the body is doubled, which makes the heart pump harder. In pregnancy the blood is also stickier, which can raise the risk for getting clots. (See also chapter 3.)
- The greater blood volume may make the heart more irritable and likely to have arrhythmias.

Women with a Fontan have a greater chance of getting these types of problems during pregnancy:

- Palpitations (changes in the way your heart beats)
- Swelling in the legs and body due to build-up of body water
- Blood clots
- Shortness of breath

Also, some of the heart medicines you take may affect you and the baby while you are pregnant. Your doctor and pharmacist will need to look at all of your medicines with care before you become pregnant to avoid causing problems for the fetus.

If you are thinking about getting pregnant, it is important to talk to your doctor to see if your heart is healthy enough for pregnancy. Your doctor may need to do certain tests to help figure this out.

Once you become pregnant, it is important for you to see your cardiologist and a high-risk obstetrician throughout the pregnancy.

Some other important facts to know about being pregnant with a Fontan include:

- Women with a Fontan carry a higher risk for miscarriage.
- They are also at increased risk of going into early labor.
- Many women on blood thinners (warfarin, aspirin, etc.) will need careful monitoring to make sure that these medicines are taken safely.
- Before getting pregnant, you may want to discuss the chances of your baby also having congenital heart disease. (See page 18, Genetics & Heredity.)

BIRTH CONTROL

Can I use any kind of birth control with a Fontan?

It is important to plan before getting pregnant. Thus, you must think about which kind of birth control is safe to use. Many of the birth control medicines have side effects that may cause problems for women with a Fontan. Such as, some may cause you to have more blood clots, palpitations, and high blood pressure. Talk with your adult congenital heart disease doctor about the best form of birth control for you.

- Often, women who have undergone a Fontan can use progesterone-only birth control. Examples of this include the shot (Depo-Provera), the minipill, or Nexplanon®, a small, straw-like device placed directly under the skin of the upper arm.
- IUDs may be looked at in women with a Fontan, as they are work well and provide good birth control for up to five years. The IUD needs to be placed with great care in women with a Fontan and under special watch, since women may have a lower blood pressure at the time the device is inserted.
- Estrogen should not be used in women with a Fontan. This is due to the greater chance of clots, high blood pressure,

and palpitations. Estrogen should be avoided, even more so, in women with a Fontan who already have an arrhythmia, a metal heart valve, low oxygen levels (cyanosis), or have a poor squeeze from their pumping chamber.

It is important for you to discuss birth control options with your cardiologist as well as a gynecologist to decide the best method for you. Both doctors may be needed to bring you the best options.

GENETICS & HEREDITY

How come I have this condition? Will I pass on my heart condition to my children?

The exact cause for most congenital heart problems is not known. We believe that most congenital heart disease is caused by both genetic (passed down through families) and factors in the environment. People with CHD have a higher chance of a heart defect occurring again in the family. The exact chance depends on what kind of heart defect you have and your family history.

Some people with CHD have a certain genetic condition that can be linked with other health problems. They may or may not know they have such a condition. These conditions can vary widely in how severe they are, so children may have less serious or more serious health problems than their parents.

Some genetic conditions are linked with CHD in adults. People with a history of learning problems, other birth defects, those that are short, or have other unexplained medical conditions on top of their heart defect may have an underlying genetic condition.

Rarely, heart defects are caused by changes in a single gene that is not working well. When this is the case, more than one person in the family most often has a congenital heart problem.

Because you have congenital heart disease yourself, you have a slightly higher chance of having a child with congenital heart disease. This chance is around 3–5 percent. That means that if you were able to have 100 children, between 3 and 5 of your children would likely have congenital heart disease.

For men with congenital heart disease, the chances of passing on your heart condition is around 2 percent. In a small number of women or men, the chances of passing on heart disease to

their children are higher. It is helpful to talk with your cardiologist and sometimes a genetics expert to find out what your specific chances are of passing your condition on to your children.

LIFE EXPECTANCY

How long will I live with a Fontan?

