

Congenital Heart Center

# Single Ventricle Congenital Heart Defects

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*A Guide for Patients,  
Parents and Families*



MICHIGAN MEDICINE

# Single Ventricle Congenital Heart Defects

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The Congenital Heart Center at University of Michigan Health C.S. Mott Children's Hospital has created this booklet to support and educate patients and families about single ventricle congenital heart defects. The terms “congenital heart disease” and “congenital heart defects” are often used interchangeably – while the ‘defect’ is what happens to the heart we know that the defect affects other parts of the body, quality of life, and the family so the term ‘disease’ is also used. Throughout this booklet there is detailed information about different types of single ventricle heart defects and what can be done to treat them. There are also diagrams to show the different heart conditions and the various surgeries that may be used to treat them. Use this booklet as a reference to support and guide you in conversations with your heart care team and to educate yourself and family members about single ventricle heart defects.

For additional information, please visit [www.mottchildren.org/congenital](http://www.mottchildren.org/congenital)

We recommend the *Your Heart Guidebook* found under Resources for Patients.

# Table of Contents

Introduction to Congenital Heart Defects and Single Ventricle Hearts .....	4
Understanding the Normal Heart .....	6
Understanding Single Ventricle (SV) Hearts .....	8
Types of Single Ventricle Hearts .....	9
Complex Single Ventricle Hearts .....	14
Diagnosing Single Ventricle Hearts .....	18
Newborn Life with Single Ventricle Hearts .....	21
Treatment for Single Ventricle Heart Disease .....	23
Stage 1.....	23
Stage 2 and 3.....	25
Outcomes with Surgery for Single Ventricle Hearts .....	26
Putting it All Together – Diagrams of Single Ventricle Hearts and Surgical Procedures .....	28
What to Expect After Surgery .....	37
Planning for Discharge .....	41
Choosing a Heart Center for Single Ventricle Heart Care .....	44
Lifelong Considerations .....	47
Family Support .....	50
Conclusion .....	54
References.....	55

# Single Ventricle

## Congenital Heart Defects

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### Introduction to Congenital Heart Defects and Single Ventricle Hearts

A **congenital heart defect (CHD)** occurs when the heart does not develop normally before birth. These defects typically happen very early in pregnancy, often before the mother knows she is pregnant. There are many different types of CHDs. While there are some genetic and possible environmental causes, the exact cause is not known for most patients and CHD is not usually preventable.

#### What is a single ventricle heart?

A normal heart has four chambers. There are two upper chambers – a right and left atrium. The **right atrium** receives blood returning from the body while the **left atrium** receives the blood returning from the lungs. Blood goes from each atrium to the two lower chambers which are the pumping chambers and are called **ventricles**. In a normal heart the **right ventricle** pumps blood to the lungs and the **left ventricle** pumps blood to the body.

Some types of CHDs result in a heart with only one working ventricle. Many children with a single ventricle (SV) heart are otherwise healthy with no other noncardiac problems. Others may have other medical problems including other heart defects, delays in normal development or genetic syndromes. **There are multiple types of single ventricle (SV) conditions, many of which are discussed in more detail in this booklet.**

## **How often do congenital heart disease and single ventricle occur?**

Reports vary on how many babies with CHD are born each year, but some approximate numbers are:

- CHD (overall): 1 in 110 babies (about 40,000 babies each year in the U.S.)
- Hypoplastic left heart syndrome (HLHS): 1 in 4,344 babies (about 960 babies each year in the U.S.)
- Hypoplastic right heart syndrome (HRHS):
  - Tricuspid atresia (underdeveloped tricuspid valve): 1 in 10,000 babies (about 400 babies each year in the U.S.)
  - Pulmonary atresia (underdeveloped pulmonary valve): 1 in 7,700 babies (about 520 babies each year in the U.S., but not all are single ventricle hearts)

# Understanding the Normal Heart

To understand a single ventricle heart, the first step is to understand the normal heart (**Figure 1: all bolded terms below are shown in the diagram**). A normal heart has four chambers. The two upper chambers are called **atria** (left atrium and right atrium) and they receive the blood returning from the body and the lungs. The two lower chambers are called **ventricles** (left ventricle and right ventricle). The ventricles are muscles that squeeze to push blood to the body and lungs. Each ventricle (pumping chamber) has an entrance and an exit valve, which are one-way doors that make sure blood does not go backwards.

A normal heart does two things every time it beats:

1. The right side takes “blue” (low oxygen) blood coming back from the body and pumps it to the lungs, where the blood gets oxygen.
2. The left side takes “red” (high oxygen) blood coming back from the lungs and pumps it to the body.

Here are the steps of how that happens:

- Blood from the body (blue blood) enters the heart through two large veins, the **superior vena cava (SVC)** and the **inferior vena cava (IVC)**. Blood enters the **right atrium** (the collecting chamber on the right side of the heart).
- From the right atrium, blood enters the **right ventricle** (pumping chamber on the right side of the heart) through the **tricuspid valve**.
- The right ventricle squeezes and the **pulmonary valve** opens. Blood flows into the lungs through the **pulmonary artery**. Blood vessels in the lungs have low pressure and expand easily (low resistance) so it is an easy job to send blood there as the right heart does not have to create a lot of pressure.
- After the lungs add oxygen and remove carbon dioxide, the blood comes back through the **pulmonary veins** to the **left atrium** (the collecting chamber on the left side of the heart).
- From the left atrium, blood enters the left ventricle (pumping chamber on the left side of the heart) through the **mitral valve**.

The ventricle squeezes and the **aortic valve** opens and blood flows to the body through the **aorta**. It takes a lot more work to send blood to the body, so the left ventricle must create a higher pressure.

- There is a wall between the right and left atria (collecting chambers) called the **atrial septum** which separates red blood (oxygen-rich blood in the left atrium) from blue blood (oxygen-poor blood in the right atrium).
- There is also a wall between the right and left ventricles (**ventricular septum**) separating the red and blue blood inside the ventricles.
- The **ductus arteriosus** is a normal blood vessel in the developing fetus that connects the lungs (**pulmonary artery**) to the body (**aorta**). Before birth it detours blood away from the lungs since they are not working yet (fetal lungs are filled with fluid in the uterus). After birth, the **ductus arteriosus** usually stays open for a few hours or days (sometimes even longer) but it is supposed to close soon after birth.
- Having a **patent (open) ductus arteriosus** (called a PDA) can be very important to babies with a single ventricle heart defect. The PDA can provide a *temporary* way to get blood to the lungs (HRHS) or blood to the body (HLHS) when the normal route of blood flow is blocked or is not adequate.

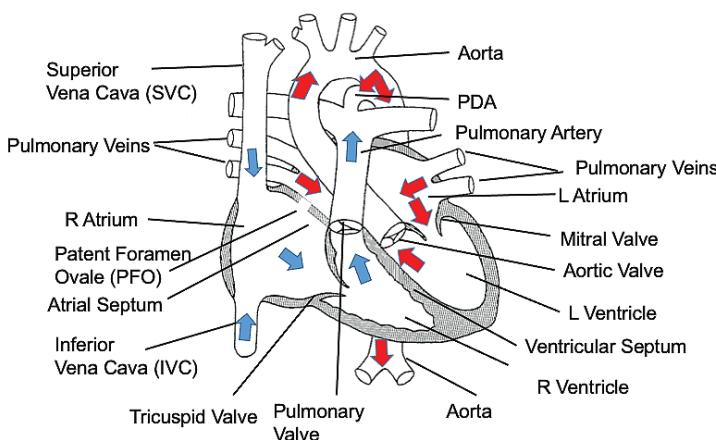


Figure 1: The normal heart (PDA)

**For all heart diagrams:**

**blue arrow** = oxygen-poor blood that has returned from the body

**red arrow** = oxygen-rich blood that has received oxygen from the lungs

**purple arrow** = a mixture of oxygen-poor blood and oxygen-rich blood

# Understanding Single Ventricle (SV) Hearts

Since there are many different types of SV hearts, it is important for us to describe the details about the exact type of heart defect to each family. All CHD conditions interfere with the normal circulation of blood in and out of the heart because one of the pumping chambers is not able to do its job.

## What are hypoplastic ventricles?

Either the right or left ventricle may be underdeveloped. This is called “**hypoplastic**.”

1. If the right ventricle is too small, it is called a **hypoplastic right ventricle**. The right ventricle moves low-oxygen (blue) blood to the lungs to receive oxygen. If it is too small, there is not a normal path for blood to get to the lungs to pick up oxygen.
2. If the left ventricle is too small, it is called a **hypoplastic left ventricle**. The left ventricle pumps high-oxygen (red) blood to the body. If it is too small, there is not a normal path for blood to get to the body to deliver oxygen.

## What causes hypoplastic ventricles?

One of the ventricles not growing to normal size (hypoplastic) can be caused by the following:

- The ventricle does not have a good entrance for blood to flow into it — the entrance valve may be too narrow (stenosis) or have no opening (atresia).
- The ventricle doesn’t have a good exit for blood to be pumped out — the exit valve may be too narrow (stenosis) or not open at all (atresia).
- Any combination of the above.

## Types of Single Ventricle Hearts

### What is hypoplastic left heart syndrome (HLHS)?

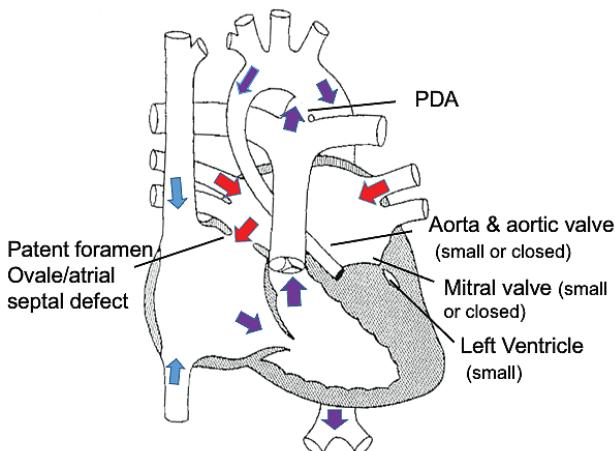
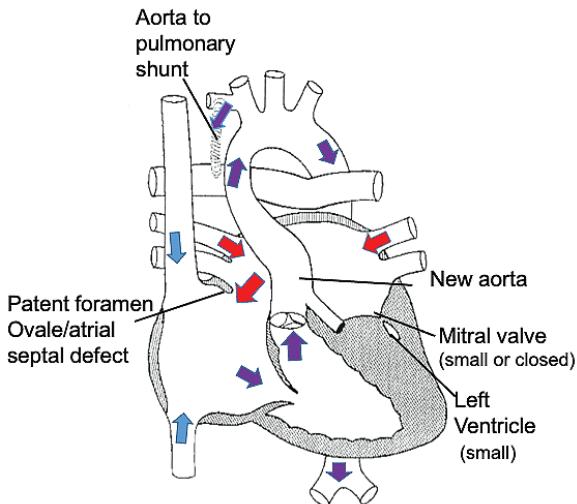


