



# The Single Ventricle Journey:

## A Guide for Parents and Families

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### CREATED BY

**National Pediatric Cardiology Quality Improvement Collaborative  
Fetal Learning Lab** *with contributions from Sisters by Heart*

### EDITORS

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*Thank you to the many families who contributed photos and stories.*



National Pediatric Cardiology  
Quality Improvement Collaborative



sistersbyheart



CHILDREN'S  
HEART ASSOCIATION  
OF CINCINNATI



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*This guide is not intended to be a substitute for professional medical advice, diagnosis, or treatment. Always seek the advice of a physician or other qualified healthcare provider with any questions you may have regarding a medical condition. In the event of a medical emergency, call a doctor or 911 immediately.*

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## Dear Families,

The National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC) is a learning network that collaborates with our parent partner organization, Sisters by Heart, to engage patients, families, clinicians, and researchers and use data for quality improvement and research. Currently made up of over 67 care centers in the US and Canada, the overarching goal of NPC-QIC is to dramatically improve the outcomes of care for children with congenital heart disease.

Our initial phase, launched in 2008, focused specifically on improving the survival and quality of life of infants with a single ventricle heart during the “interstage” period following a Norwood operation (between their first and second heart surgeries). Our efforts led to significant improvements in interstage growth and survival. Our second phase launched in 2016 and broadened the scope of our work together, focusing on the care and outcomes of these infants from prenatal diagnosis through the child’s first year of life.

When you learn your baby has Hypoplastic Left Heart Syndrome (HLHS) or another single ventricle heart problem, you may experience a range of emotions from shock to sadness. You may also feel overwhelmed by the amount of information you are being asked to comprehend and remember. We know that this is an extremely stressful and emotional time for you.

As a quality improvement initiative, the NPC-QIC Fetal Learning Lab clinicians and parents created this book to help inform you as you prepare for the journey ahead. Please read it, share it with others and keep it with you throughout your baby’s first hospitalization and onward. We hope that by arming you with this amazing resource, you will feel informed, empowered and therefore comforted as you begin parenting a baby with a single ventricle heart problem.

Every child is different, and your journey will be filled with its own unique challenges and celebrations. We want you to know that while you are focused on caring for your baby, we remain committed to continuing our

---

efforts to improve outcomes for your baby and all children with single ventricle heart problems.

Please contact us (*see below*) if you have any questions.

SINCERELY,

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*Support for NPC-QIC is provided by participating care centers, the Children's Heart Association of Cincinnati, and individual philanthropy.*

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### MORE INFORMATION

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# 1

## Diagnosis

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

## Hypoplastic Left Heart Syndrome

Single ventricle **congenital heart disease (CHD)** is the term used to describe abnormalities in the formation of the heart when one side of the heart does not form normally. These are serious conditions that require life-long care. This guide focuses on **hypoplastic left heart syndrome (HLHS)** and its different forms, in which the left side of the heart is severely underdeveloped. HLHS is a rare condition, affecting about 1 in 5,000 babies. In the United States, about 1,000 babies with HLHS are born each year.

This section describes how a heart is normally put together and how a heart with HLHS is different. A normal heart has four chambers: two upper filling chambers, called the left and right **atria**, and two lower pumping chambers, called the left and right **ventricles** (see *Figure 1A*). The left side has the important and harder job of pumping **oxygenated** blood, which has a **red** color, to the body. The body uses the oxygen in the blood, and after the oxygen is removed from the blood, it turns **blue** in color. The job of the right side of the

Figure 1A – Normal Heart

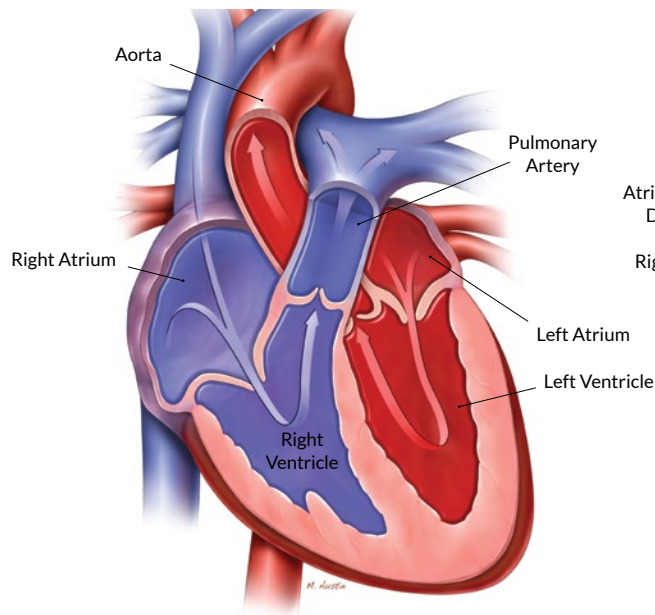
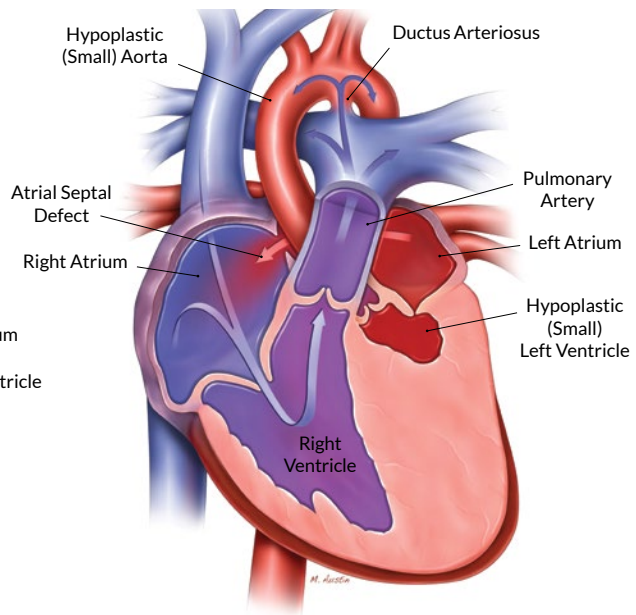


Figure 1B – Hypoplastic Left Heart Syndrome (HLHS)



heart is to send **blue** blood to the lungs so that the blood can pick up oxygen again. After the blood leaves the lungs, it returns to the left side of the heart, and the cycle repeats itself over and over again.

In HLHS, the left side of the heart is very underdeveloped (*see Figure 1B*). The word **hypoplastic** means small. In HLHS, the structures on the left side of the heart are so small that they are unable to do the job of pumping **red** blood to the body. The inflow valve (called the **mitral valve**) and the outflow valve (**aortic valve**) might be narrowed, which is called **stenosis**, or completely blocked, which is called **atresia**. In addition, the left-sided pumping chamber (ventricle) itself is very small. The result is that only the structures on the right side of the heart have formed normally, including the inflow valve (**tricuspid valve**), the outflow valve (**pulmonary valve**), and the right ventricle. Many babies with single ventricle CHD also have an **atrial septal defect (ASD)**, which is a hole (connection) between the two atria. This connection is used by the surgeons to help with changing the blood flow pattern as is described in the next chapter.

Of the two jobs that the heart has to do—pumping blood to the body and pumping blood to the lungs—pumping blood to the body is the harder job. In HLHS, three surgeries are required to re-direct the blood flow so that the ventricle that did form normally, the right ventricle, can do the important job of sending blood to the body. You will read more about these surgeries in upcoming sections.

## Causes

Families affected by a diagnosis of single ventricle CHD like HLHS often want to understand what caused it. HLHS happens for unknown reasons while the baby is in the mother's womb. Most babies with HLHS are otherwise healthy, but some may have other medical problems. About two-thirds of children with HLHS are boys.

For starters, nothing one or one's partner did caused this to happen: nothing that one ate or drank; no medications taken; and nothing that occurred during conception. Most forms of CHD, including HLHS and other types of single ventricle CHD, happen for no single particular reason.

So, what did cause this? The reason is not clear – it is thought that a few issues that might be seen in a person's **genetic** make-up and his or her **environment** may result in a heart defect. Researchers have looked at various **exposures** during pregnancy such as drugs, infection, chemicals, and known illnesses of the mother, but a true connection between these and single ventricle CHD has not been proven.

Some cases suggest that blockages to parts of the left side of the heart and blood vessels may run in families. These heart problems are known as *left-sided obstructive lesions*. This group includes minor issues of the left-sided heart valves (that may not require surgery), blockage in the great vessel called the *aorta*, and HLHS. Recent studies show that there is an increased chance of a sibling of someone with HLHS having a heart problem on the left side of the heart. This possibility increases if one of the parents also has a left-sided heart problem. There are many forms of CHD with differences in how bad the heart defect can be, and sometimes parents may not know that siblings (or even themselves) have heart problems unless they have an *ultrasound* of the heart.

## Genetic Abnormalities

Genetic syndromes result from a defect in a person's genes. **Genes** contain specific codes that are unique to every person. Each person's genes are contained within structures (**chromosomes**) in every cell. Single ventricle CHD such as HLHS is usually not related to *genetic syndromes* or **chromosomal defects** like Down syndrome. One exception is Turner syndrome, a rare genetic defect that occurs only in girls. Turner syndrome happens when one "X" chromosome is missing. Turner syndrome is known to be associated with different left-sided obstructive lesions, including HLHS. A heart abnormality occurs in approximately one-third of all infants with Turner syndrome, but only a small number will actually have HLHS. Testing for genetic abnormalities can be performed **prenatally** in some cases and also after birth.