The Fontan operation changed life expectancy for babies born with single ventricle hearts. Before the operation, most children did not survive into the teenage years. The Fontan operation has given children born with a single pumping chamber a very good chance at life. We now have evidence that patients with a Fontan are living for 30–40 years after their Fontan surgery. The only reason that we don't see patients who have their Fontan for longer is that the operation only began in the early 1980s on a world-wide basis. People with a Fontan have simply not had long enough to prove that they can live for longer.

With the newer forms of the operation (the lateral tunnel and extracardiac Fontans), we expect almost 100 percent of affected children to get to adult life with a Fontan. The times during surgery carry the most risk. This is before and after children receive “shunt” operations as they are being prepared to get the Fontan operation. Once the Fontan operation is done and the child has had a successful operation, we can expect very good chances of living into the 30s and 40s, and likely beyond that as well. It is crucial that you see an adult congenital heart disease (ACHD) specialist on a regular basis to watch for, prevent and treat issues.

LIVING WILL & HEALTHCARE PROXY

Many people with lifelong conditions like to have a plan in place should they ever be in a position where they cannot make their own decisions. For example, if you were ever unable to speak for yourself, then your wishes about how you want to be treated can still be carried out. This is very helpful for doctors and nurses and caring for you. Speak to your cardiologist, spouse, family members and those close to you to help you come up with a good plan should this ever happen.



CHAPTER 4

Being Heart Healthy

Being Heart Healthy

Is there anything I can do to improve my health with a Fontan circulation?

PHYSICAL HEALTH

It almost goes without saying that a healthy lifestyle is vital to be and feel healthy. Eating well, being active, keeping a healthy weight, getting enough sleep, dealing with stress and attention to your spiritual and emotional health are all key parts of a healthy lifestyle. In this section, we address how to live a healthy lifestyle with a Fontan circulation.

HEALTHY WEIGHT

You can tell if you are at a healthy weight by looking at your body mass index or BMI. Your healthcare provider can do this, or you can do it yourself by taking your height and weight and putting the numbers in the Centers for Disease Control (CDC) online BMI calculator: http://www.cdc.gov/healthyweight/assessing/bmi/adult_bmi/english_bmi_calculator/bmi_calculator.html.

If your BMI is between 25 and 30, you are overweight. If it is 30 or above, you are in the obese grouping. Changing your lifestyle and getting help from your doctor can help you get and stay at a healthy weight.

DIET

Most of the time, eating well means eating only a small amount of processed food [e.g. hot dogs, ham, ready meals], sugar, and saturated fat [e.g. fried foods, fries]; eating proper amounts of whole grains, fruits and veggies, lean proteins, and low-fat dairy products; and drinking water instead of sugary drinks such as sodas or juices. More information on healthy eating can be found at the U.S. Department of Agriculture websites:

- <http://www.cnpp.usda.gov/2015-2020-dietary-guidelines-americans>
- <http://www.cnpp.usda.gov/MyPlate>

Appropriate Food Portions

The MyPlate image shows how an ideal meal plate is made up of one-half of the plate with fruits and veggies, one-quarter of the plate as whole grains, and the final quarter of the plate with a protein source such as a lean meat, fish, or eggs. Added

sugars and saturated fats should be less than 10 percent of your total calories. Limit salt to 2,300 milligrams per day. Food label reading can help you meet these guidelines. You can learn more by going to: http://health.gov/dietaryguidelines/dga2005/healthieryou/html/tips_food_label.html.

Around five servings of fruits and veggies should be included in your daily diet. Fruits and veggies are a great source of nutrients, are low in salt, low in fat, and low in calories. MyPlate also suggests making half of your grain use for the day whole grains. Whole grains are a good source of vitamins, minerals, and fiber, and lead to heart health. The suggested amount of dietary fiber per day is 25–35 grams. Fiber has cardiovascular benefits, and is good for digestive health.

Those on medically prescribed diets may have to adjust these general recommendations.