Figure 2: Hypoplastic left heart syndrome (HLHS)

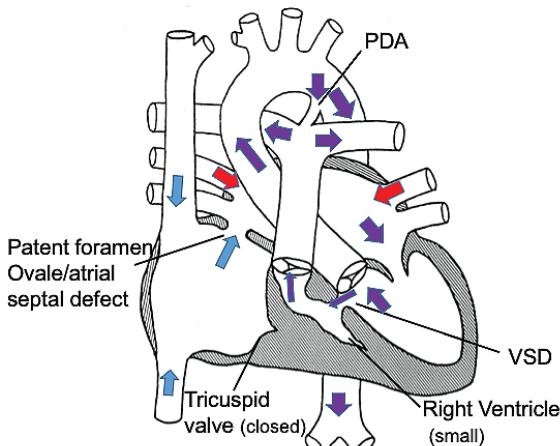
- The left ventricle is too small and does not squeeze (pump) normally.
- The left ventricle's entrance (mitral valve) and exit (aortic valve) are very small or closed completely.
- The aorta (artery to the body) is also small.
- Because of these problems, there is not a normal path of blood from the heart to the body. The body must rely on the patent (open) ductus arteriosus (PDA) to get blood to the aorta, so the body gets the oxygen and energy it needs. The PDA is a short-term solution (discussed later in this booklet).
- The first surgery that is done for HLHS is called a Norwood procedure.
- The **Norwood procedure** will create an unobstructed pathway to get oxygenated blood to the body, and a pathway for blue blood to get to the lungs.



**Figure 3: Norwood with Blalock Taussig shunt**

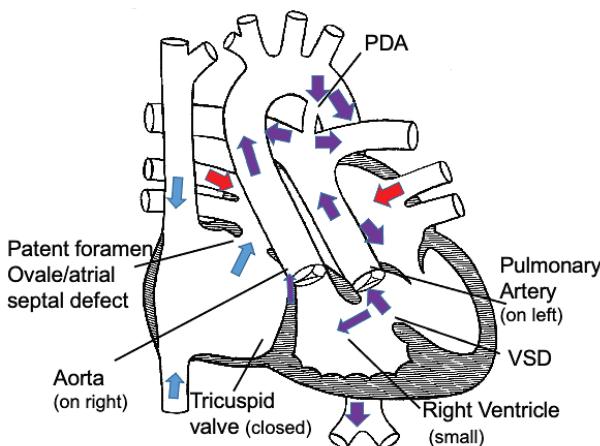
## What is hypoplastic right heart syndrome (HRHS)?

### Tricuspid atresia



**Figure 4: Tricuspid atresia with normally related great vessels & VSD**

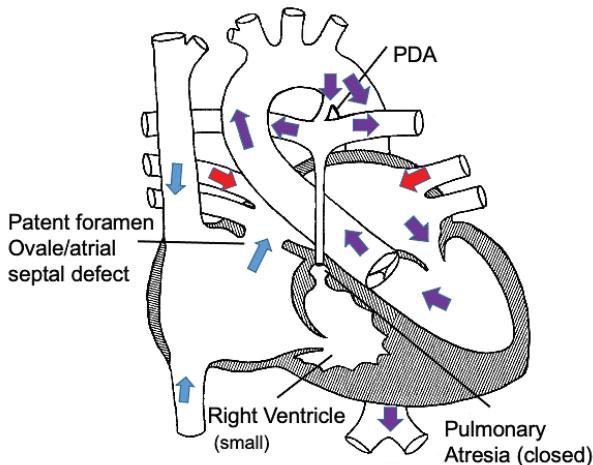
- The entrance to the right ventricle (the tricuspid valve) does not develop at all and blood cannot flow into the right ventricle normally.
- The right ventricle itself is very small (hypoplastic).
- The normal exit (the pulmonary valve) may be small (pulmonary stenosis) or completely closed (pulmonary atresia) or may be normal in size.
- Because of these problems, there is not a good path of blue blood to the lungs, so the body must rely on the **patent (open) ductus arteriosus (PDA)** to get blood to the lungs to pick up oxygen.
- With tricuspid atresia there may also be a **ventricular septal defect (VSD)**, which is a hole in the wall between the right and left ventricles. Blood can cross this hole and get out to the lungs through the small right ventricle and the pulmonary artery. This blood flow may allow the pulmonary valve and artery to develop to a more normal size.
- Tricuspid atresia may be found with an additional CHD, **transposition of the great arteries (TGA)**. With TGA the pulmonary artery comes off the left ventricle and the aorta comes off the right ventricle, which is the opposite of normal. When the aorta comes off the small right ventricle, it may also be small and there may not be a good path of blood to the body. If the aorta is small, the body must rely on the **PDA** for blood flow to the aorta, so the body gets the oxygen and energy it needs.



**Figure 5: Tricuspid atresia with transposition of great vessels & VSD**

- The type of surgery that is done depends on which blood vessel is coming off the right ventricle (the pulmonary artery or the aorta) and the size of the blood vessel. The first surgery could be a pulmonary artery banding, a systemic artery to pulmonary artery shunt, or a Norwood procedure (these surgeries are described later in this booklet).

## Pulmonary atresia with intact ventricular septum



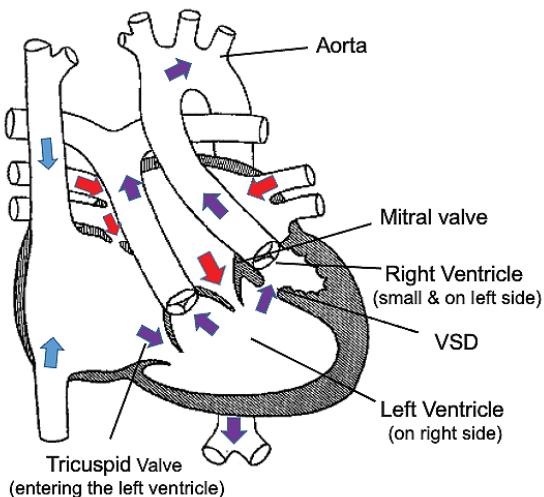
**Figure 6: Pulmonary atresia with intact ventricular septum**

- The right ventricle's exit (pulmonary valve) does not form normally. The valve is completely closed (atresia) so blood cannot flow normally from the right ventricle to the lungs. Pulmonary atresia can occur with or without a ventricular septal defect (VSD), but the focus here is without a VSD, which can cause a single ventricle heart.
- Intact ventricular septum means that the wall between the ventricles formed completely so there is no hole (ventricular septal defect) in the wall between the ventricles.

- When there is pulmonary atresia and no VSD, the right ventricle has decreased blood flow, may not grow well and may be too small (hypoplastic) when the baby is born. In some babies, the right ventricle may be big enough and may not be considered a single ventricle heart. With pulmonary atresia, there is not a good path for blood to flow to the lungs, so the body must rely on the **PDA** to get blood to the lungs to pick up oxygen.
- The arteries that provide blood flow to the heart muscle itself are called **coronary arteries**. In some cases of pulmonary atresia with an intact ventricular septum, the coronary arteries in the muscular walls of the small right ventricle are very abnormal and can cause major problems in the flow of blood to the heart muscle in the left ventricle. These abnormal coronary arteries are called **sinusoids**. They can increase the child's risk of problems both before and after surgery.

# Complex Single Ventricle Hearts

## Double inlet left ventricle



**Figure 7: Double inlet left ventricle with levo transposition of the great vessels**

- The upper chambers of the heart (atria) empty through the mitral and tricuspid valves into only one ventricle (the left ventricle) instead of both the left and right ventricles.
- The right ventricle doesn't have an entrance valve, so it is often small and underdeveloped.
- The ventricles may be in the wrong place. The left ventricle may be on the right side of the heart while the right ventricle may be on the left side. The position of the ventricles does not affect the ability of the heart muscle to pump blood.
- The arteries leaving the heart (pulmonary artery and aorta) can come off either ventricle. The type of surgery that is done depends on which vessel is coming off the small right ventricle (the pulmonary artery or the aorta) and the size of the blood vessel. The first surgery could be a pulmonary artery banding, a systemic artery to pulmonary artery shunt, or a Norwood procedure (these surgeries are described later in this booklet).

## Shone's complex

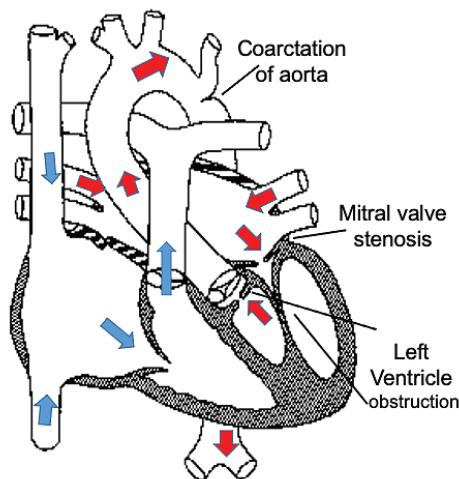


Figure 8: Shone's complex

- Many parts on the left side of the heart do not develop normally, but the left ventricle may be more normally developed.
- The mitral and aortic valves may not be big enough or open well.
- There can be tissue blockages in the left atrium, above the mitral valve or below the aortic valve.
- The aorta may be too small and may have a narrowed area (coarctation of the aorta).
- Depending on the severity of these abnormalities, Shone's complex can be a type of hypoplastic left heart syndrome (HLHS). Please refer to the HLHS section above.

## Unbalanced atrioventricular septal defect

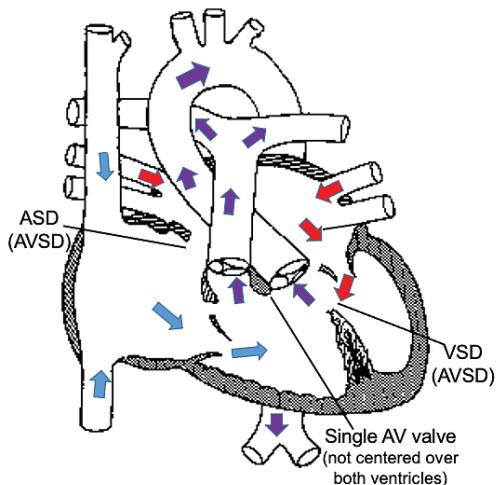
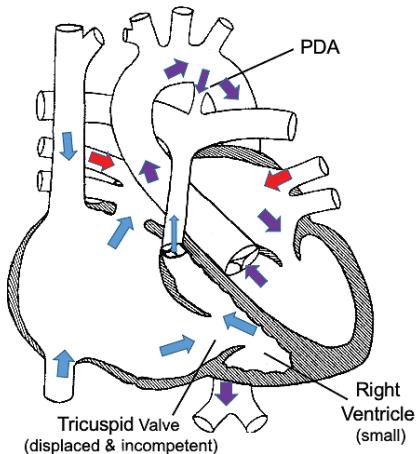


Figure 9: Unbalanced atrioventricular septal defect (AVSD)

- The mitral and tricuspid valves start out as a single valve early in the development of the heart before they eventually divide. Sometimes the division into two valves doesn't happen. In some cases, the single common valve drains mostly to just one ventricle. This defect is called an **unbalanced atrioventricular septal defect (AVSD)**. If the unbalance is severe, this results in a single ventricle heart.
- There are two types of unbalanced AVSD:
  - Unbalanced AVSD to the right ventricle may result in a small left ventricle. If severe, this creates a type of hypoplastic left heart syndrome (please refer to HLHS section above).
  - Unbalanced AVSD to the left ventricle may result in small right ventricle. If severe, this creates a type of hypoplastic right heart (please refer to HRHS section above).