Your doctor might recommend that you have **prenatal** testing to identify any genetic abnormalities before the birth of your baby. The following tests are currently available:

- **Non-invasive prenatal testing (NIPT) or Cell-free fetal DNA testing:** Your doctor can take a sample of your blood while you are pregnant to look for copies of **fetal** (baby) DNA. This is only a screening test. A positive result means that there could be problems with the DNA. This should be confirmed with another test to be sure of the result. Other testing can be done before or after birth, depending on the needs of the baby.
- **Amniocentesis:** This is an optional test that is done while you are pregnant to check for chromosomal defects like Down syndrome and also to check **lung maturity** in the baby. This invasive procedure involves taking a small amount of amniotic fluid with a thin, hollow needle with the guidance of ultrasound. The fluid is sent to the lab for testing (**karyotype** and/or **microarray**, see below) and the doctor will tell you when to expect results. There is a small risk of miscarriage with amniocentesis, so be sure to ask your **obstetrician (OB)** about risks and complications.

- **Karyotype or Microarray:** This test can be performed before birth using the amniocentesis sample or after birth by testing your baby's blood directly to look for genetic problems and to confirm the prenatal testing results. A karyotype confirms each cell has the right number and size of chromosomes (46) with no larger missing pieces or **duplications** of DNA. A microarray takes a more detailed look at the DNA for smaller **deletions** of genetic material.

## Prenatal Diagnosis

Pregnancy is an exciting time of anticipation and preparation, but fear can easily override that excitement with a diagnosis of CHD. Getting the news that your unborn baby has CHD is extremely hard, and parents will naturally have a lot of questions. Many families receive the diagnosis of HLHS before birth. This is called prenatal diagnosis, and we hope this provides families with time to better prepare themselves to welcome a baby with a special heart. From prenatal diagnosis to birth, expectant parents will have several opportunities to meet with their heart team and to be educated about this diagnosis to better prepare themselves for life with a child with HLHS.

A question often asked by parents is if there is a chance that the baby's heart defect will get better during pregnancy. Parents should know that heart defects like HLHS will not get better during pregnancy. A very small number of babies with HLHS may be considered for a procedure while still in the womb (called **fetal intervention**), which is done in some centers. This procedure may improve **outcomes** in some cases. Your cardiologist can determine if your baby might be able to have this type of procedure.

## Heart Testing During Pregnancy

The following is a description of some of the heart testing that may be done during pregnancy:

- **Fetal Echocardiography:** This is a test that uses ultrasound waves to look at how the baby's heart is formed and how it is working. The test is typically performed around 18 to 20 weeks of pregnancy. It is a painless test, and it is safe for both the mother and the baby. It is usually done by an ultrasound technician (sonographer), but it may be done by an OB, **maternal-fetal medicine specialist**, or **fetal cardiologist**. The test itself takes about 30 minutes. In most cases, a doctor will meet with you after the test to discuss the results in detail. This is a great time to ask questions. Usually, multiple **fetal echocardiograms** are performed throughout the course of the pregnancy to check on the baby's heart.

- **Non-stress test:** This is a fetal test that allows the OB to check the baby's heart rate and to see whether a baby's heart rate pattern is normal. This information helps the OB determine the well-being of the baby and if further testing is required, such as a follow-up ultrasound. This test is usually done at least once or twice after 26 weeks of pregnancy.

For patients with a prenatal diagnosis, the delivery is not usually scheduled before the 39th week of pregnancy. The decision about timing of delivery is usually made with your OB. The heart team will be prepared for your baby's delivery. Once the baby is born and stabilized by doctors called **neonatologists** in the delivery room, he or she will be transferred to a **neonatal intensive care unit (NICU)** where a whole team of healthcare providers will care for the baby. An echocardiogram will be performed on the baby to confirm the diagnosis, and additional preoperative testing will be performed (**electrocardiogram, or ECG**; x-ray; blood work; ultrasound of the head, kidneys). Additional testing such as **cardiac catheterization** or **cardiac computed tomography (CT)** or **magnetic resonance imaging (MRI)** may be done in some babies before having surgery.

## Postnatal Diagnosis

Many heart conditions can be diagnosed by ultrasound during pregnancy, but for a variety of reasons, not all heart defects are diagnosed before a baby is born. Some babies are diagnosed after they are born (**postnatal diagnosis**), when a caretaker is concerned that a baby may be unwell. Babies with unrecognized HLHS may appear to have difficulty breathing or eating, appear pale or gray, have decreased wet diapers, or be excessively sleepy or lethargic. If this happens, your providers will probably consult with a **pediatric cardiologist** to make the diagnosis and quickly arrange transfer to the appropriate level of care for your child.

When parents are expecting a healthy baby, receiving the diagnosis of HLHS after delivery can be shocking and terrifying. It is natural to worry and to be overwhelmed by suddenly being notified that the baby will need not one, but at least *three* open-heart surgeries, and by all of the information that has been shared. However, parents won't be alone in this journey as the whole team – including the cardiologists, surgeons, nurses, social workers, nutritionists, therapists, etc. – will be there to provide support.

# 2

## Surgical Procedures for Hypoplastic Left Heart Syndrome (HLHS)

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There are usually three surgeries that are required for babies who will undergo HLHS procedures:

- The Stage 1 Norwood surgery
- The Stage 2 Glenn surgery
- The Stage 3 Fontan surgery

In this chapter, we will briefly describe these procedures.

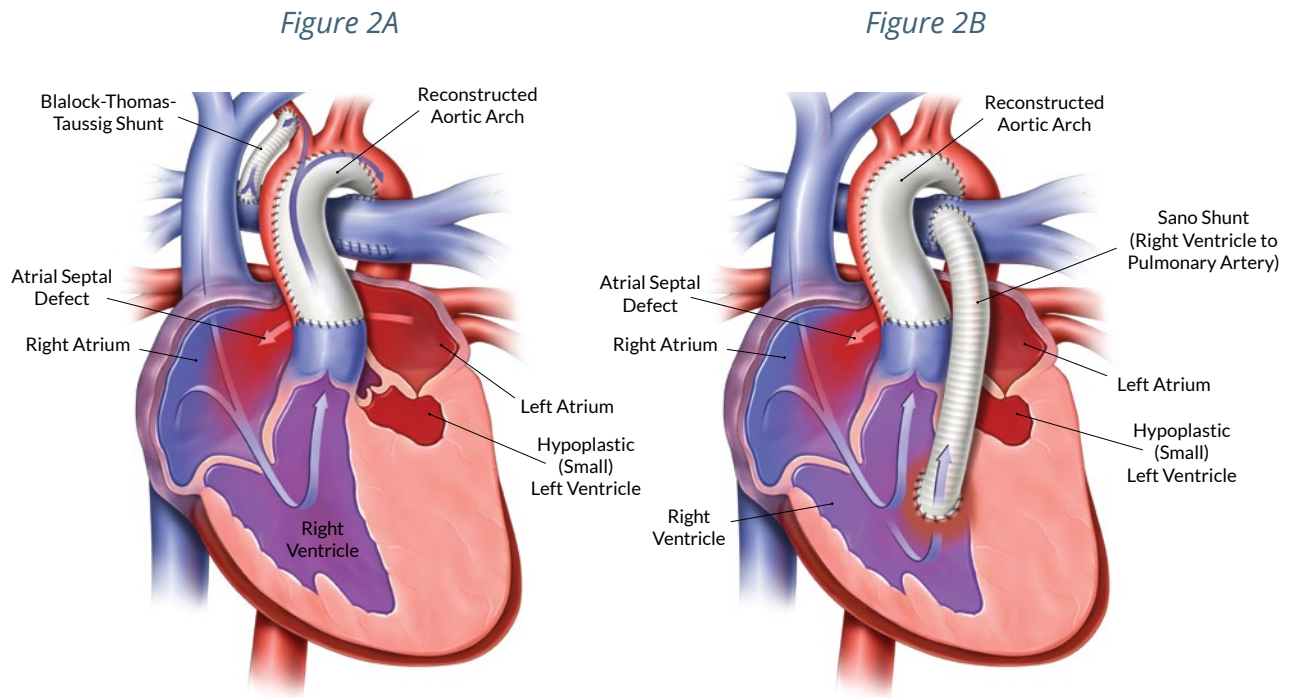
## Initial Surgery — Stage 1 Norwood (or Hybrid Norwood)

In HLHS, the left-sided pumping chamber (left ventricle) is very small and unable to eject enough blood to the body. This means that the right ventricle (the right-sided pumping chamber), which did form normally, becomes the main pump that pumps blood to both the body *and* the lungs. So, the right ventricle will now have two jobs. It will be able to pump blood (1) to the lungs through the main lung artery (main **pulmonary artery**) and (2) indirectly to the body across a connection called the **patent ductus arteriosus (PDA)**. The PDA is a normal blood vessel in the unborn baby. In a heart that has formed normally, the PDA usually closes in the first few days after a baby is born. In HLHS, however, the PDA is very important and needs to stay open to make sure that blood gets to the baby's brain and body. To keep the PDA open, a special medicine called **prostaglandin** (sometimes called prostin or PGE1) is given that allows the PDA to remain open until surgery. The goals of the Stage 1 surgery (either Norwood or hybrid Norwood) are to re-establish good blood flow to the body directly from the heart and to send enough blood to the lungs to keep oxygen levels high enough. This is what is called a "balanced **circulation**": just enough blood goes to the body, and just enough blood goes to the lungs.

The Norwood is usually done within the first week of life. Through this procedure, the surgeons provide blood flow to the body by attaching the main pulmonary artery to the underdeveloped aorta, making a new aorta, or *neo-aorta*, (see *Figure 2*) and creating a way for the red blood to travel to the body without any blockages. The PDA is no longer needed and will be closed by the surgeon. In addition, if a baby was born with an **atrial septal defect (ASD)**, that hole is enlarged by the surgeons to allow the red blood from



Figure 2 – Norwood Surgery



the lungs to pass freely to the right side. If a baby was born without an ASD, the surgeon will create one. Since the main lung artery is used to enlarge the aorta, blood flow to the lungs must be re-created. This can be accomplished in one of two ways: (1) a small connection from the neo-aorta to the arteries in the lungs, called a **Blalock-Thomas-Taussig (BTT) shunt** (see Figure 2A) or (2) a connection from the right ventricle to the arteries feeding the lungs, called a Sano shunt (see Figure 2B). Both operations have been very successful, and both are used at many different centers. Both surgeries require the use of a **heart-lung bypass machine** and the use of **blood transfusions**. Both surgeries will allow mixing of red and blue blood.