Sodium

A high-sodium (salt) diet will raise blood pressure over time and in turn raise the risk of heart disease and stroke. The American Heart Association suggests less than 1500 mg of sodium each day. To help stay on track with your intake, it is important to look at the nutrition facts label and buy items that have less than 140 mg of sodium per serving. Most of the sodium in our diet comes from processed foods and foods we order when we are out at restaurants. Cooking your own meals and choosing more fresh foods will help keep your salt intake down. Try avoiding adding salt during cooking or while at the table and that will help to lower your daily intake.

Fats

Dietary fat is important in the everyday diet, but you have to look at the type of fat you are eating. Saturated and transaturated (trans) fat raise LDL, or “bad,” cholesterol levels, which lead to heart disease. Unsaturated fats like canola and flaxseed are helpful when eaten in moderation. Saturated fats are found in foods such as desserts, cheeses, ice cream, fried foods, pizza, whole milk, and full-fat dairy foods. Limit and switch out these foods to include healthier versions as often as possible.

Remember these few things when you start each day. Eat to live healthy and keep your heart healthy.

SLEEP

Almost everyone agrees that most Americans do not get enough sleep. Poor sleep can lead to feeling tired, decreased activity, mood changes, and weight gain. Adults should get 7–8 hours of sleep a night. The National Institutes of Health (NIH) offers these recommendations for good sleep:

- Go to bed and wake up at the same time each day.
- Try to keep the same sleep schedule on weeknights and weekends. Limit the change to no more than about an hour. Staying up late and sleeping in late on weekends can disrupt your body clock's sleep–wake rhythm.
- Use the hour before bed for quiet time. Avoid hard exercise and bright artificial light, such as from a TV or computer screen. The light may signal the brain that it's time to be awake.
- Avoid heavy and/or large meals within a couple hours of bedtime. (Having a light snack is okay.) Avoid alcohol before bed.
- Avoid nicotine (such as, cigarettes) and caffeine (which is found in things like soda, coffee, tea, and chocolate). Nicotine and caffeine are stimulants, and both can lead to problems with sleep. The effects of caffeine can last as long as eight hours, so a cup of coffee in the late afternoon can make it hard for you to fall asleep at night.
- Spend time outside each day (when possible) and be physically active. Keep your bedroom quiet, cool, and dark (a dim night light is fine, if needed).
- Take a hot bath or use techniques to help you relax before bed.

For more information, visit the NIH website: <http://www.nhlbi.nih.gov/health/health-topics/topics/sdd/strategies>.

EXERCISE

How much exercise can I do? Are there certain kinds of exercise that are better for me?

Like with any other forms of heart disease, it is important to exercise on a routine basis if you have a Fontan. The benefits of exercise are many. They involve having a healthier and stronger mind, strong muscles, and a cardiovascular system that works better as a result of exercise. For many, this means that they feel better and also live longer.

Limitations

In most cases, Fontan patients have few limitations, but there may be some patients who may have to be more careful than

others. Talk with your doctor about the level of exercise that's right for you. If your Fontan function is not the best, you may need to choose more mild aerobic exercise where you raise your heart rate only to around the 110–130 range in adults (when you don't have a pacemaker). If this is the case, you must also avoid lifting heavy weights. For some Fontan patients, swimming may be better than other forms of exercise, because it helps to take the back pressure off the liver and bowels when you exercise.

Strength Building

Building strength of the muscles around the chest wall is important. This can be done with special exercises that your adult congenital heart disease doctor and exercise physiologist can suggest. Swimming and rowing are good forms of exercise that may help make your muscles strong, like your chest muscles.

Types of Activities



Physical activity is vital for good health and feeling good. Being active can improve your mood as well. Try to find activities that you like to do. Remember that any amount of activity is better than none. To maintain good health benefits, the CDC suggests a blend of aerobic activity and activity to make your muscles stronger:

Aerobic Exercise

Do 2.5 hours each week of moderate-intensity aerobic physical activity such as brisk walking, swimming, or cycling.

Strength Exercise

To strengthen muscles, do activities such as mild lifting weights or using resistance bands that are moderate intensity and involve all major muscle groups. Aim to do these two or more days per week.

If you have any medical restrictions, follow your doctor's advice. You can find more information on the CDC website: <http://health.gov/paguidelines/guidelines/chapter4.aspx>.