## Ebstein's anomaly



**Figure 10: Ebstein's anomaly**

- The formation of the tricuspid valve (entrance to the right ventricle) is abnormal.
- The tricuspid valve is not in the normal position and it usually leaks. The leakage can be mild to severe. In a more severe form, the right atrium is very big, and the right ventricle can be small or not work well. If severe, this can result in a type of hypoplastic right heart (please refer to HRHS section above).

## Heterotaxy syndrome with single ventricle heart defects

- Heterotaxy is when some parts of the body in the chest and abdomen (belly) do not form in the right place. People with heterotaxy often have complicated congenital heart defects (CHDs) and can have single ventricle defects.
- Other possible defects include:
  - The veins that drain blood from the body (superior and inferior vena cava) and the lungs (pulmonary veins) may connect to the wrong part of the heart.
  - The heart may be located more on the right side of the chest than the left side.
  - The liver and the stomach may be in the wrong position in the belly.
  - There may be a normal, an absent, or multiple spleen(s).

# Diagnosing Single Ventricle Hearts

CHDs can be found in several ways at different times in the pregnancy or after birth.

## Prenatal (before birth) diagnosis

Sometimes a routine prenatal ultrasound may show a possible congenital heart defect (CHD). An **ultrasound** is a procedure that uses high-energy sound waves to look at tissues and organs. Sound waves produce an echo that forms pictures of the tissues and organs on a computer screen (sonogram). If an ultrasound shows a suspected heart problem, a referral is made for further evaluation by a pediatric cardiologist (a heart doctor) who can perform a fetal heart echocardiogram. A **fetal heart echocardiogram (echo)** is an ultrasound that specifically looks at all the parts of a fetal heart in great detail. Once the diagnosis is made, parents can receive information on the specific heart defect, what to expect during pregnancy and after delivery, and a generalized long-term outlook.

## Time to prepare

Learning about a heart problem while still pregnant gives the parents and family time to prepare for the baby's birth. The family and cardiology team can discuss the plan and location of delivery of the baby that will work the best for the situation.

Here are some ways a family can prepare for a child's CHD journey:

- Determine who will be the baby's pediatrician/primary care provider and the pediatric cardiologist (heart doctor). It is best to select these health care providers before the baby is born. Family or friends may have recommendations about primary care doctors in your area, or you can ask your heart care team for recommendations.
- Be sure all family members and caregivers are up to date on vaccines as well as the flu shot when recommended. The baby will not be able to get vaccines right away and could be at greater risk than other babies.

- Connect with other families who also have a child with a heart defect. Some nationally recognized social media groups may have local chapters (see Family Resource section of this booklet).
- Consider the support available through family and friends to assist through these challenging times. Think about who can help with other responsibilities while you need to take care of your baby. Planning ahead can help reduce some stress.

## Role of fetal intervention

In rare cases, the prenatal diagnosis may lead to consideration of a fetal intervention. A **fetal intervention** is a procedure on the baby's heart before birth. Fetal intervention is limited to a small number of patients that would benefit from this very high-risk procedure. At the current time, most fetal interventions are for one of two diagnoses:

1. Critical aortic stenosis which is leading to a hypoplastic left heart.
2. A high-risk form of hypoplastic left heart syndrome (HLHS) with a restrictive atrial septum (the hole in the atrial septum is too small to let blood flow through).

If fetal intervention is an option, the fetal cardiology team will discuss these options with the family.

- In **critical aortic stenosis**, when the aortic valve is thickened and doesn't open well, but the left ventricle is still a good size, a procedure to open the aortic valve before birth may be done to hopefully promote continued growth of the left ventricle and aortic valve. This may or may not prevent HLHS.
- A fetal intervention that may be done for **HLHS with a restrictive atrial septum** (small opening or no opening between the upper heart chambers) is to open the hole. This allows blood from the lungs to cross the hole and flow out to the body through the good-sized ventricle. It may improve a baby's heart and lung function in the newborn period and increase the chance of survival.

## Postnatal (after birth) diagnosis

Most newborns with serious heart defects look well initially after delivery but will usually show symptoms in the first few days after birth. Routine newborn screening for critical CHDs might show

changes in oxygen levels before any symptoms are seen. Other babies may have abnormal findings on the newborn exam performed by the baby's health care team.

Symptoms of a CHD may include:

- Bluish skin tone.
- Fast or hard breathing.
- Poor feeding.
- Cool skin.
- Lethargy – very sleepy and less responsive.
- Heart murmur.

These symptoms will lead the nurse or doctor to look further for a diagnosis. If a problem isn't detected before a newborn is discharged from the hospital, the same symptoms may be seen at home and parents should seek medical attention.

## Diagnostic Tests

Typical tests used to assist the health care team in making a diagnosis would be an EKG, chest X-ray and echocardiogram. These tests are safe and painless. In addition, thorough physical examinations by pediatric heart care professionals will help lead to a diagnosis and treatment plan.

In most cases, all the diagnostic information can be obtained from the echocardiogram. Patients with a single ventricle heart may also benefit from other tests after the echocardiogram such as magnetic resonance imaging (MRI) or computerized tomography (CT) scans to provide additional images of the heart. Rarely, a cardiac catheterization may be done to get more information. This is a procedure done in a cardiac catheterization laboratory. A thin tube (a catheter) is inserted into the blood vessels leading to the heart. This test can provide additional diagnostic information and, in some cases, special catheters and devices can be used to treat parts of the heart defect. For more information, visit [www.mottchildren.org/chcdiagnostics](http://www.mottchildren.org/chcdiagnostics).

# Newborn Life with Single Ventricle Hearts

## What happens after birth

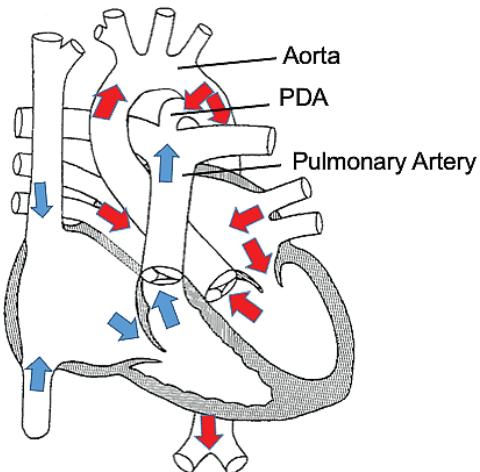
Remember, a normal heart does two things every time it beats:

1. The right side takes “blue” (low oxygen) blood coming back from the body and pumps it to the lungs, where the blood gets oxygen.
2. The left side takes “red” (high oxygen) blood coming back from the lungs and pumps it to the body.

The patent ductus arteriosus (PDA) is a normal blood vessel in the developing fetus that connects the lungs (pulmonary artery) to the body (aorta). Before birth it detours blood away from the lungs since they are not working yet (the fetal lungs are filled with fluid in the uterus). In babies with single ventricle hearts, the PDA can provide a temporary way to get blood to the lungs or blood to the body when the normal route of flow does not exist.

Because the PDA is still open for a period after birth, most newborns with single ventricle heart defects actually look well initially after delivery. However, the PDA will start to close in the first hours to couple of days of life. If it closes, it is life threatening to a baby with a single ventricle heart defect that is dependent on the PDA for blood flow to the lungs or body.

While waiting for surgery, the first step for a newborn with a single ventricle heart dependent on the PDA is to keep the PDA open. An intravenous (IV) medicine, called Prostaglandin (PGE), is used to keep the PDA open temporarily. PGE medicine is only a short-term treatment and babies must remain in the hospital and usually in the intensive care unit (ICU) when getting this medicine. There may be side effects from the medicine, especially in babies who are on it for longer periods of time. When babies first start on the medicine, it can cause them to have pauses in their breathing. Babies are given the medication form of caffeine to counteract this side effect.



**Figure 11: Patent ductus arteriosus (PDA)**

## First stage of surgery

The first surgery is done to provide a sure pathway to get blood flow to the lungs or to the body, based on which flow is inadequate. In newborn babies the goal is simply to get blood to both places even though blue and red blood are still mixing within the heart. In later surgeries, more permanent connections are made to separate the blue and red blood. The series of surgeries is necessary because newborn babies are not able to handle all the stages of surgery at once.

Depending on the anatomy of the heart defect, the heart care team will determine the best surgical plan for the first stage. The first surgery is typically scheduled in the first week of life, and will generally be one of the following (see further descriptions in the Treatment for Single Ventricle Heart Disease section):

- Norwood procedure.
- Hybrid procedure.
- Shunt from aorta to pulmonary arteries.
- PDA stent.
- Pulmonary artery banding.

## Treatment for Single Ventricle Heart Disease

When a single ventricle heart defect diagnosis is made, the family will have a discussion with the pediatric cardiologist, pediatric heart surgeon, and other members of the heart care team about the treatment options for the child.

To stay alive, we must have blood flowing to our body and to our lungs. Single ventricle hearts typically do not have both. For most single ventricle hearts, a series of palliative surgeries (usually three) can be done over the first few years of life to get blue blood to the lungs and to get red blood to the body using only the single good ventricle. These surgeries are called “palliative” because they do not “fix” the heart problem. They create alternate pathways using the working parts of the heart for blood to get to the lungs and body, bypassing the small left or right ventricle.

### **Stage 1**

There are several options for the first stage of surgery or intervention depending on the child’s specific heart defect. Each option is focused on getting adequate blood flow to the body and to the lungs. Procedures you may hear about (and are pictured later in the book) are:

#### **For hypoplastic left heart syndrome (HLHS)**

- **Norwood procedure (see pg. 29):** This is an open-heart surgical procedure requiring the use of the heart lung machine (the machine that oxygenates and pumps blood during a heart surgery). With this procedure a stable path of blood flow from the single ventricle to the body is created. It also establishes a path of blood flow to the lungs through a tube called a shunt. The shunt may be from the aorta to the pulmonary artery or from the single ventricle to the pulmonary artery.