Some centers instead perform what is called the hybrid procedure. The hybrid procedure avoids the heart-lung bypass machine by performing part of the procedure using **cardiac catheterization**. The hybrid procedure includes placing a **stent** in the PDA, allowing blood flow to be sent to the body. If needed, a bigger hole (ASD) or connection is made between the top chambers of the heart using a **balloon catheter** or stent. The last step does involve surgery and includes placing bands around the artery to each lung (*left and right* pulmonary arteries) to protect the lungs from excess flow and pressure.

## ADELINE



Our HLHS heart warrior, right after her first surgery, the Norwood, and then once we arrived home one month later. She was so curious and in love with her mobile. Home, sweet home.

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The hybrid procedure is a way to create balanced blood flow without using the heart-lung bypass machine, and it can sometimes be performed without the need for blood transfusions.

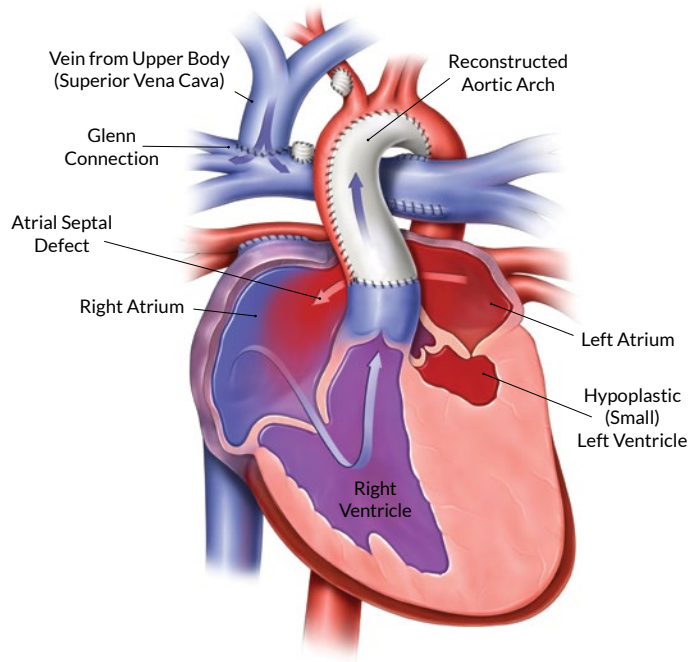
Some centers primarily use the hybrid procedure, whereas others primarily use the Norwood procedure. Be sure to ask your providers if you have questions about the type of surgery your doctors perform.

## Stage 2 Bidirectional Cavopulmonary Shunt (Glenn) and Stage 3 Fontan Completion

There are typically two more surgeries for the patient with HLHS: the Stage 2 cavopulmonary shunt, or **bidirectional Glenn** procedure, and the Stage 3 Fontan procedure. The purpose of these two surgeries is to re-route the **blue** blood so that it flows directly into the lungs without passing through the heart. These procedures separate the **blue** and **red** blood allowing normal or near-normal oxygen levels to reach the brain and body. Also, they make the circulation much more stable and decrease the amount of work the right ventricle has to do. The time period between the first and second surgeries is called the **Interstage**.

The Glenn is usually performed at 4 to 6 months of age, although it can be performed earlier or later in certain situations. The surgery involves attaching the **superior vena cava (SVC)**, the main vein that returns blood from the upper body, directly to the left and right pulmonary arteries (*see Figure 3*), re-routing **blue** blood from the top part of the

Figure 3 – Glenn Surgery

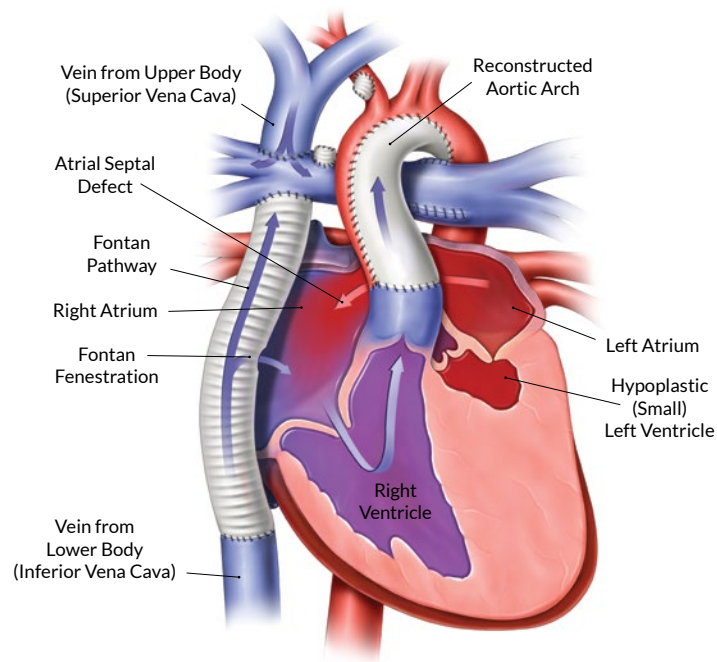


body so that it does not need to pass through the heart. Sometimes, there is more than one SVC (one on the right and one on the left). If there are SVCs on both sides of the body, both are usually surgically connected to the lungs ( **bilateral** bidirectional Glenn shunts). The previously placed BTT shunt or Sano shunt is removed, as it is no longer needed.

The **blue** blood from the lower body continues to return to the heart by way of the **inferior vena cava (IVC)**, mixing with the red blood from the Glenn shunt. Oxygen levels after the Glenn surgery are still below normal at 75% to 85%, similar to the Interstage between the Norwood and the Glenn; however, babies can grow and develop well after this procedure. For infants who had a hybrid Norwood procedure, the second surgery is called a **comprehensive** Stage 2 surgery, which includes the aortic arch re-creation (creating a neo-aorta) that was not performed at the time of the hybrid operation.

The Fontan surgery usually takes place when your child is 2 to 4 years old, although this varies somewhat from center to center. The timing of the surgery is dependent on how your child is doing, as well as center preference. The Fontan surgery involves directing the **blue** blood from the lower body to the pulmonary arteries, either through the

Figure 4 – Fontan Surgery



heart using a tube (*intracardiac baffle*) or along the outside of the heart (*extracardiac conduit*) (see Figure 4). A small connection to the heart may be created at this time (*fenestration*), which allows some blue blood to bypass the lungs; this can improve blood flow to the body immediately after surgery. This fenestration often will close on its own with time but can be closed in the cardiac catheterization lab later as well. Use of the heart-lung bypass machine is required for the Fontan completion procedure and is usually used for the Glenn procedure as well.

# 3

## Planning for Your Baby's Arrival

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

## Delivery Location—Where will I deliver my baby?

Your baby will require specialized care in the newborn period. Your OB team and fetal cardiologist will make recommendations about which centers are best suited to care for your baby after birth. In addition to the input from your current care team, NPC-QIC and Sisters by Heart created a document entitled [\*Single Ventricle Q&A: A Guide to Communicating with Your Child's Care Team\*](#) that can help you interview care teams and gather more information about potential hospitals for your child's birth and surgery. See [Chapter 18](#) for the Single Ventricle Q&A: A Guide to Communicating with Your Child's Care Team.

## How will I deliver my baby?

Many women ask about whether a special type of delivery is recommended for the birth of their baby because they are worried about how their baby will do during labor. In general, babies with HLHS will do well and can tolerate labor and delivery just like any other baby. You will be followed very closely during pregnancy by both the OB and fetal cardiologist. If special delivery plans need to be made, your care team will explain those to you.

For the vast majority of women, the mode of delivery will be based on the recommendation of the obstetric team. You should expect to receive the same type of consideration that any woman has about delivery mode. All delivery decisions will be based on the specifics of the individual mother, current pregnancy considerations (baby size, baby position, mom's health factors, etc.), and birth history for any prior children. Fetal cardiologists do not make the final decision about how a baby should be delivered, but they do give guidance to the obstetrical team about any special care that your baby might require after birth.

Timing of delivery depends on the unique needs of both you and your baby, but the current recommendation, based on current research, is to take a pregnancy as close to 40 weeks as possible. Sometimes for planning purposes (for example, if you live hours away from the hospital), an induction of labor may be recommended for the delivery of your baby. This will be a discussion you will have with your care team.

## Coordination of Care

The delivery and postnatal care of a baby with HLHS requires the coordinated efforts of many different specialists and care providers. This is why many centers use a model in which nurses or other patient care coordinators are key members of the team. They help members of the team communicate about ongoing obstetric care, individualized delivery plans, and both your and your baby's specific needs. Members of both your and your baby's care team will meet throughout the pregnancy to make sure that there is a proper delivery plan in place for when your baby arrives.

You should be offered the opportunity to meet with members of your care team in the prenatal period. Examples of people you might meet before your baby arrives include:

- **Fetal cardiologist, fetal cardiology nurse and/or nurse practitioner** (specially trained members of pediatric cardiology involved in prenatal diagnosis of CHD)
- **Pediatric cardiothoracic surgeon** (surgeon who operates on your baby's heart after birth)
- **Neonatologist, neonatal nurse practitioner** (individuals who provide specialized intensive care to babies)
- **Maternal-fetal medicine specialist, perinatologist, or OB** (members of the team delivering your baby and caring for you through pregnancy)
- **Social workers** (individuals who help you and your team communicate and ensure that you and your family are coping well through the entire process)

## What happens in the delivery room?

On the day of delivery, your care team will be ready for your baby's arrival. Centers that deliver babies with HLHS will be very used to the type of care required for your child after birth. For the most part, the actual labor or **cesarean section** (C-section, if needed) delivery of your baby shouldn't differ too much from the standard practices used in the delivery of any baby. Each center might differ on which care team members are actually present in the delivery room, but it is helpful to know that there will be separate teams of physicians and nurses to care for both you and your baby. If you have the opportunity during a prenatal visit to meet with a member of the neonatology team (the team that is most typically present to assess and care for babies in the delivery room), you can ask about the specific delivery room practices at your hospital.