MENTAL HEALTH & COPING

How do I cope with feeling different from others and feeling isolated at times? How do I manage guilt about my heart conditions?

Managing Guilt and Shame

Guilt can be a sign that we need to make a change in the way we behave. It helps us form relationships and keeps us from repeating mistakes. Some people may feel guilty about things that they cannot change or that are not their fault. This guilt is not healthy, because it does not serve a purpose.

Shame is a bad feeling about one's self. Shame comes from the view that we are not worthy or not good enough. Shame and guilt are very painful, strong feelings that work together in a hurtful way. People living with a single ventricle heart face many challenges. Limits and restrictions highlight differences and may make it harder to feel like you fit in. Over time, fear of being judged or being disappointed can lead to avoiding social activities and isolation. This cycle can have harmful effects on mental health and well-being. That is why it is so important to challenge harmful thoughts and feelings when dealing with a heart condition.

To cope with feelings of shame and guilt, and develop a healthy self-view, the first step is to understand common sources of shame and guilt:

- Needing support or depending on others
- Causing other people worry
- Having to cancel plans or miss work
- Worry about not fulfilling roles as an employee, student, parent, friend, or partner

The second step is acceptance and connection. Talking about medical issues and emotions can be hard for some people, even more so if you are a very private person. It may be helpful to talk with a social worker or psychologist for guidance and support.

Finally, understand your role as part of your care team and make a plan. A good state of mind about treating heart conditions has been found to improve health. Start by making a list of your interests and what you enjoy most. This will help you understand your strengths and how to use them. Then, focus on what behavior you can change. Setting small goals improves mood and trust in yourself. As you learn to problem-solve, you will feel more connected, hopeful, and motivated.

Managing Stress



Stress has almost become a way of life. Reducing stress is vital to good health and a few simple tips may help. In addition to the other healthy lifestyle recommendations, the CDC suggests to:

Avoid drugs and alcohol.

They may seem to be a brief fix to feel better, but in the long run drugs, and alcohol can create more problems. This will add to your stress—instead of taking it away.

Find support.

Seek help from a partner, family member, friend, counselor, doctor, or clergy person. Having someone with a caring, listening ear to share your problems with can really lighten the burden.

Connect socially.

After a stressful event, it is easy to isolate yourself. Make sure that you are spending time with loved ones. Think about planning fun activities with your partner, children, or friends.

More information is available at: <http://www.cdc.gov/features/copingwithstress/>.

ALCOHOL AND DRUGS

Another very common question that patients ask is “Am I allowed to drink alcohol, smoke or use recreational drugs if I have a Fontan?”

While there are no national or international recommendations on drinking alcohol, smoking, or using recreational drugs in people with Fontans, we recommend great caution with these because of the effects they may have on the heart function.

Alcohol

Chronic use of alcohol may damage the liver, which is of special concern in the Fontan patient who is likely to already have some liver damage. Alcohol is also known to cause arrhythmias, which is not safe for Fontan patients. This is even more unsafe in those who have a history of arrhythmias.

Smoking

Smoking is known to cause lung damage and makes it more likely for you to develop lung cancer. The Fontan needs the lungs to work very well, in order for the blood to flow healthily through the lungs. When the lungs are damaged, the Fontan does not work as well. It starts to cause problems with your ability to exercise, but also causes your Fontan to create more damage to organs such as your liver.

Recreational Drugs

Recreational drugs are very dangerous even with casual use. For example, cocaine and other stimulants are known to damage the heart muscle and in some cases can cause heart attacks. Other drugs, like heroin and LSD, may cause high blood pressure or cause serious blood infections that will damage the heart. Among all drugs, IV (intravenous) drugs, such as heroin, are the most dangerous as they raise your risk of bloodstream infections, which can then lead to life-threatening infections of the heart.

Marijuana is increasingly being shown to have dangerous side effects in some patients with heart disease. For example they may have heart attacks, arrhythmias, or some have even died unexpectedly. Despite the use in patients with a single ventricle, there is no data yet about the safety of marijuana in patients with a single ventricle and a Fontan circulation.