- **Hybrid procedure (see pg. 29):** This procedure combines surgery and catheterization interventions to accomplish the same goals as the Norwood procedure but does not require use of the heart lung machine. The hybrid Norwood procedure is considered when newborn babies are too high risk to undergo the Norwood operation right away. Babies who have a Hybrid procedure will still require the full Norwood operation when they are a few months of age.

To perform the hybrid, the chest is opened like in standard heart surgery, but the heart is not opened and continues to beat. A stent is placed in the PDA to keep it open, and bands are placed around the pulmonary arteries to the lungs to prevent too much blood flow going to the lungs. If needed, the ASD (hole in the atrial septum) is enlarged.

## **For hypoplastic right heart syndrome and other single ventricle hearts (see pg. 30)**

Different surgeries may be needed depending on the specific anatomy to get the right amount of blood to the lungs and to the body:

- **Aorta to pulmonary shunt:** Provides blood flow to the lungs by placing a tube from the aorta to the pulmonary artery.
- **PDA stent:** Provides blood flow to the lungs or to the body by placing a stent to keep the PDA open.
- **Pulmonary artery band:** A band or tie cinches down the pulmonary artery to prevent too much blood flow going to the lungs.
- **Norwood procedure:** Some babies with a hypoplastic aorta will require a Norwood procedure like those babies with hypoplastic left heart syndrome.
- **No initial surgery:** No surgery may be needed if there is good balance of blood flow to the lungs and body.

In some babies with complex heart defects, there may be no surgery or procedure (including heart transplant) that can give any meaningful chance of helping the baby survive and therefore no surgery may be indicated. This is a very difficult decision and one

that is only made after thorough review and discussions among cardiologists, surgeons, and the family. Second opinions from other centers may be sought to help in this decision. In this situation, compassionate care will focus on the baby's comfort and quality of life. This can mean days, months or even years. Families have the support of our social work and palliative care teams, in addition to the heart care team, throughout this difficult process.

## **Stage 2 and 3**

Regardless of the initial surgery, babies with single ventricle hearts have a very similar second stage of surgery (either a hemi-Fontan procedure or bidirectional Glenn procedure) and third stage of surgery (either lateral tunnel or extracardiac Fontan procedure). Though the details of these surgeries may be a little different depending on each child's anatomy, the result is the same. The goal of these two stages of surgery is to separate the blue blood going to the lungs from the red blood going to the body by creating separate pathways in the heart.

**Stage 2** (hemi-Fontan or bidirectional Glenn procedure) is performed at around 4-6 months of age, but timing can vary depending on the needs of each child. The specific type of surgery will be determined by the child's specific anatomy.

**Stage 3** (lateral tunnel or extracardiac conduit Fontan procedure) is typically performed around 2-4 years of age. The Fontan procedure is elective, which means it does not have to be performed at a specific age and timing can vary depending on the needs of each child. The lateral tunnel Fontan is often planned around 18 months to 2 years of age. Surgery for patients getting an extracardiac conduit Fontan may be done at a little older age (closer to 3 to 4 years). The child's cardiologist and surgeon will determine the type of surgery based on the child's anatomy and condition.

Though the three stages of surgery are what we plan for children with single ventricle heart defects, there may be additional surgeries and procedures that are needed between the operations or after the Fontan operation.

# **Outcomes with Surgery for Single Ventricle Hearts**

## **Survival with surgery**

Before the 1980s, a child born with a single ventricle heart defect had little to no hope for survival beyond infancy. Pediatric cardiac surgeons and cardiologists at U-M and other heart care centers pioneered the surgeries and treatments described in this book. The result has been improved survival for patients with single ventricle heart defects. The surgeries and treatments we now offer give hope to so many children and their families and can allow for survival into adulthood with a good quality of life.

Even though most children with single ventricle heart defects will survive the surgeries, it is important for every family to understand that single ventricle heart defects and their treatments are very high risk. Some children do not survive the surgeries, even if they are felt to be good candidates. Other children may develop complications that could lead to their death at any point after the surgeries. The likelihood that any given child will survive through the surgeries and into adulthood will depend on each child's specific heart defect. Specific heart anatomy, as well as other problems outside the heart, can all affect a child's survival. **The cardiologist caring for your child will go over these specific risk factors and help you to understand what to expect.**

Since the time of early treatments for single ventricle hearts, our center has consistently strived to enhance outcomes for treatment options in single ventricle heart disease. As a high-volume center, we have achieved a leading role in improving single ventricle care and outcomes. For an overall look at the outcomes for U-M Congenital Heart Center, visit [www.mottchildren.org/chcoutcomes](http://www.mottchildren.org/chcoutcomes).

## **Long-term survival**

Even when children survive the planned stages of surgery, a single ventricle heart defect is never “fixed”. Patients with single ventricle hearts will need lifelong care with a cardiologist. The Fontan circulation creates problems for the body that can arise at any age after the surgery. Some problems can be treated with medical or surgical options, while others don’t have good treatments at this time. Our hope is that through continued research studying single ventricle patients, new treatments will be created to help more single ventricle patients survive and do well. For more about the Michigan Congenital Heart Outcomes Research and Discovery (M-CHORD) Program, visit [www.mottchildren.org/chcresearch](http://www.mottchildren.org/chcresearch).

## **Long-term quality of life**

Our goal is to help children with single ventricle hearts survive into adulthood. We also strive to help them thrive and enjoy an excellent long-term quality of life. In addition to medical concerns, some children with single ventricle hearts may need extra help in school or have emotional or behavioral challenges. Programs and resources are available at the U-M Congenital Heart Center to assist with early recognition and treatment of potential problems, and to optimize functioning and quality of life for those at risk for these challenges. One important example is the Cardiac Neurodevelopmental Clinic, [www.mottchildren.org/chcneurodev](http://www.mottchildren.org/chcneurodev).

# Putting it All Together – Diagrams of Single Ventricle Hearts and Surgical Procedures

## Normal heart with patent ductus arteriosus (PDA) and patent foramen ovale (PFO)

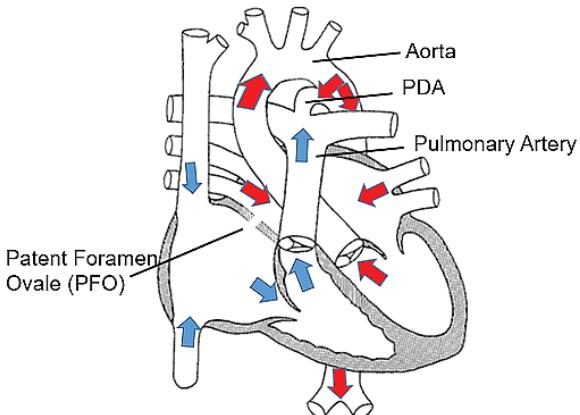


Figure 12: Normal heart with patent ductus arteriosus (PDA) and patent foramen ovale (PFO)

## Hypoplastic left heart syndrome and procedures

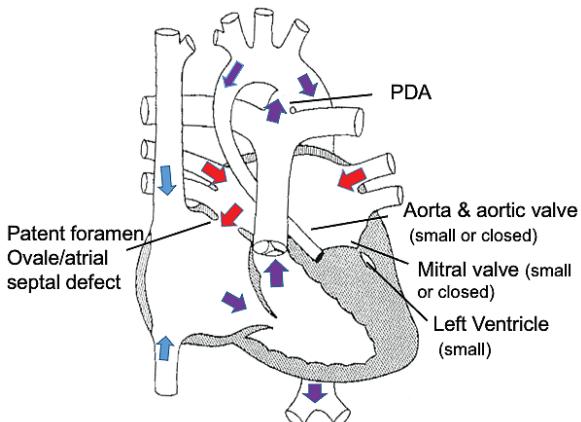


Figure 13: Hypoplastic Left Heart Syndrome (HLHS)