Much like any other delivery, the baby will receive an assessment in the delivery room. This includes a quick exam of the baby from head to toe and an evaluation of the baby's oxygen levels (**oxygen saturations**). Most babies born with HLHS look healthy in the delivery room. If, for any reason, your baby requires extra support, like oxygen, this can be done in the delivery room. It is important for you to know that many babies without CHD require oxygen or extra treatments in the delivery room, and this might not mean that your baby is sick. He or she might just be transitioning to life outside the womb like any baby needs to do after delivery. Other treatments, like putting in **intravenous (IV) lines** or **umbilical lines**, performing an echocardiogram of the baby's heart, and giving medications like **prostaglandin** will most likely happen later, once the baby has moved out of the delivery room.

In most cases, the team delivering and stabilizing your baby will make every effort to allow you to see, touch, or even briefly hold your baby in the delivery room. Questions about how much bonding typically occurs in the delivery room differ between centers and should be asked about during prenatal visits. Although all efforts are made to ensure that you have the most normalized delivery experience possible, your care team recognizes that bonding in the delivery room is shorter than is typical. Your experience is still unique, and families will be supported and encouraged to bond with their child in the **intensive care unit (ICU)** once mom and the baby are stable after delivery.

## Where does my baby go after delivery?

Each center will have a slightly different plan for where your baby goes after delivery, but your baby will go to the place with a team that is able to deliver the best and most comprehensive care for your child. Typically, this place is an ICU (either the neonatal intensive care unit [NICU] or **cardiac intensive care unit [CICU]**). Some hospitals are set up for women to deliver in the same building where the ICUs for newborn babies are located. Other hospitals have a labor and delivery unit that is located within one building and an ICU in a neighboring building or a short transport away from where the baby is delivered.

The majority of centers will allow a support person to accompany your baby to the ICU while mom is stabilized after birth. Care teams will be mindful of how stressful it is to be separated from your baby and support person, and they should give you updates on your baby throughout the post-birth recovery period. Once you are stable, the care team will take you to see your baby.



## Delivery and the First 24 Hours— One Parent’s Perspective



*“Between the diagnosis and birth, I went to weekly ultrasounds at a closer location to monitor the baby’s progress. The findings would then be sent to Children’s in Minneapolis. As the due date approached, we met with the cardiac surgeon, the hero in our tale, who went over the details of the first surgery and answered any questions we may have had regarding the first stage.*

*We also met with the birthing team to discuss what the day would be like, and the days leading up to the first surgery, which typically takes place 3 to 4 days after birth. One of the goals of the team is to prolong the pregnancy as long as possible; for, the bigger the baby, the better. I was able to make it to full term and was induced a week after my due date.*

*Another goal they had was to be able to have the mother deliver vaginally, so we were able to achieve that goal as well. Since this was not my first child, I was pleased to know that much of the delivery would be like a “normal” birth. The only difference would be that I would be moved to an operating room right before I started pushing. This way, the entire team could be right there to take over after birth. One commonality between many heart families is that we rejoice for any chance at “normal.”*

*I loved that my family could be present for much of the delivery and that my husband would be allowed to partake in every part of the*

delivery. The greatest comfort I had was that my husband could follow our baby to the next room for monitoring.

I was able to hold my baby for a minute before she was taken by the cardiac team, with my husband then following them to watch over AND take about one hundred pictures along the way. From my understanding, HLHS babies with no complications are monitored until the day of surgery and are able to stay awake until being **intubated** right before surgery. Our daughter did not respond well to one of the drugs, so she had to be intubated from her day of birth until the day of surgery. After the fact, I heard that this reaction is quite common, but I was not aware of this at the time. That made for a very difficult first night for all involved.

Once she was intubated, she remained that way until surgery day. Although a limited number of guests were allowed, we were blessed to be alongside her until she was taken away for surgery. Surgery was a huge success and went without any complications. This would be the first of many times, Miss Adeline proved to be a heart warrior!”

**Tara**

MOTHER OF ADELINE

# 4

## Preoperative Plan

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**David Parra, MD**, *Monroe Carell Jr. Children's Hospital at Vanderbilt*

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

This section of the guide offers information about what to expect in the first days and weeks of your child's life.

## Arrival in the ICU

After delivery, your baby will be admitted to the NICU or CICU, where IV or ***intra-arterial lines*** may be placed. Some lines may be placed in the umbilical cord stump (umbilical lines). If your baby is born at a hospital that does not do heart surgeries for children, your baby will be transferred to a pediatric cardiac surgical center. Special intensive care teams transport these infants by ambulance or by air. Some centers allow an adult family member to ride along if the baby is doing well. Some infants may need a breathing tube (***endotracheal tube***) and breathing machine (***ventilator***) during long transports.

## Your Child's Care Team

**Fetal cardiologist:** Fetal cardiologists are heart doctors who have had special training and use ultrasound pictures to recognize heart problems in children *before* birth. They help families plan for their child's care during the pregnancy.

**Primary pediatric cardiologist:** Pediatric cardiologists are the doctors who provide medical care for your child's heart defect. Your child will have check-ups with a primary cardiologist during childhood. Your child's primary cardiologist may be the cardiologist you saw during your pregnancy or a cardiologist you meet after birth.

**Cardiothoracic surgeons:** Cardiothoracic surgeons have had special training and are experts in performing surgery for heart problems.

**Intensive care specialists:** Intensive care specialists are doctors who are experts in caring for critically ill children and who will be in charge of the medical care of your child while he or she is in the ICU.

All of these physicians, along with bedside nurses, respiratory therapists, hospitalists, physician assistants, and nurse practitioners (specialized nurses) are part of the team caring for your child. You will also meet ***physical therapists, occupational therapists, speech therapists***, social workers, and other staff members who are there to support your child's development and your family's well-being.

Use this section to write down the names of your care team members:

**Fetal Cardiologist:** \_\_\_\_\_

**Primary Pediatric Cardiologist:** \_\_\_\_\_

**Cardiothoracic Surgeon(s):** \_\_\_\_\_

\_\_\_\_\_

**Intensive Care Specialist(s):** \_\_\_\_\_

\_\_\_\_\_

**Nurse(s):** \_\_\_\_\_

\_\_\_\_\_

**Respiratory Therapist(s):** \_\_\_\_\_

\_\_\_\_\_

**Nurse Practitioner(s):** \_\_\_\_\_

\_\_\_\_\_

**Physical Therapist(s):** \_\_\_\_\_

\_\_\_\_\_

**Occupational Therapist(s):** \_\_\_\_\_

\_\_\_\_\_

**Speech Therapist(s):** \_\_\_\_\_

\_\_\_\_\_

**Social Worker(s):** \_\_\_\_\_

\_\_\_\_\_

## Understanding the Intensive Care Unit (ICU)

Infants with HLHS or other single ventricle heart defects are admitted to a NICU, a CICU, or a **pediatric intensive care unit (PICU)**. Your baby will be on a monitor that continuously shows your baby's heart rate, blood pressure, oxygen level, and breathing (**respiratory**) rate. Your baby will require frequent blood tests, many of which may be drawn using the IV lines placed in the delivery room or in the ICU.

Your ICU team understands the importance of involving families in their child's care. Though visiting practices may vary, all of our centers assure that families have time to visit, hold, and bond with their child. Parents are encouraged to help care for their baby. Families are part of daily rounds in the ICU where the team makes plans for your child and answers questions.

## Care Before Surgery

Most infants with HLHS are stable in the first days of life. The time before the first surgery is used for testing and preparing for surgery. Your child will have at least one complete echocardiogram (ultrasound of the heart) to confirm all of the details of the heart anatomy. Your child will also have at least one electrocardiogram (ECG, EKG) to look at the heartbeat pattern (rhythm). He or she will be watched on a heart monitor (**telemetry**) to look for abnormal heartbeat patterns (**arrhythmias**). In some cases, additional tests may be required to look more closely at parts of the heart or lungs. These tests may include cardiac catheterization and **angiography, cardiac MRI, or cardiac CT scan**.

Some infants with HLHS may be quite ill in the days before the first surgery. This is particularly true for infants with a restrictive or intact atrial septum (no ASD), a condition which limits the return of blood from the lungs. Infants with a restrictive or intact atrial septum may require special procedures immediately after birth to relieve this blockage. Other infants may be unstable because they have too much blood flow to their lungs and not enough blood flow to the body. These infants may require support with a breathing machine (ventilator), with oxygen, or with special inhaled gas mixtures. Some will require IV heart medications, which may help with heart performance and blood pressure.

## Testing Before Surgery

Infants diagnosed with HLHS or other single ventricle heart defects may have additional

health problems beyond the heart and will be checked for these after birth. Head and kidney (**renal**) pictures (ultrasounds) are commonly included during the period before heart surgery, as problems in the brain or kidneys are important to know about when planning for surgery. If your child's genes were not examined during your pregnancy, tests can be performed on cord blood or on your baby's blood after birth. Some infants have syndromes where there are multiple health problems in addition to the heart defect. Genetic testing may help to identify other potential problems, which may be important in your child's care.

---

**TOBY**



Toby was born with double outlet right ventricle, transposition of the great vessels, and pulmonary stenosis. He had his first heart surgery at 20 days old. He is now over five months and loves playing with his grandparents all day and keeping his parents up all night.