MEDICATIONS

What kind of medicines should I expect to take if I have a Fontan?

Most people with a Fontan will need medicines of some sort. Those prescribed most often are:

- Aspirin and/or warfarin—these are blood-thinning medicines used to prevent life-threatening blood clots. See also chapter 3.
- Beta blockers (e.g. metoprolol or carvedilol) and ACE inhibitors (e.g. lisinopril or Enalapril) or ARB's (e.g. Losartan or Irbesartan)—these are used to take the strain off the heart by lowering and controlling heart rate and blood pressure. These medicines can also prevent stiffening of the heart muscle and blood vessels over time.
- Furosemide and spironolactone—these are diuretic medicines that are used to control extra fluid build-up and therefore prevent swelling of the legs and other parts of the body.





Appendix



Appendix

GLOSSARY

ACHD (adult congenital heart disease): refers to healthcare provided to adults who were born with heart disease. Gains in surgery and medical management of children born with congenital heart disease, have increased the number of adults 18 years and older with CHD. Adult congenital heart disease is now recognized as a specialized field like electrophysiology (heart rhythm doctors) or heart failure. The medical knowledge needed to care for adults with CHD is large and complex, and it calls for special training for doctors to become good at it and to know the latest and best treatments available. There are now also specialized centers across the USA and worldwide that focus on the treatment of adults (and children) with CHD. See <http://www.achaheart.org/home/clinic-directory.aspx>.

Aorta: The main artery that leads fresh blood from the heart to the body.

Arrhythmia: When the normal heartbeat changes to an abnormally fast, or abnormally slow, or is not normal in some other way. When this happens, the patient feels “palpitations” or what some call “flutter.”

Atrium (plural is atria): The filling chamber in the heart. The atria help to fill the ventricles so that they can do their job of pumping well.

Fontan: A heart operation performed in people born with a heart problem in which there is only one ventricle.

Glenn operation: In 1958, Dr. William Glenn made an operation where the superior vena cava (SVC) is stitched straight to the right lung (pulmonary) artery by the surgeon. In this way the superior vena cava no longer goes back to the heart, but instead brings used (oxygen poor) blood straight back to the lung (pulmonary) arteries.

IVC (inferior vena cava): The main vein that drains blood from the lower half of the body (legs, belly, and lower half of chest) back to the heart.

SVC (superior vena cava): The main vein that drains blood from the upper half of the body (head, neck, arms, and upper chest) back to the heart.

Ventricle: The pumping chamber of the heart. It has thickened muscular walls that help it pump blood to the body or to the lungs. There are two ventricle chambers—one on the right and one on the left. Most Fontan patients only have one normal sized working ventricle.

Support & Resources



When you are faced with a lifelong heart condition such as adult congenital heart disease, it can help to connect with others who get what you're going through. Support groups offer a way to bring people together to share information. This can also help reduce stress and worry, and provide helpful education. Support groups that may be of interest to adults with congenital heart disease include:

Adult CHD Survivors of Cincinnati

Offers support and teaching to adult congenital heart disease (ACHD) patients and their family and friends, including a monthly support group.

survivingchd@gmail.com

Congenital Heart Network of Greater Cincinnati

Offers direct support and teaching to improve the lives of those with CHD and their families.

pcha.oh@conqueringchd.org | conqueringCHD.org/Ohio

Mended Little Hearts

Offers teaching, resources, and access to local support groups for families and caregivers of children with CHD.

[888-432-7899](tel:888-432-7899) | mendedlittlehearts.org

The Adult Congenital Heart Association (ACHA)

Offers teaching and resources for adults and adolescents with CHD.

[888-921-ACHA](tel:888-921-ACHA) | achaheart.org

Cincinnati Children's Fontan Management Clinic

[513-803-2243](tel:513-803-2243)

THE HEART INSTITUTE



Contact Us At...

513-803-2243

Monday–Friday, 8 am–4:30 pm

513-636-4200

Weekends, Nights & Holidays for the hospital operator

3333 Burnet Avenue, Cincinnati, Ohio 45229-3026



KN-1128 09/18
BRV296310 0519 000300