## Stage 1 procedures

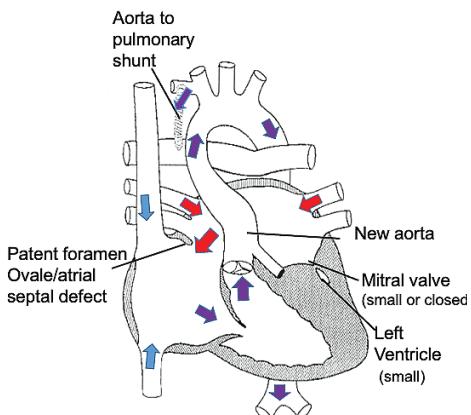


Figure 14: Norwood with Blalock Taussig shunt

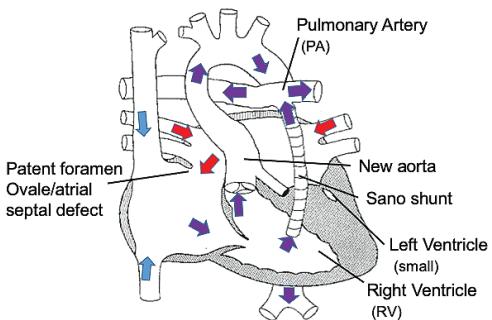


Figure 15: Norwood with Sano (RV to PA) shunt

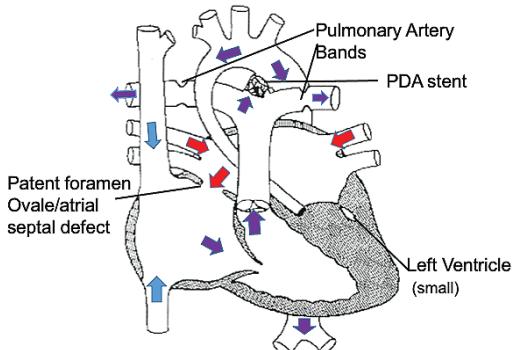


Figure 16: Hybrid

## Stage 2 procedure

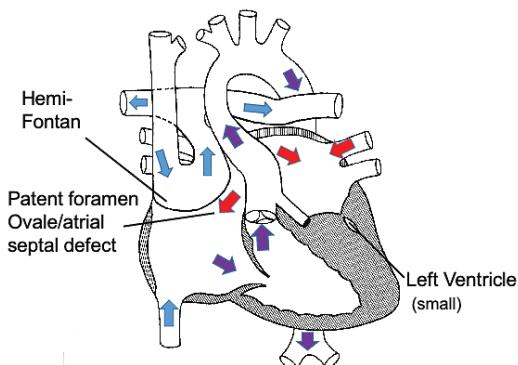


Figure 17: Hemi-Fontan

## Stage 3 procedures

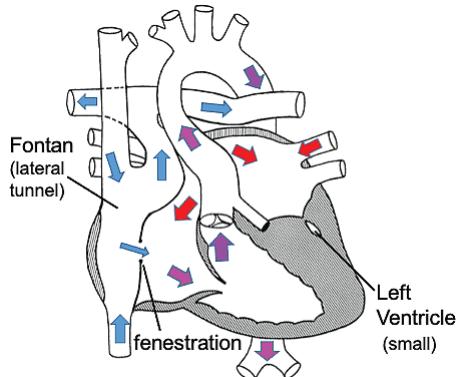


Figure 18: Fontan – lateral tunnel

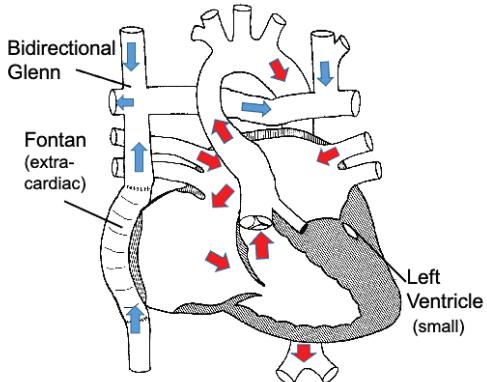


Figure 19: Fontan (extracardiac) and bidirectional Glen

## Hypoplastic right heart diagnoses and procedures HRHS: Tricuspid atresia diagnoses

**HRHS: Tricuspid atresia with normally related great vessels and VSD**

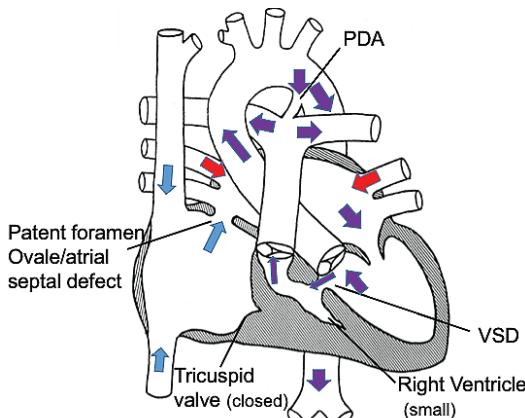


Figure 20: Tricuspid atresia with normally related great vessels & VSD

## Stage 1 procedures

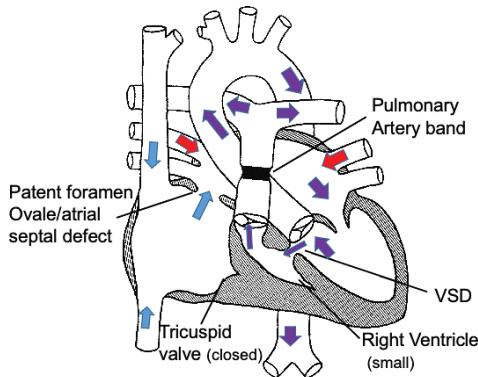


Figure 21: Stage 1 — pulmonary artery band

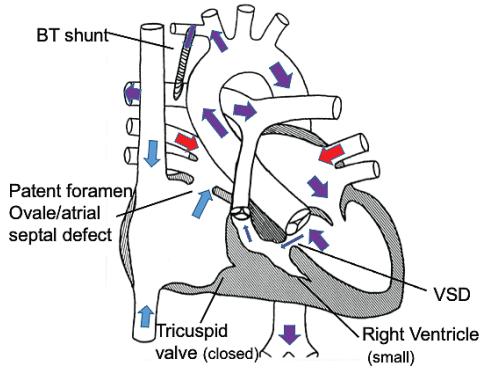


Figure 22: Stage 1 – Blalock Taussig shunt

\*Choice of stage 1 surgery depends on if pulmonary blood flow needs to be decreased or increased. If pulmonary blood flow is satisfactory this stage may not be necessary.

## Stage 2 procedure

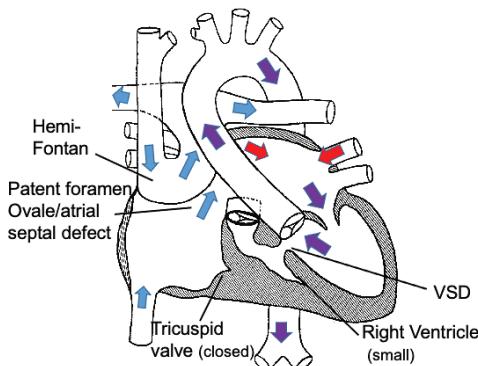


Figure 23: Stage 2 – hemi-Fontan

## Stage 3 procedure

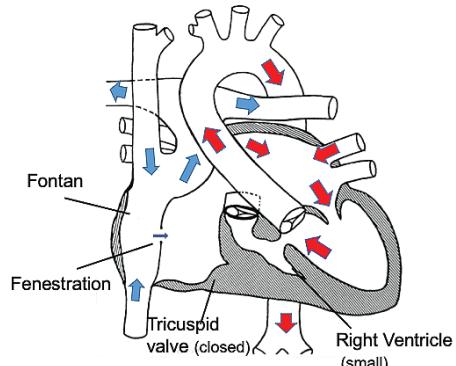


Figure 24: Stage 3 – Fontan

# HRHS: Tricuspid atresia with transposition of great vessels and VSD

## Stage 1 procedure

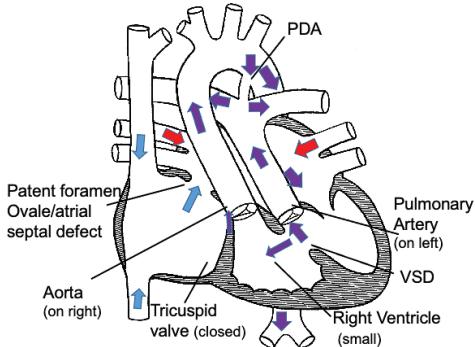


Figure 25: Tricuspid atresia with transposition of great vessels and VSD

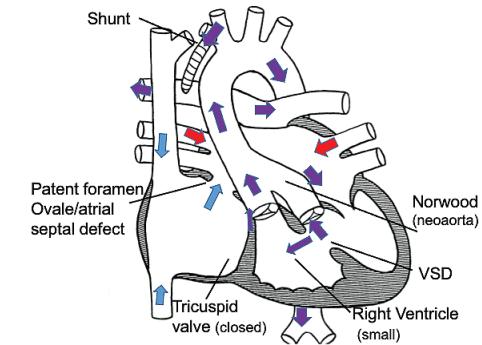


Figure 26: Stage 1 – modified Norwood with shunt

## Stage 2 procedure

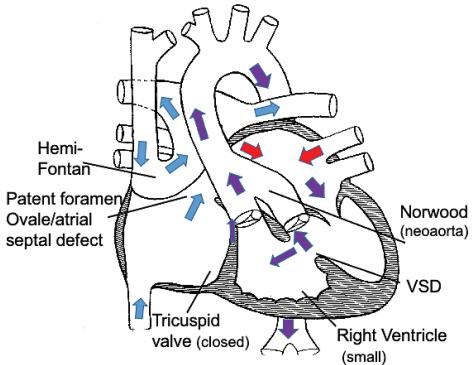


Figure 27: Stage 2 – hemi-Fontan

## Stage 3 procedure

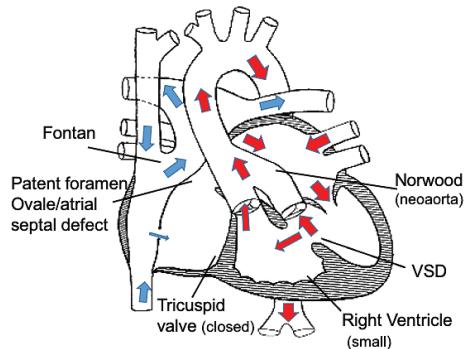


Figure 28: Stage 3 – Fontan (lateral tunnel)

## HRHS: Pulmonary atresia with intact ventricular septum

### Stage 1 procedure

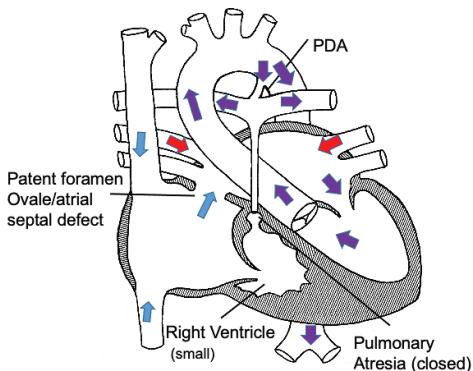


Figure 29: Pulmonary atresia with intact ventricular septum

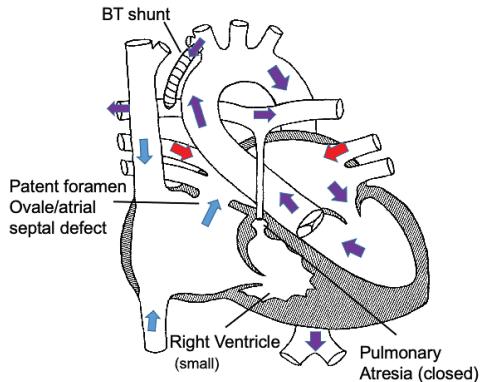


Figure 30: Stage 1 – Blalock Taussig shunt

### Stage 2 procedure

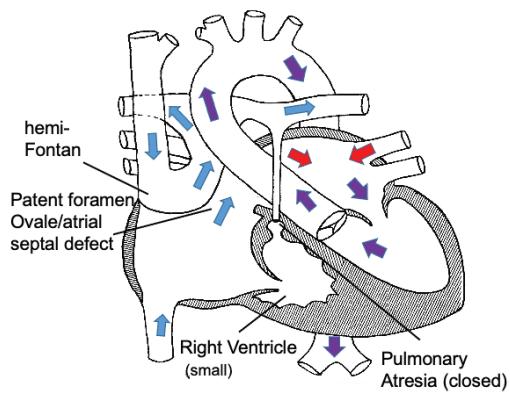


Figure 31: Stage 2 – hemi-Fontan

### Stage 3 procedure

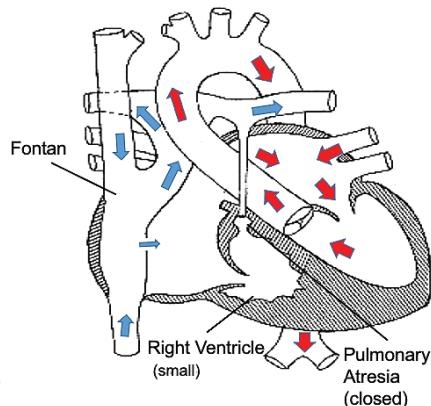
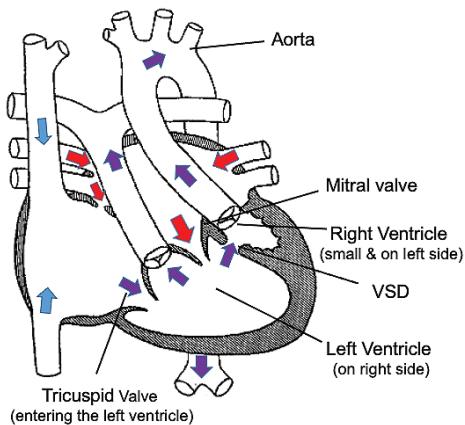
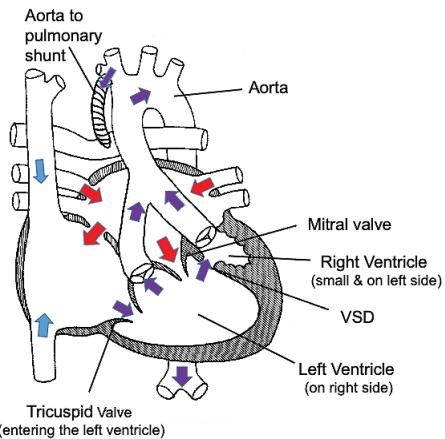