---

## Nutrition Before Surgery

Your baby will need nutrition in the days before the first surgery. The type of nutrition your baby will receive depends on his or her unique situation. If possible, breastmilk or formula may be offered to your baby. If your child is too sick or unable to eat by bottle or breast, nutrition may be given through an IV or by a feeding tube placed through the nose that delivers small amounts of breastmilk or formula. Many babies will be able to feed before surgery. Feeding practices vary between centers. At some centers, breastfeeding is allowed in the days before surgery. In others, infants may be given

pumped breastmilk by bottle so that the amounts can be tracked closely. Your baby's nurses will help with holding and positioning your baby for feeding.

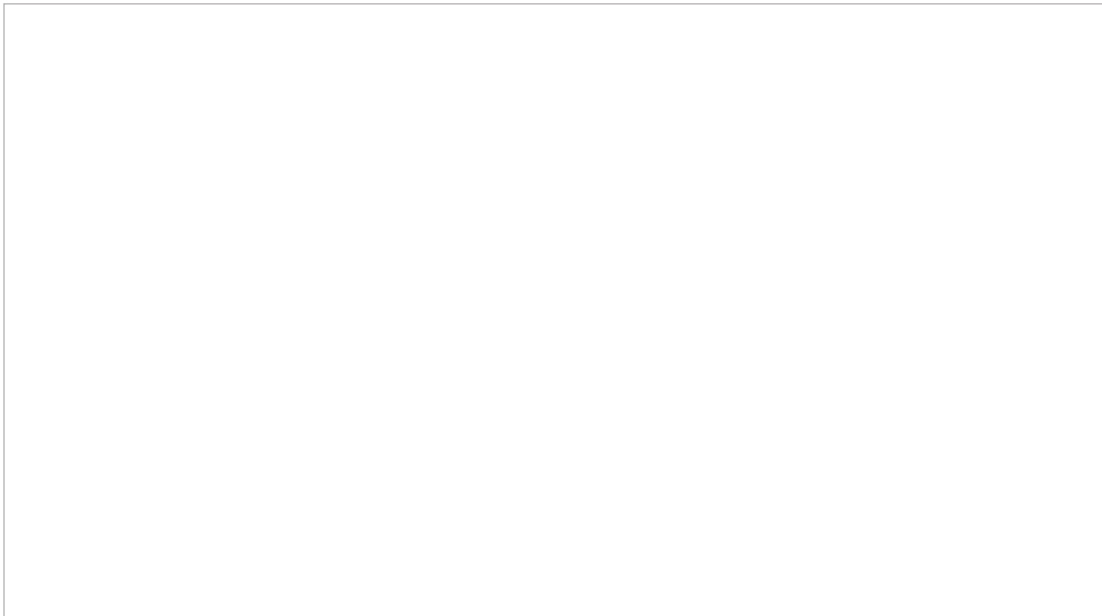
## Family Support

These early weeks of life are challenging for families. Members of your care team will have information about resources that may be helpful to you during this time. The details will differ between centers but may include information about places to stay during your child's time in the hospital (including sleep rooms, a nearby Ronald McDonald house, or low-cost hotels). There may be places for laundry, showering, and meals. There will be resources for breastfeeding mothers to pump and to store breastmilk. There may also be information about local support groups for families of children with heart disease.

You may have questions about what you should bring for your child. Both before and after surgery, your child will have special needs in terms of clothing and his or her bed space. These will be met by the hospital. Your team can let you know when clothing or special family items like blankets can be brought in from home.

## Your Hospital

At your hospital, there may be specific unique approaches that your care team may want you to know about. They are added here:





# 5

## Stage 1 Hospital Stay

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

This section of the guide offers information about what to expect in the hospital after your baby has had his or her Stage 1 surgery through hospital discharge. This section provides a general overview of your baby's hospital stay.

## Day of Surgery

Your baby will likely be taken to surgery from the intensive care unit (ICU) early in the morning, and regardless of the type of surgery planned, will likely be in the **operating room (OR)** for most of the day. During the surgery, you will be updated on a regular basis by a member of the surgery team.

### In the Operating Room

Your baby will be taken to the OR by the anesthesia team. **Anesthesia** is the medicine that is given to allow your baby to sleep through the operation and be comfortable. After your baby is put to sleep, small flexible tubes, usually called lines, will be placed in some veins and arteries. These lines serve different purposes. Some are used to monitor pressure and blood flow in the heart and blood vessels. Others are used to give important nutrition and medicines to your baby. A breathing tube (endotracheal tube) will be placed in your baby's air passage to help with breathing. Another tube, called a **Foley catheter**, will be placed into the bladder to monitor urine output, which is an important way of checking blood flow to the kidneys. Your baby will also be connected to a heart rhythm monitor, called an electrocardiogram (ECG or EKG), which will constantly evaluate the baby's heart rate and rhythm. Sometimes, a special ultrasound scope is placed in the **esophagus** to allow for an echocardiogram at the beginning and end of the surgery. During surgery, your baby's heart and lung function will be supported by a heart-lung machine called the **cardiopulmonary (heart-lung bypass) machine**. This machine performs the work of the heart and lungs, and the machine is operated by a technician called a **perfusionist**.

Your baby will be cared for by the **anesthesiologist, cardiothoracic surgeon, thoracic surgery nurses**, and perfusionists throughout the surgery. When the surgery is complete, your baby will be transferred back to the ICU.

## Postoperative Course

Once the surgery and transfer to the ICU are complete, there will be connections to monitors and intravenous lines (IVs), and the incision will be covered with a dressing or bandage. Often, after this type of operation, the chest wall is not completely closed, and a special dressing is left in place. This is done because, after surgery, a lot of fluid can build up, causing the heart and chest wall to swell. Leaving the chest open can help allow space for this fluid and swelling. If your baby is left with an open chest after surgery, it is typically closed within 2 to 4 days.

To help better prepare you for what to expect after surgery, each piece of equipment is discussed in detail below.

### Lines and Tubes

A **central venous line** is a special, deeper IV that has extra ports (places to give medicines). This line is usually placed in the side of the neck or in the groin area. Blood samples can also be taken from this line.

Special lines, called right or left **atrial lines**, are IVs that are placed in the top of the belly, through the skin and into the atria (top chambers of the heart), by the surgeon at the end of surgery. Measurements of blood pressures inside the heart can be obtained through these lines. These lines also provide an additional place to give medications and take blood samples.

At least one IV line will be inserted into the hand, arm, foot, ankle, or sometimes scalp. This allows for fluids and medications to be given after the surgery, especially when the more specialized lines are no longer needed. IV pumps control how much medicine and fluids to be given through the IV lines at a specific rate based on weight.

An **arterial line (A-line)** is a special IV inserted into an **artery** (a blood vessel where you can feel the heartbeat). This allows the team to continuously measure the blood pressure. Blood samples are also taken from this line. If your baby had an arterial line in his or her belly button placed right after birth, this is sometimes kept to be used after surgery. If the arterial line was placed later after birth or in the OR, it is usually in the wrist, groin area, or sometimes in the ankle.

An endotracheal tube (breathing tube) is inserted into the mouth or nose and placed in the **trachea** (windpipe). The breathing tube connects to the ventilator (breathing machine) that will breathe for your child. How your child recovers from surgery determines when the breathing tube is removed.

There will be at least one **chest tube**. A chest tube is a tube that drains fluid or air from around the lungs or the sac around the heart into a rectangular collection box. The tube is usually placed through the skin at the bottom of the incision/in the upper belly. On occasion, chest tubes are placed on the side of the chest, if there is a need for drainage from around the lungs.

Temporary **pacemaker wires** are small wires placed through the skin at the top of the belly that attach directly to the surface of the heart. These wires can be used to attach to a pacemaker machine, if necessary, to temporarily generate heartbeats at a desired rate.

A urinary (Foley) catheter is a tube placed into the bladder that drains urine into a collection bag. This allows your child's medical team to measure the amount of urine produced each hour more accurately.

A **nasogastric (NG) tube** is a tube placed in the nose and down into the stomach. This will keep the stomach empty to decrease the chance of vomiting. If your child has this tube after surgery, it can be used to give breastmilk, formula, or other nutrition.

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## BODIE



Two weeks old, recovering from his Norwood surgery. Among the many tubes and wires placed, Bodie was fed through an NG tube.

Although initially overwhelming, in the days and weeks to come, Bodie learned to eat orally, and lost not just the NG tube, but all of the other tubes and lines in preparation for his discharge home.

---

**Vital signs** are displayed on the monitor located above your baby's bed. This can include heart rate, respiratory rate, blood pressure, oxygen level, temperature, and other specialized measurements if needed.

**ECG/EKG leads** are stickers that are placed on the chest and belly that detect the heart rate and rhythm and breathing rate to be displayed on the monitor.

A **pulse oximeter** is a sticker that resembles a Band-Aid with a red light that measures the amount of oxygen or oxygen saturation level in the blood. This is usually placed on a finger or toe. On small babies, this may be placed around the hand or foot.

## Pain Control

A priority of your doctors is to make sure that your baby is as comfortable and calm as possible. Immediately after surgery, your baby will likely receive pain medications continuously through a pump that controls one of his or her IV lines. As-needed doses of pain medication can also be given if you or the team feel that your baby is uncomfortable. As your baby becomes ready to have the breathing tube removed (**extubation**), these pain medicines will be decreased to allow him or her to wake up more and breathe on his or her own. Pain medications can also be given through a feeding tube or orally (by mouth) as your baby continues to recover.

## Medications

After surgery, your baby will likely be on several medications. All babies will need a diuretic after surgery – a medication to make them pee (urinate) – to help them get rid of extra fluid that always collects after a big surgery and being on the heart-lung bypass machine. Most babies with HLHS will go home on some form of diuretic by mouth, typically a medication called **furosemide**, to continue to help keep extra fluid out of the lungs until the next surgery.