Figure 32: Stage 3 – Fontan (lateral tunnel)

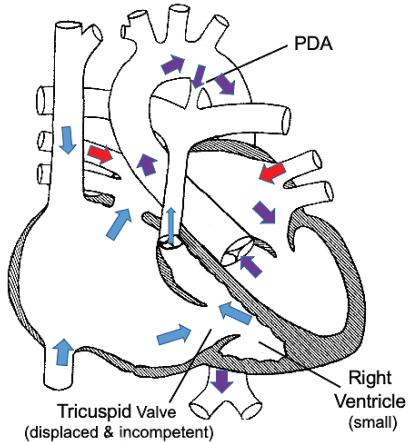
## Complex single ventricle diagnoses



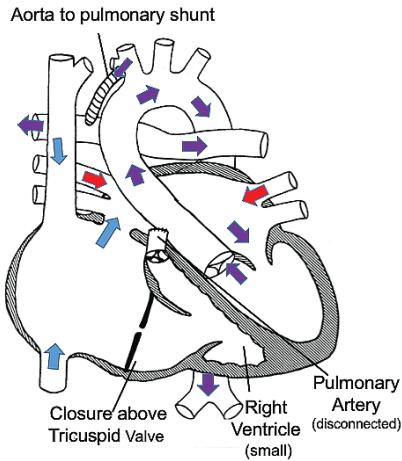
**Figure 33: Double inlet left ventricle with levo transposition of the great vessels**



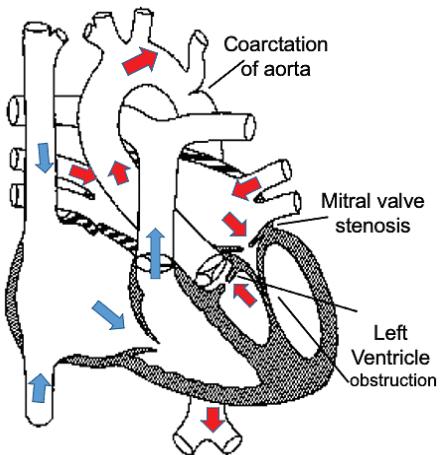
**Figure 34: Double inlet left ventricle — postoperative — Damus Kaye Stansel and shunt**



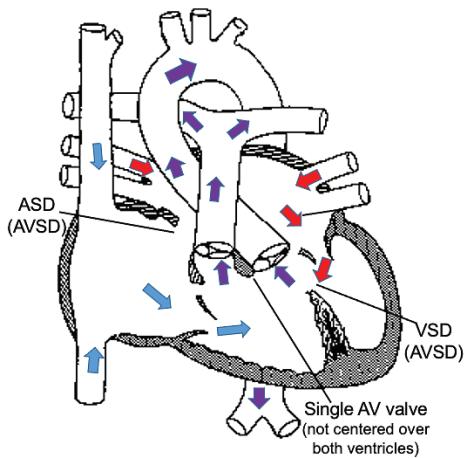
**Figure 35: Ebstein's anomaly**



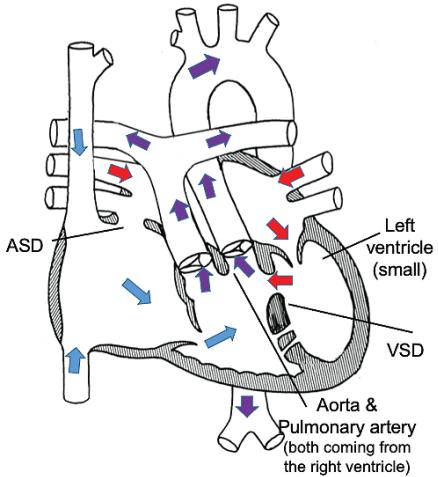
**Figure 36: Stage 1 – Starnes' procedure, aorta to pulmonary shunt**



**Figure 37: Shone's complex**

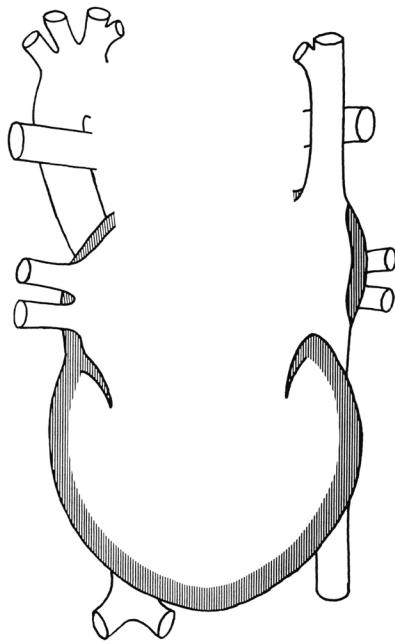
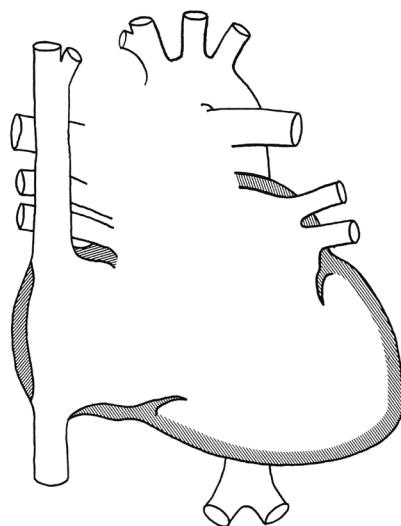


**Figure 38: Unbalanced atrioventricular septal defect (AVSD)**



**Figure 39: Double outlet right ventricle with hypoplastic left ventricle**

## Modifiable heart diagrams



## What to Expect After Surgery

After surgery, your child will be admitted to the pediatric cardiac intensive care unit. The length of time required to recover is different for every child and their unique heart defect. Seeing your child critically ill in the ICU is extremely difficult and stressful. Our heart care team including doctors, nurses, social workers, and many others will be there to support you and provide regular updates on how your child is doing.

Since you know your child best, you can help us take the best care of your child. Throughout the post-procedure time we want you to tell us anything you notice and to ask questions. We always want open communication with you. Having the heart care team and you working together is always helpful to your child.

There are events that happen after heart surgery that we deal with routinely. These include:

- post-surgery pain.
- bleeding in the area of the procedure.
- changes in heart rhythm (EKG).
- changes in oxygen saturations or blood pressure.
- breathing or airway difficulties.
- feeding difficulties.

Although complications do occur, life-threatening complications are rare. Our team is always prepared to address them and will keep you informed about them.

**Pain:** As one would expect, some pain will be present after a surgery or procedure. Pain is always addressed by the heart care team and medications are available. You can help by letting us know the things you think can help your child feel more comfortable.

**Bleeding:** There is always a degree of bleeding at the site of a procedure with most any type of surgery or procedure. When surgery involves opening the chest, it is routine to place chest tubes to drain

any blood or fluid from the chest that might otherwise take space that is needed by the heart and lungs. When the heart lung machine (bypass pump) is used for open-heart surgery, there is typically more bleeding. Most blood loss slows down within the first few hours of surgery. If the bleeding continues, the surgeon may need to explore in the area to see where it is coming from and work to stop it. This is typically done in the intensive care unit without going back to the operating room.

**Heart/circulation complications:** Low oxygen saturations can occur if there is not enough blood flow to the lungs through the pulmonary arteries (that flow may be through a surgically created shunt). An echocardiogram and/or cardiac catheterization may be done to look for the cause and guide treatment.

After surgery, there are times that the heart's pumping function is depressed (weaker) and medication is used to improve heart function. If the heart function is too weak or cardiac arrest occurs, our heart care team is prepared to use more extreme measures to try to support your child. One example of a life-saving technique is **extracorporeal membrane oxygenation (ECMO)**. This is a type of heart and lung machine that can provide blood flow and oxygenation when needed to allow the child to recover.

**Heart rhythm abnormalities (arrhythmias):** Irregular heartbeats or abnormal heart rhythms can happen after some heart surgeries. These usually resolve within days to weeks after surgery. Occasionally, they may remain even at discharge. If the abnormal rhythm continues and/or causes problems with blood pressure, medications may be used to control the rhythm. Rarely, a pacemaker may be needed.

**Infections:** Infections are uncommon but can occur after surgery. We take preventative measures to reduce the risk of infection. These preventions include antibiotic ointment placed inside both sides of your child's nose for five days before or immediately after surgery and giving antibiotics for 24 hours after surgery. Our heart care

teams carefully track all blood, urine, surgical site, and respiratory tract or lung infections and treat them right away.

**Breathing problems:** Anesthesia and chest surgery affects the lungs. The airway can have swelling from the **endotracheal tube** (the breathing tube that is used during and often after a procedure). The small air sacs in the lungs may be a bit collapsed after surgery. Encouraging deep breaths and clearing excess mucous from airways with coughing or suctioning are ways to help the lungs recover. We can support breathing using an endotracheal tube, a **ventilator** (breathing machine), and oxygen as long as necessary. At the end of some procedures the endotracheal tube and ventilator are no longer needed and for many others they are only needed for a short time after the procedure. Airway or lung issues such as a pneumonia or congestion can occur and require medications to treat.

**Feeding/nutrition/gastrointestinal challenges:** Surgery and anesthesia can upset your child's digestion. Loss of appetite, nausea and vomiting can occur with any child. For infants there are several things that may cause them not to eat. A baby must coordinate sucking, swallowing, and breathing all at the same time in order to eat. This can be tough for a baby with CHD after surgery. Some things that can make it tougher for a baby to eat after surgery are:

- If a newborn has not had a chance to feed before surgery, they may need time to learn to eat.
- Less commonly, there may be an injury or partial injury during surgery to the nerves associated with the vocal cords which help to protect the airway when swallowing.

We have a specialized feeding and swallowing team, including occupational and speech therapy and ear/nose/throat specialists, who evaluate your baby and work with you and your baby to help develop feeding skills. If it isn't possible for your child to eat enough by mouth, a tube is used to provide breast milk or formula into the stomach. The tube inserted in the nose and then into the stomach is called a **nasogastric (NG) tube**. Over time it is hoped more of the feeding can be by mouth and the amount going through the tube is

decreased. If tube feedings are needed to help your child at the time of discharge, you will be taught all you need to know to do the tube feedings at home.

For children who continue to need this type of feeding support, a procedure can be done to place a tube directly into the stomach (a **gastrostomy or G-tube**) to support them for longer until they gain feeding skills to have all feedings by mouth. Your heart care team will provide guidance in these cases.

There are instances when a child may develop bloody stools. Following complex surgeries (particularly after stage 1 surgeries) blood in the stool can be related to intermittent decreased blood flow in the intestine. This can make your child very sick and will require additional treatment.

**Neurological complications:** Rarely, seizures and/or a stroke can occur after heart surgery. If your child undergoes a complex procedure or has had multiple procedures, there is a higher risk for these complications. To diagnose these problems as quickly as possible and prevent further problems, your child may be connected to a brainwave monitor (**EEG**). A pediatric neurologist will be consulted to assist with diagnosis and treatment if these complications occur. Seizures are treated with medicine. Other tests may be necessary including head ultrasound, MRI, or CT scan to evaluate and consider treatment options.

# Planning for Discharge

## After stage 1 surgery

Before your child is discharged after the first surgery for single ventricle heart defects, you will be given a lot of information about how to care for your child. There are teaching videos and written material. You will practice:

- Giving medications.
- Feeding with a feeding tube, if needed.
- Weighing your child.
- Checking your child's oxygen level.

To prepare for being at home, you will spend at least a full day “rooming in” with your child. During this time, you will take care of your child completely (like care will be at home), but there is the assurance of knowing the heart care team is available to answer questions or concerns.

In addition, before discharge there will be a phone call with you, the hospital care team, the pediatrician/primary care provider, and the heart doctor (cardiologist) to review all of your child's care issues. Follow up appointments will be scheduled.

Discharge from the hospital will not happen until all of this has been done and your child has been gaining weight over several days.

Once discharged home, your child will need close monitoring and follow-up. It is important to follow-up regularly with your child's pediatrician, cardiologist and interstage nurse practitioner. We will focus on feedings, your child's ability to gain weight, and medications. Common childhood illnesses may be tougher for a child with heart disease to handle.

## **Interstage monitoring program (between stage 1 and stage 2 surgeries)**

To help support care at home during the interstage period, a specialized home monitoring team follows your child with a single ventricle heart defect to monitor their growth closely and guide you in how to watch for symptoms that could require further evaluation or care. Home monitoring begins the day you and your child go home from the hospital and continues until the second stage operation.

The home monitoring team includes a pediatric nurse practitioner who specializes in cardiology and a dietitian. A **nurse practitioner** is a registered nurse who has additional education and training in how to diagnose and treat disease. A **dietitian** may provide nutritional counseling, meal planning, medical nutrition therapy (such as a special diet, dietary supplements, or intravenous or tube feedings), and nutrition education programs.

Before discharge, you will receive:

- A scale to weigh your child at home.
- A monitor to check your child's oxygen saturation.

Your child will need to be weighed and have their oxygen saturation checked every day. To check the blood oxygen saturation (also called “pulse ox” or “sat”) you will wrap a small piece of tape around the baby’s finger, wrist, or toe. You will be instructed to record the weight, oxygen saturation, and amount your child eats in a written log or on your child’s portal online. The interstage team will review all this information and communicate with you, your child’s cardiologist, and pediatrician weekly (or more often if necessary). The team is available whenever there are questions or concerns. Since we have started home monitoring, we have seen improved growth in babies and have been able to recognize and treat potential problems earlier.

## **After stage 2 surgery**

After the second surgery (the Hemi-Fontan or bidirectional Glenn), your child may look bluer initially and may be fussy or irritable after waking up from surgery. This is normal and will typically improve in the first few weeks after surgery. If the oxygen levels are much lower than expected right after surgery, we may need to evaluate your child further. This may include an echocardiogram and possibly a heart catheterization to identify if there is a problem with the blood flow to the lungs.

After the second surgery, your child will have a more stable lung blood flow and their heart will have to do less work overall. Parents frequently say that their babies are happier and eat better after recovery from this surgery. It is no longer necessary to do daily weights or oxygen checks at home after the second stage of surgery. Visits to the cardiologist are typically less often, but still occur every 3-6 months.

## **After stage 3 surgery**

This surgery may have a hospital stay of 2 weeks or possibly longer. Children usually do very well with this surgery and are transferred from the ICU to the acute care unit within 1-2 days of surgery. Most of the hospital stay is because of chest tube drainage that can take time to go away. Your child can be active and spend a lot of the day in the playroom or walking around the hospital. The oxygen levels after this surgery are usually in the upper 80s or low 90s but may rise to the upper 90s over the months after a Fontan surgery.

After the Fontan surgery, visits to the cardiologist occur every 6-12 months.

## Choosing a Heart Center for Single Ventricle Heart Care

Consultation with a congenital heart center is an important way for you to learn about the diagnosis and options for care. No matter where you end up receiving care for your child, the most important thing is to gather information and ask questions. Of course, this is much easier to do if your situation is not an emergency, but in any case, asking questions will help you understand the information better and feel more comfortable with your heart care team and the decisions you and the team will make together.

Most often, the more experience a heart care team has, the better patients recover. Excellent surgical outcomes are important. The best results are dependent on the entire heart care team. That includes surgeons, cardiologists, anesthesiologists, nurses, pharmacists, social workers, respiratory therapists, and many more. If there are any complications or surprise findings, it is important that the heart care team is prepared to provide the intensive care necessary, preferably in a cardiac intensive care unit where everyone is specialized in congenital heart care.

Listed below are some suggestions on where you can go for information on your congenital heart disease diagnosis and what you can do to make sure you are choosing the right heart center for your needs.

- **Check online for heart education and resources:**
  - Congenital Heart Defects (Centers for Disease Control and Prevention): [www.cdc.gov/ncbddd/heartdefects/living.html](http://www.cdc.gov/ncbddd/heartdefects/living.html)
  - Congenital Heart Defects (National Heart, Lung and Blood Institute): [www.nhlbi.nih.gov/health-topics/congenital-heart-defects](http://www.nhlbi.nih.gov/health-topics/congenital-heart-defects)
  - Congenital Heart Center Overview (C.S. Mott Children's Hospital): [www.mottchildren.org/congenital](http://www.mottchildren.org/congenital)
  - Conquering CHD Hospital Navigator (provides information on various heart centers): [www.conqueringchd.org/learn/hospital-navigator/](http://www.conqueringchd.org/learn/hospital-navigator/)

- **Review more information (data) on heart centers:** Congenital Heart Surgery Public Reporting (Society of Thoracic Surgeons) has surgical outcome data for participating heart centers: [publicreporting.sts.org/chsd](http://publicreporting.sts.org/chsd). Enter the name of a hospital or select your state from the list and click “apply” to access the information. This can be hard to understand on your own.  
**It is very important to go over this information with your pediatrician or heart care doctor to help you understand what it means.**
- **Ask questions:** The Conquering CHD group has a good list of questions families can ask any heart center: [www.conqueringchd.org/guided-questions-tool/](http://www.conqueringchd.org/guided-questions-tool/)
  - At a minimum, you can ask a doctor:
    - How many times they have done the procedure?
    - How many times their team has done the procedure?
    - How long they expect a child to be in the hospital?
    - What may be expected during the hospital stay?
    - Is there more than one surgeon and a complete team to give the care needed?
- **Social media and online sites:** These can be helpful, but it is very important to check any site to be sure it is a responsible site. It's important to remember that every patient's heart defect and heart journey is unique; your child's situation may be very different from those discussed on social media. For more information, see the section on Family Support.
- **Ask other families and those you trust:** You (and your child, if old enough) must feel they are making the right decision for their situation.
- **Ask for another opinion from another heart center:** This is something that heart care doctors are very willing to do to help make decisions especially for more complicated patients. If a doctor is hesitant to answer your questions or does not support

you in getting a second opinion, that may be a reason to seek another program for your child's care.

A family may decide to travel to a center far from their home to get care at an experienced center of their choice. There are resources to help you in this process. Social workers are an excellent resource for help with this and other needs. To contact the social work team at the Congenital Heart Center, please call the clinic at 734-764-5176.

This can be a stressful time of information gathering and discussions with doctors and your family. We encourage you to use all resources to meet your needs for information and support including doctors, nurse practitioners, nurses, social workers, clergy, family doctors, close friends or family members, and other parents of children with single ventricle heart defects.

## Lifelong Considerations

The overall goal of caring for single ventricle heart patients is to help them live life as normally as possible. Many things may be necessary to achieve this goal, such as:

- Tube feedings.
- Hospital stays.
- Medications.
- Procedures.
- Appointments.

Maximizing quality of life is always the focus for your child. Children with single ventricle hearts, regardless of which surgeries are done, will need to be followed by a pediatric cardiologist or adult congenital heart cardiologist for the rest of their lives. How often appointments are necessary will be determined by the doctors. Things to discuss with your heart care doctor as your child gets older include:

- Exercise and physical activity recommendations.
- Prevention of endocarditis (an infection in the heart).
- Future surgeries.
- Role of heart transplant.
- Peer support groups.
- Birth control and pregnancy.
- Career choice.
- Insurance issues.

The care team will help you and your child with these issues as they arise over your child's lifespan. The next section will discuss some of these topics.

### Exercise and play

While there may be some limitations for exercise, normal play is typically fine. As your child grows, the heart care team will help establish guidelines for exercise and activities. Testing is typically

done in older children to assess how well their heart tolerates exercise. Exercise is important to a healthy heart and is also helpful to the single ventricle circulation. Therefore, it is good to discuss how to safely exercise with the heart care team.

## **Immunizations and vaccines**

Infants, children and adults with single ventricle hearts should receive all recommended immunizations (including flu shots). Consult with the heart care team when needed, as there may be occasions when the timing needs to be changed due to surgeries, procedures, childhood illnesses or other events.

For babies and toddlers under one year of age who haven't completed all three surgeries, Synagis (palivizumab) is recommended. This is a monthly injection to help them fight respiratory syncytial virus (RSV), which can make babies with single ventricle hearts very ill. These injections are required from October or November through March or April depending on when the virus is present in the community. Babies receiving Synagis still require flu shots.

## **Developmental considerations**

Some children with single ventricle hearts have developmental challenges that may affect a wide range of functions including muscle control, speech, emotion, learning ability, and memory. Having your child evaluated early can be helpful to check for differences and get recommendations to help your child reach their potential. You may be referred to a specialist to help with your child's development.