Your baby may also go home on additional medications that will be given by mouth or through a feeding tube. Many babies with HLHS will be on a small dose of **aspirin** used as a mild blood thinner. There are other types of blood thinners that can also be used, some of which are given as an injection. Some babies with HLHS will need to be on other medications for their heart to either keep blood pressure under control, help the heart deal with leaky valves, or to help the heart squeeze/function. These medications will be determined on a case-by-case basis by your baby's cardiology team. You will receive teaching on all of your baby's medications – what they are for and possible side effects, as well as how often and how to give them – before your baby leaves the hospital. Please see the *Single Ventricle Guide* section on Medications in [Chapter 16](#) for more information.

## Feeding and Nutrition

After surgery, it will be important for your baby to get enough nutrition. Initially, this will

be given through an IV and/or feeding tube. As your baby recovers, working on feeding – both tolerating feeds and learning to eat by mouth – will be an important part of his or her recovery and preparation for going home. Most babies with HLHS will at least temporarily require an NG tube to provide some nutrition as they recover from surgery and work on learning to eat by mouth. Every baby is different, and your baby's team will work to make sure that your baby's nutritional needs are met and that your baby is fed safely and effectively. Please see the *Single Ventricle Guide* sections on Nutrition and Growth in [Chapter 7](#) and Tube Feeding in [Chapter 8](#) for more information.

## Transfer to the Floor from the ICU

Once your baby no longer needs a breathing tube and isn't requiring any IV blood pressure medicines, he or she will be moved to the surgical ward, or **step-down unit** where the care provided is less intensive. This is a good thing and means that you are getting closer to going home. The team will not let your baby leave the ICU until they are comfortable that he or she is ready. All hospitals will do a detailed hand-off between the ICU team and the surgical ward team to make sure that the new team of doctors and nurses knows everything that they need to know about your baby. In some hospitals, you may be invited to be a part of this hand-off. This is a great time to ask questions and point out things that you've noticed about your baby since you know your child best!

## The Journey Toward Home

To get ready to go home, you will gradually be expected to do more and more to care for your baby. There will likely be several classes that will help prepare you to be at home, including **CPR**. Your team will also spend a lot of time in the weeks and days leading up to discharge teaching you how to use any equipment that you might need at home. You will also be taught a list of *red flags* – signs and symptoms to watch for in your baby that may signal that something is wrong. Use the time in the hospital to get to know your baby well so that you will notice when something changes! At most hospitals, the parents are expected to complete a **rooming in** that typically lasts 24 to 48 hours. During this period, you will provide all of the care to your baby, including feeding and giving medicines, while in the safety of the hospital with the nurses and other providers nearby. This is a time to practice and make sure that all of your questions are answered.

## Your Hospital

At your hospital, there may be specific unique approaches that your care team may want you to know about. They are added here:







# 6

## Interstage Monitoring

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### **PARENT PARTNERS**

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

## Getting Ready to Go Home

Going home from the hospital can be stressful for families. Families become used to having the medical team close by, and many families feel better knowing that their babies are being watched closely. Babies are particularly at risk between the Norwood procedure and Glenn procedure because they typically rely on either the Blalock-Thomas-Taussig (BTT) shunt or the Sano shunt to give blood flow to the lungs, and their heart has to work harder than a normal heart.

You will go home with a few simple tools, including a scale to weigh your baby and a monitor to check your baby's oxygen levels (saturation). Your care team will also give you specific things to watch for and tell you how to reach them if you are worried. The reason for this extra monitoring is to catch any potential problems as early as possible. Although this may seem like a lot at first, your care team will work with you to make sure you are as comfortable as possible with the equipment you bring home with you. Many families tell us that these tools make them feel safer to be at home with their baby. Usually, you will not need to keep using these tools as often after your baby has his or her next surgery, the Glenn procedure. There is more information about each of these tools below.

### Oxygen Saturation

As mentioned in the last chapter, a pulse oximeter (or "pulse ox") is a machine with a sticker that resembles a Band-Aid with a red light that is usually placed on a finger or toe. The machine tells you the oxygen level, usually referred to as the oxygen saturation, or "O<sub>2</sub> sat," and allows your heart care team to see if your baby has the right amount of blood flow going to the lungs and body. Your care team will tell you how often to check your baby's O<sub>2</sub> sats and will ask you to communicate that information to your team. Most parents become familiar with this number and learn what their baby's usual O<sub>2</sub> sats are during the hospital stay. Your care team will give you the O<sub>2</sub> sats that are normal for your baby and tell you how to contact them if they are out of range.

### Weight Gain

Another good way for your care team to monitor if your baby is doing well is to see if he or she is gaining weight. Babies usually do not gain weight well if their heart or lungs are



Pumps help staff to give various medications and flushes that need to be delivered through IVs. Patients are often on various medications to help with heart pumping, blood pressure, and pain.

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working too hard. As part of the home monitoring program, you will be asked to weigh your baby each day to see if he or she has gained or lost weight. Your care team will let you know how much weight your baby should gain and when to become concerned.

### **Recording Progress**

In order to keep track of how your baby is doing, you will be asked to communicate your baby's O<sub>2</sub> sats and weight and how much food your baby is getting (by nursing, bottle feeding, or through a feeding tube). The team will tell you what is normal for your baby and when to call them. You will need to notify the care team if the baby's saturations are too low, they are not gaining enough weight, they are losing weight, or for other reasons that will be explained, depending on your child's specific issues. Writing down all of this information will also let your providers see how your baby is doing between your visits to the cardiologist's office.



# 7

## Nutrition and Growth in Patients with Single Ventricle CHD from Birth to Fontan and Beyond

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*Chelsea Scaramuzzino, University of Rochester Medical Center*

*With contributions from members of NPC-QIC Nutrition and Growth Lead Group*

DYLAN



Dylan was born with HLHS. He is now 6 years old, and loves to swim, play the piano, and practice karate. His favorite food is pizza.

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

Nutrition and growth in children with single ventricle hearts are important, yet at times can be very challenging. This section divides nutrition into the pre- and post-Norwood stages and the Interstage. Each stage is unique, given the changes in blood flow and complications that can occur. The overall goal is to ensure healthy weight gain and normal eating habits and to give families tools, resources, and confidence for an overall healthy lifestyle.

## Human Milk/Donor Milk/Formula

There are many different ways to feed your baby. Breastmilk is the preferred milk because it provides special benefits to your baby's health. Breastmilk can help protect your baby against infections, is easily digested, and may potentially reduce the severity of some allergies. The first milk that your breasts make is called **colostrum** and it is important because it helps your baby build a strong immune system, creates a tough coating on your baby's stomach and intestines to keep germs from causing illness, and helps to prevent low blood sugar in your newborn.

If you are unable to provide breastmilk or choose to use formula, banked donor milk, or another milk, they will also provide full nutrition and meet the needs of your growing baby. Whichever form of nutrition is provided, the most important factor is that it promotes your baby's brain growth, nervous system, and eye development.

- Some centers may allow you to breastfeed or bottle feed if your baby is stable. This

will depend on several things, like how fast your baby is breathing, if he or she has a breathing tube, and the overall well-being of your baby.

- Most centers encourage the use of breastmilk. Lactation consultants and staff should provide you with the right equipment and teach you the process of pumping breastmilk for your baby when feeding directly from the breast is not possible.
- If you choose to use donor milk, you will need to sign a consent form. Banked donor milk comes from lactating moms who give their milk to a milk bank. Milk is collected from only healthy mothers who have passed a screening process. The milk is heat-treated (pasteurized) to kill known viruses and bacteria that may have been present in the milk. Donor milk may be an option if your supply is low or as your baby's primary nutrition if you desire.
- Breastmilk or formula can also be used for mouth care, like swabbing your baby's mouth, if your baby is not able to take feedings by mouth. Caring for your baby's mouth helps prevent infection and provides comfort.

## Pre-Norwood

### Non-nutritive Exercises

If oral feeding (feeding by mouth) is limited or not allowed, it is important to continue to encourage the developmental part of feeding, which includes sucking and swallowing. Ask your center what their guidelines are regarding holding, skin-to-skin or **kangaroo care** to promote bonding with your baby.

Some centers will allow/encourage your baby to develop his or her **oral motor skills** with the following:

- **Pacifier dips:** dipping a pacifier in breastmilk or formula for your baby to practice sucking
- **Non-nutritive breastfeeding:** allowing your baby to suck on your nipple after you have pumped
- **Oral motor exercise:** special exercises done at the bedside to strengthen your baby's oral motor skills

### Feeding/Nutrition in the Intensive Care Unit (ICU) Setting

The goal in the pre-Norwood stage is to provide adequate nutrition to your baby before surgery. Depending on your center's practice, your baby may be started on some feeds (you may hear this referred to as **enteral feeds**) if he or she is stable before surgery. It is

common that IV nutrition, also known as **total parenteral nutrition (TPN)** or **Intralipids (IL)**, is used to provide full or supplemental nutrition. Common methods used include:

- **TPN/IL:** A special liquid food mixture that is given through an IV into your baby's blood. The mixture contains proteins, carbohydrates, fats, vitamins, and minerals to provide optimal nutrition to your baby before surgery.
- **Feeding tube:** A small tube that typically goes through your baby's nose or mouth into his or her stomach or small intestine to provide breastmilk or formula:
  - **Nasogastric tube: NG tube, nose to stomach**
  - **Orogastric tube: OG tube, mouth to stomach**
  - **Nasoduodenal tube: ND tube, nose to first part of small intestine (duodenum)**
  - **Nasojejunal tube: NJ tube, nose to second part of small intestine (jejunum)**
- **Nasogastric tube (NG tube):** This is a small tube that goes through your baby's nose to his or her stomach to provide breastmilk or formula. Your team will teach you how to use this tube and replace it if necessary. If your baby cannot eat by mouth or finish a full feeding by mouth, the remainder of the feed can be given through the NG tube.
- **Nasojejunal tube (NJ tube):** Sometimes babies may need the feeding tube advanced past the stomach into the jejunum (small intestine), usually due to problems such as reflux. This is called Nasojejunal feeding with an NJ tube.
- **Gastrostomy tube (G tube):** A G tube is a feeding tube that is placed directly into the stomach by a surgeon. If your baby cannot eat by mouth or finish a full feeding by mouth, the remainder of the feed can be given through the G tube. Again, some babies will need the tube advanced past the stomach into the jejunum, and this is called a **gastrojejunal (GJ tube)**.
- **po (Latin term that means by mouth):** Allowing your baby to eat by mouth based on cues or limited volume depending on your baby's stability and your center's practices.

## Post-Norwood

### Nutrition Management After Surgery

The main goal for post-Norwood nutrition is to start nutrition early in an effort to promote healing and allow your baby to grow. There will likely be various ways to make sure that your baby is getting adequate nutrition:



- Oftentimes, the initial source of your baby's nutrition will be from TPN/IL through an IV. This is a concentrated source of nutrition that can be given in a small amount of fluid.
- After surgery, fluid may be limited, and nutrition may continue to be delivered through an IV. As recovery starts, medications are decreased, and your baby becomes more stable from surgery, the ICU team will begin introducing oral or tube feeds (breastmilk or formula). Some centers may have a feeding protocol they follow when starting feeds. Most centers begin feeds through a feeding tube, which could be inserted through the nose or mouth and into the stomach or jejunum (small intestine).
- Over the next several days, the amount of feeds your baby is given will increase, and the TPN/IL will be weaned off.
- Once your baby has the breathing tube removed and is at a safe oxygen level, your team will check the safety and readiness for your baby to start taking feeds by mouth. Your feeding therapist should help begin this process and continue to monitor your baby's progress.
- Your baby might have difficulty feeding at first, which is not uncommon.

### Possible Post-Norwood Complications That Can Affect Feeding/Nutrition

- **Vocal cord dysfunction:** The *recurrent laryngeal nerve*, which is an important nerve that coordinates normal breathing and swallowing, is located next to the aorta. During surgery, particularly surgery where the aorta is reconstructed (like with the Norwood), the recurrent laryngeal nerve can be stretched or injured, causing vocal cord dysfunction. If this happens to your baby, he or she may have a hoarse cry or high-pitched, wheezing sound (*stridor*). If so, he or she may be at a higher risk for *aspiration* (where the feedings can go into the lungs). Your medical team will decide if there is enough concern to order an evaluation of your baby's vocal cords. A doctor who specializes in surgery related to the throat (*otolaryngologist or ear, nose, and throat [ENT] specialist*) may come to evaluate your baby's vocal cords with a small scope.
- **Swallowing problems:** Babies with single ventricle CHD can have difficulty swallowing. Problems that can affect swallowing include vocal cord dysfunction, decreased muscle strength, and breathing difficulties. A feeding therapist (usually a speech therapist or occupational therapist) will work with your baby to improve swallowing function. A common test used to evaluate your baby's swallow function is a *video fluoroscopic swallow study (VSS)* also known as a *modified barium swallow test*. This test is done using x-rays to actually watch how your baby is swallowing.

- **Gastroesophageal reflux disease (GERD):** Babies with single ventricle CHD may have increased reflux that can cause a burning sensation in the throat during or after feeds. Babies with reflux may be irritable, take less feedings by mouth, or even vomit. They may need special positioning and formulas or medications to help make the symptoms better.
- **Chylous effusion:** This is a milky, high-fat drainage from the chest tube that can occur when the lymphatic system in the body has been damaged. Some babies with single ventricle CHD are born with abnormal lymphatic systems. This is treated with dietary changes. Your baby's diet will be changed to either de-fatted breastmilk or a low-fat formula.
- **Poor incision healing:** Your team may change your baby's breastmilk or formula to provide more calories and protein for your baby to heal properly. Your baby may need extra vitamins and minerals to aid in incision healing.

## Feeding Evaluation

There are several types of tests to make sure your baby is safe when taking feeds by mouth. These tests can vary, depending on how your baby is doing and what the practice is at your center. Some examples of these tests are:

- **Bedside evaluation by feeding therapists:** Occupational and speech therapists may be called to check your baby's feeding safety when he or she starts eating by mouth. If your baby is feeding, therapists are concerned about his or her ability to swallow; they will discuss with the team the need for testing or changes to feeding.
- **Laryngoscopy:** This test is done by an otolaryngologist, or ENT who can look directly at your baby's vocal cords with a small scope.
- **Video fluoroscopic swallow study (VSS)/modified barium swallow study:** This test is routine at some sites before they will allow your baby to eat by mouth. It is also performed if there is a concern that your baby is aspirating when eating. Aspiration is dangerous because breastmilk or formula goes into the lungs and can cause serious breathing problems and lung infections. If your baby does have aspiration, then he or she may require a special bottle nipple, thickened feeds, or a feeding tube for complete nutrition if it is not safe to eat by mouth. This test is done using x-ray to watch how your baby is swallowing the breastmilk or formula.
- **Fiberoptic Endoscopic Evaluation of Swallowing (FEES):** This test is done by an otolaryngologist, or ENT. It is used to look at vocal cord function and to determine risk for aspiration. Some centers use this test instead of the swallow study.

## Importance of Weight Gain and Feeding Goals

As your baby gets close to hospital discharge, the team will teach you how to mix formula, feed and weigh your baby, and monitor his or her nutrition. The main goal in the Interstage is for your baby to gain weight safely. Your baby will need to gain an average of 20 to 30 grams per day. If your baby cannot take all of his or her feeds by mouth, he or she may need supplemental (extra) feedings using a feeding tube. Your team will determine what type of feeding tube your baby will need. See the [\*Feeding/ Nutrition in the Intensive Care Unit \[ICU\] Setting\*](#) section earlier in this chapter.

If your baby has to go home with a feeding tube, your baby's ability to safely eat by mouth and maintain good nutrition will determine how long he or she needs a feeding tube. At the time the tube is placed, your team should set up a plan with you for eventually removing the feeding tube. That way, you will understand what the expectations are in order for the tube to be removed as soon as it is safe to do so. It is our goal to have all babies with single ventricle CHD safely eating by mouth and with their feeding tube removed by their first birthday.

WINSTON



Winston was born with HLHS, diagnosed at 36 hours old. He is now 18 months old and 1 year post Glenn. Winston was NG tube fed from birth until 7 months of age when he was weaned to orally feeding from a bottle. Winston is a very curious boy who enjoys exploring his world. He loves being outside, playing with his “power tools” and adores his baby sister.

## The Interstage

The Interstage is the time between the Stage 1 Norwood heart surgery and the bidirectional Glenn procedure (second heart surgery). The Interstage could mean Interstage at home or in-hospital Interstage for some babies, depending on their stability. It is the most fragile period for babies with single ventricle CHD and the most

high-risk time for growth failure. The nutritional goal during this time is to maintain growth and to ensure adequate daily weight gain for your baby's second stage surgery.

## Home Monitoring Program

You will receive education on how to weigh your baby daily on a scale before being discharged home from the hospital. You will be taught how to report your baby's weight to your center's Interstage team. This team will be monitoring your baby's progress. If there are any concerns with your baby's weight, he or she may have to be readmitted to the hospital.

## Red Flags and Feeding Intolerance

You will be taught signs to look for that may indicate your baby is not tolerating his or her feedings. Some of these signs include:

- Increasing fussiness with feeds
- New-onset vomiting
- Bloody stools
- Significant changes in breathing during feeds

If your baby shows these signs, you should notify your Interstage team immediately, as these could be signs of an urgent medical need.

LAKYN



Lakyn was born with double inlet left ventricle (DILV), pulmonary atresia, and discontinuous branch pulmonary arteries. She was diagnosed after birth and underwent central pulmonary repair and BTT shunt placement. She is full of surprises and keeps her family on their toes! "Bee ready for me to pull my tube out!" says her mommy. Instead of focusing on bows, her mom has turned her efforts to the most amazing tape designs for those pink cheeks! She is a busy bee!

JACK



Jack was born with DILV, transposition of the great arteries, a hypoplastic aorta, interrupted aortic arch, and a ventricular septal defect (VSD). He is now almost 3 years old and loves being on the playground, pretending to be a fireman, and playing with his cousins. His favorite foods are pizza and grapes.

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## Re-admission

If your baby has poor weight gain over several days, then your team may have your baby readmitted to the hospital. This is not a setback, and you should not feel discouraged. It is common for the team to want to monitor your baby in the hospital to make sure that your baby's heart is still doing well after surgery. They may also make changes to your baby's feeds so that he or she gains weight.

## Outpatient Feeding Therapies

After your baby is discharged home, he or she may be referred to outpatient therapists to continue working on taking feeds by mouth. If you have not been referred automatically, ask your care team about available outpatient services. The feeding therapists will work with you and your baby to make sure he or she is making progress with feeds. Your center or your baby's pediatrician can refer you to the **Early Intervention (EI)** program in your area. Each state has a unique program that provides a wide range of services to help meet children's **developmental milestones**.

## Supplemental Feedings

If your baby cannot take all of his or her feeds by mouth, he or she may require supplemental tube feedings during the Interstage. Again, do not be discouraged. It is common for some babies with single ventricle CHD to require feeding assistance until after their second stage surgery. You and your team will continue to evaluate your baby's nutrition and growth in order to discontinue tube feedings according to your established tube weaning plan.