Routine visits to the Cardiology Neurodevelopmental Follow-Up Clinic are recommended at certain ages to assess your child and recommend appropriate therapies so they can start as early as possible when needed. For more information about this clinic, visit [www.mottchildren.org/chcneurodev](http://www.mottchildren.org/chcneurodev).

## **Routine post-Fontan follow-up**

Although Fontan surgery allows children with single ventricles to continue to grow and develop, the circulation of blood following a Fontan procedure is not normal and can cause some serious changes in the body as your child grows. Routine testing of the liver, kidney, immune, and endocrine systems in addition to heart function is needed from childhood through adulthood. Some children, particularly those followed outside our center, may benefit from being checked at our multi-disciplinary Single Ventricle Clinic. At this clinic, patients are evaluated by multiple specialists, in addition to their routine primary cardiologists' care. For more information about this clinic, visit [www.mottchildren.org/chc-sv-clinic](http://www.mottchildren.org/chc-sv-clinic).

## **Endocarditis prevention**

Endocarditis is a serious heart infection caused by bacteria in the blood stream. People who have major heart defects or have had heart surgery have a higher risk of developing endocarditis. Prevention is key! Doctors or dentists will prescribe antibiotics before teeth cleaning and some other procedures to help prevent endocarditis. Be sure all health care providers for your child are informed of their heart history. Symptoms of endocarditis are vague, but if there is a fever that will not go away after at least 3 days or if your child is losing weight make sure that the doctor treating your child knows they have a heart defect. Blood tests and blood cultures to check for infection are often needed. Good dental care and brushing teeth at least once a day can help prevent this infection.

## **Adult congenital heart disease care**

Single ventricle heart disease is a lifelong condition. Fortunately, there are specialists in cardiology for adults with congenital heart disease. They can provide advice as children grow into adults and need to consider adult decisions like family planning, employment, and aging. They also monitor heart changes over time as other treatments may become necessary. Care with an adult congenital heart team is necessary throughout adult life. For more about this program, visit [www.umcvc.org/achd](http://www.umcvc.org/achd).

# Family Support

While this booklet is a very important educational resource, there are many other types of resources and support available to you and your family. Understanding the social and emotional stress that often go along with single ventricle heart disease and knowing where to get support is important.

## Family care resources

You must care for yourselves to be able to care for your children. The reality of having a child diagnosed with a heart defect, experiencing extended hospital stays, and having a sense of uncertainty can feel very overwhelming. Lack of sleep can affect your ability to cope and think clearly. Some families report having symptoms of post-traumatic stress disorder. It is important to practice self-care including:

- Good nutrition.
- Getting enough sleep.
- Mindfulness.
- Positive thoughts.

The heart care team is available to support you and your family. It is important to reach out to them and ask for help for physical and emotional needs. Please remember you are not alone.

The heart care team works with several departments and groups to help provide a full system of support for families. A few of these support systems include:

- **Social work:** Provides support through logistical planning, emotional coping, resource identification and much more.
- **Child life:** Provides support through medical play, art therapy, music therapy and other activities.
- **Spiritual care:** Provides support from clinically-trained chaplains representing a variety of religions and faith traditions. Spiritual care is available even for those who don't participate in organized religion.

- **Palliative care:** Provides special guidance in identifying quality of life concerns, long- and short-term goals and help with difficult decisions. This support can be available at any point of serious illness.
- **Connecting with other families:** Provides support through helpful relationships that may occur with families you meet through your/your child's journey. You may also develop these relationships through online groups, and with hospital volunteer and support programs.
- **Pediatric psychologists:** Provide patient and family support for coping with the emotional side of your child's CHD.

## Online patient/family resources

The internet and social media can be helpful and often provide very useful information. However, there can be confusing or misleading information and comments. It is very important to check any website to be sure it comes from a responsible source. **Find out if the site has a medical advisory board, or connection to a pediatric heart center or other reputable national organization.**

Comments on social media or about online articles may be well-intentioned but may not be correct for all families and children. We recommend you always clarify recommendations with your own health care team who knows your situation best. For general information, use websites that you know are reputable with good sources.

Below are some websites and social media information.

*Please note that all online resource sites are subject to change over time. Some of the organizations are on social media platforms, also.*

## National and local websites

- Congenital Heart Center at University of Michigan Health C.S. Mott Children's Hospital: [www.mottchildren.org/congenital](http://www.mottchildren.org/congenital)
- Adult Congenital Heart Association: [www.achaheart.org](http://www.achaheart.org)
- ACTION Network (Advanced Cardiac Therapies Improving Outcomes Network): [www.actionlearningnetwork.org](http://www.actionlearningnetwork.org)

- American Heart Association: [www.americanheart.org](http://www.americanheart.org)
- Cardiac Networks United: [cardiacnetworksunited.org](http://cardiacnetworksunited.org)
- Children's Heart Foundation: [www.childrensheartfoundation.org](http://www.childrensheartfoundation.org)  
Local Michigan Chapter: [www.childrensheartfoundation.org/michigan](http://www.childrensheartfoundation.org/michigan). Use the search bar to search for events happening in your state's chapter.
- Conquering CHD: [conqueringchd.org](http://conqueringchd.org)  
Local Michigan Chapter: [www.conqueringchd.org/michigan](http://www.conqueringchd.org/michigan)
- Cove Point Foundation – Congenital Heart Disease (includes diagrams) – [www.pted.org](http://www.pted.org)
- Enduring Hearts (serving transplant families): [enduringhearts.org](http://enduringhearts.org)
- Fontan Outcomes Network: [www.sistersbyheart.org/fontan-outcomes-network](http://www.sistersbyheart.org/fontan-outcomes-network)
- iHeartChange (designed to help young people who are “transitioning” from pediatric to adult care): [iheartchange.org](http://iheartchange.org)
- Mended Hearts and Mended Little Hearts: [www.mendedhearts.org](http://www.mendedhearts.org)
- NPC-QIC (National Pediatric Cardiac-Quality Improvement Collaborative): [www.npcqic.org](http://www.npcqic.org)

This site includes a guide created by NPC-QIC Fetal Learning Lab with contributions from Sisters by Heart – “The Single Ventricle Journey: A Guide for Parents and Families”: [www.npcqic.org/tools-resources](http://www.npcqic.org/tools-resources)

- Saving Tiny Hearts: [savingtinyhearts.org](http://savingtinyhearts.org)
- Sisters by Heart (HLHS): [www.sistersbyheart.org](http://www.sistersbyheart.org)

## Social media

Local and national social media sites are available. Some are hosted by organizations and others are managed by family volunteers. Other than the specific C.S. Mott Children's Hospital Facebook and Twitter pages, these sites are not managed by C.S. Mott Children's Hospital.

Sites include:

- C.S. Mott Children's Hospital:  
[www.facebook.com/mottchildren](http://www.facebook.com/mottchildren)  
[www.twitter.com/mottchildren](http://www.twitter.com/mottchildren)
- Action Learning Network (focused on heart failure and transplant): [www.facebook.com/actionlearningnetwork](http://www.facebook.com/actionlearningnetwork)
- Children's Heart Foundation Michigan: [www.facebook.com/CHFMI](http://www.facebook.com/CHFMI)  
Children's Heart Foundation (national organization): [www.facebook.com/TheCHF](http://www.facebook.com/TheCHF)
- Conquering CHD – Michigan: [www.facebook.com/groups/ConqueringCHDMichigan](http://www.facebook.com/groups/ConqueringCHDMichigan)  
Conquering CHD (national organization): [www.facebook.com/conqueringchd](http://www.facebook.com/conqueringchd)
- Hearts of Hope of SE Michigan: [www.facebook.com/groups/113575315332](http://www.facebook.com/groups/113575315332)
- Heart Families from Mott: [www.facebook.com/groups/432331583522633](http://www.facebook.com/groups/432331583522633)
- Heart Families of Western Michigan: [www.facebook.com/groups/4021250814615971](http://www.facebook.com/groups/4021250814615971)
- Mended Little Hearts:  
[www.facebook.com/MendedLittleHeartsNationalOrganization](http://www.facebook.com/MendedLittleHeartsNationalOrganization)  
[www.twitter.com/MLH\\_CHD](http://www.twitter.com/MLH_CHD)
- Michigan Heart Families: [www.facebook.com/groups/198643373492259](http://www.facebook.com/groups/198643373492259)
- Sisters By Heart (a group focused on HLHS): [www.facebook.com/Sisters-by-Heart-156971814350396](http://www.facebook.com/Sisters-by-Heart-156971814350396)

## Conclusion

Advances in congenital heart care have improved the survival rate and quality of life for children with even the most complex CHDs. As children are better able to survive with single ventricle heart defects, we can expand the focus of care to improving quality of life through focus on developmental outcomes, recognizing and preventing long-term complications of single ventricle hearts, and enhancing the transition to become independent adults.

We highly value the thoughts and ideas of patients and families who have lived with a single ventricle heart defect. They have experienced what it means to:

- Survive a major diagnosis.
- Grow up with the physical challenges of a single ventricle heart.
- Struggle with the choices and concerns of adolescence and young adulthood in the face of the single ventricle diagnosis.
- Face the realities of how the diagnosis impacts relationships with parents, siblings, significant others/spouses, family members and peers.

We know those born with single ventricle heart defects can have a good quality of life and will have friends, be able to play, go to school, and grow up like other children. Our hope for all our patients is for them to live a happy life well into adulthood. The journey may be different for every child and family, but the support and guidance that we will provide as members of your heart care team will be constant.

## References

1. C.S. Mott Children's Hospital – Congenital Heart Disease: [www.mottchildren.org/congenital](http://www.mottchildren.org/congenital)
2. Centers for Disease Control – Congenital Heart Defects: [www.cdc.gov/ncbddd/heartdefects/specificdefects.html](http://www.cdc.gov/ncbddd/heartdefects/specificdefects.html)
3. Hoffman JE, Kaplan S. The incidence of congenital heart disease, *JACC*, Volume 39, Issue 12, 19 June 2002, Pages 1890-1900
4. Hypoplastic Left Heart Syndrome / HLHS | Cincinnati Fetal Center: [www.cincinnatichildrens.org/service/f/fetal-care/conditions/hlhs](http://www.cincinnatichildrens.org/service/f/fetal-care/conditions/hlhs) (source of some incidence data in the intro)
5. Mullins CS, Mayer DC. *Congenital Heart Disease: A Diagrammatic Atlas*. Wiley Liss. 1991. *Original drawings adapted from Congenital Heart Disease: A Diagrammatic Atlas by C.S. Mullins, and D.C. Mayer. Used with permission.*
6. The Single Ventricle Journey: A Guide for Parents and Families created by NPC-QIC Fetal Learning Lab with contributions from Sisters by Heart: [www.npcqic.org/tools-resources](http://www.npcqic.org/tools-resources)

Additional links/references are in the “choosing a heart center for single ventricle heart care” and “online patient/family resources” sections.

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