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*“After our son’s prenatal diagnosis, we researched all we could about HLHS. However, nothing prepared me for how difficult it would be to accept not being able to feed my son the way I intended. And I don’t mean breast vs bottle, I mean being able to feed him at all and bond with him through that very primal mother/baby experience.*

*After the Norwood, my son was reliant on a feeding tube for all his nutrition. Despite being discouraged and upset, I was very thankful for his feeding tube because I knew he was receiving not only the nutrition necessary to recover well, but also the nourishment necessary for him to be as strong as he could be in time for the Glenn. In a way, the feeding and growing aspect became one less worry and more of a sure thing. And sure things were hard to come by those days.*

*The feeding tube was a difficult life. We faced challenges. Just like overcoming the challenge of two open-heart surgeries, we successfully weaned from the feeding tube after the Glenn. Remember that a feeding tube is for the short term, not the long haul. Also, remember that you, as the parent, are the expert when it comes to feeding your baby. The feeding tube may take away the experience you were expecting, but it doesn’t take away your expertise.*

*I’m happy to tell you that my son loved his first birthday cake and today he lives to eat.”*

**Lacie**

MOTHER OF DYLAN

# 8

## Tube Feeding

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

For some children with CHD, total or supplemental nutrition by a feeding tube may be necessary. Navigating this new way of feeding your child can feel overwhelming and scary. This section was designed by parents of tube-fed children with CHD, together with mental health and healthcare professionals, to provide information and resources that may be helpful as you begin your journey with a feeding tube.

## Feelings

Many parents of tube-fed children experience a roller coaster of emotions. While parents may feel relief and comfort knowing their child is safely receiving the nourishment he or she needs, it is also natural to feel frustrated, defeated, and unprepared.

Many parents experience feelings of uncertainty about their child's future with a feeding tube and anxiety regarding feeding schedules, weight gain, and managing new medical equipment and supplies. Also, some parents report that they often feel alone and isolated. These feelings are normal, and you are not alone. Here are some things that other parents of tube-fed children have found helpful:

- Connect with other families of tube-fed children through your medical center, local support groups, or online resources
- Discuss expectations with your child's care team—it's never too early to discuss your child's tube weaning plan
- Be easy on yourself; a feeding tube does not mean you or your child have failed
- Take time to acknowledge the positive progress in your journey
- Identify a trusted caregiver who can help to give you much-needed breaks

## Stress

It is natural for parents to have stress regarding their child's nutritional needs, and parents with a tube-fed child have unique hurdles. Knowing what to expect can help you better prepare yourself and your child. Parents often identify some of these things as stressors as they adjust to life with a feeding tube:

- Experiencing tube feeding side effects such as vomiting, retching, food refusal, **oral aversion**, **gastroesophageal reflux**, and/or difficulty with weight gain



- Uninvited advice from family or friends on how to “get your child to eat”
- Pressure and expectations from providers regarding caloric intake or weight gain
- Frustration caused by lacking a strong support network
- Difficulty in identifying the provider who is your go-to for feeding tube issues and concerns
- Additional responsibilities, feeling more like a medical provider than a parent
- Uncertainty of the future and/or lack of a detailed plan
- Learning how to use new equipment and adjust to it in everyday life

You may not be able to get rid of all of these stressors, but you can learn how to best manage them and be aware of how you react to them moving forward. Connect with your child’s care team in discussions regarding your concerns. Remember that you are all on the same team and one team member’s input is just as valuable as another. As the parent, you know your child best.

## Bonding & Mealtime Relationships

Bonding with your baby may be difficult when you feel you are unable to feed your baby the way you may have planned. It is normal to wonder when and how your child will develop their feeding and drinking skills and no longer need a feeding tube. Remember that a feeding tube is only a tool, just like a spoon or a bottle. As a parent, your role is still a meaningful and important one. There are ample opportunities for bonding and developing healthy food and mealtime relationships at any age, no matter how your child is being fed:

- A foundation of trust and support between you and your child is important
- Promote regular, positive, and fun opportunities around food and eating
- Manage your expectations—it is common that a tube-fed child will not show interest in oral eating right now
- Celebrate all successes; none is too small
- Remember that sometimes less is more—pushing or being forceful can prevent rather than promote progress
- Encouraging safe oral stimulation through exploration or play can allow your child this important connection with eating food by mouth
- Tube feeding can still be “normal” feeding and is not a “medical” event—holding

your baby while he or she is tube fed and having your child be a part of the family mealtime is still possible

## Self-Doubt & Empowerment

Feeling disconnected or doubting yourself can be common as a parent of a tube-fed child. This is especially true if you are also preparing for another heart surgery. The stakes feel high, and this new world of feeding your baby can be intimidating and/or overwhelming. As a parent, you are the expert on your child, and therefore the most important part of their feeding/nutrition plan:

- Trust your instincts
- Do not hesitate to ask questions; and be a strong advocate for your child
- You are enough! Align your resources at your medical center and revisit the development plan that is right for your child's age often
- Don't be afraid to think outside of the box, discuss new ideas, explore all options
- Give yourself credit for all that you've learned

## Support

There are many supports available for families with tube-fed children with CHD.

### Clinical Resources

Resources may vary by medical center. It is important to understand your therapists and their role in the present and long-term care for your child's nutrition, feeding tube management, and weaning. Here are some of the therapy resources parents have used:

- Speech-language pathology (SLP)
- **Physical therapy (PT)** and **occupational therapy (OT)**
- Early Intervention or Baby Net Services—services vary by state.  
Seek referral before discharge
- Formal tube weaning programs

### Community Support

Finding online forums and support groups that allow you to connect with other families going through the same challenges can be greatly helpful. Check out:

- Local and national CHD parent support groups like Sisters by Heart, Mended Little Hearts, Pediatric Congenital Heart Association, etc.: [sistersbyheart.org](http://sistersbyheart.org), [facebook.com/Mended Little Hearts National Organization](https://www.facebook.com/MendedLittleHeartsNationalOrganization), [conqueringchd.org](http://conqueringchd.org)
- Feeding Tube Awareness Foundation: [feedingtubeawareness.org](http://feedingtubeawareness.org)
- National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC): [npcqic.org](http://npcqic.org)
- Inside Out Care: [insideoutcare.com](http://insideoutcare.com)



# 9

## Pre-Glenn Evaluation

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The time after the Norwood surgery, leading up to the Glenn surgery, is known as the Interstage. It can be a stressful time for many families. There is fear in waiting for another heart surgery, recovery, and hospitalization.

## When will my baby have the Glenn surgery?

- Glenn surgery usually occurs at 6 months of age but can be done any time between 3 and 9 months old.
- Timing of the surgery will depend on your baby's size and oxygen levels.
- As your baby gets bigger, he or she will start to outgrow the shunt placed during the first surgery. Your baby may have lower oxygen levels as he or she outgrows the shunt.

ELISE



Elise was born with HLHS, and this picture reflects how she was just a few days after her first open heart surgery, at 2 days of life. She's now just over a year old and has the most infectious smile ever.

## Why does my baby need a cardiac catheterization before the Glenn?

- During the Interstage, babies are followed closely by their cardiologist with echocardiograms, **ECGs**, chest x-rays, **pulse oximetry**, and weight checks. Before the Glenn surgery, there are several additional tests that are performed.
- Cardiac catheterization is one of these tests. It helps the **cardiologists** and surgeons get measurements that show how well the heart is working and how well-prepared your baby is for the Glenn.

- Pressure and resistance through the blood vessels in your baby's lungs are the most important measurements taken at the pre-Glenn catheterization. Results from this test help guide the surgical plan.
- The doctor performing the cardiac catheterization will meet with you before the test to discuss the details and answer any questions you have.

## What happens during the cardiac catheterization?

- Before the procedure, a nurse will put an IV in your baby to give fluids and medicine to help him or her stay calm, relaxed, and asleep during the procedure.
- A specially trained cardiologist will use a flexible, long, thin tube called a **catheter** during the procedure. The catheter is inserted in a blood vessel near your baby's groin. This tube is then followed with x-ray pictures as it is moved through blood vessels that eventually lead to the heart.
- Detailed, high-resolution pictures of the heart are taken, and measurements of oxygen levels and pressures are made.
- If needed, the cardiologist may also perform additional procedures. There are special catheters, balloon catheters, and other devices that the doctor can use to make narrow blood vessels bigger and get rid of any abnormal or extra vessels.

STRIVER



Striver was born with Double Inlet Left Ventricle (DILV) with Transposition of the Great Arteries (TGA). He had the Damus-Kay-Stansel (DKS)/partial Norwood at 3 days old and the Glenn at 5-1/2 months old. He is now ONE! Striver loves reading, wrestling, and walks to fountains. He laughs all the time and knows how to flirt, especially with his nurses at CHLA.

## Are There Any Risks?

- As with any procedure, there are risks. A cardiologist will explain all the risks that can occur.
- Common complications include bleeding, injury to blood vessels, blood clots, and abnormal heart rate or rhythm. Rare complications include brain injury, infection, damage to the heart, stroke, blood vessel tears or rupture, and death.

## What is a good pre-Glenn result?

- The best candidates for Glenn surgery have well-developed blood vessels and blood flow to the lungs.
- Additional good test results are a widely open aortic arch, normal heart function, and minimal valve leakage.
- If results are not good, surgery may be delayed to let your baby grow more. This allows the pulmonary arteries to grow during this time, or to provide medications to improve your baby's condition.

## How do I prepare my baby for the test?

- A nurse will call you before the test with detailed instructions, like when to stop feeding your baby and what medications you can give before the test.
- Be prepared for your baby to be admitted to the hospital after the test is completed.
- Your baby will be monitored overnight and can usually go home the next morning if there are no problems or concerns.
- The nurses will give you instructions on how to care for your baby after the test.

## Why does my baby need a cardiac MRI before the Glenn?

- To get more detailed pictures of the heart, a cardiac MRI helps cardiologists and surgeons determine the surgical plan for the Glenn.
- Cardiac MRI is sometimes used instead of a cardiac catheterization.
- This does not involve any radiation, and no procedures are performed during the MRI.



## What happens during a cardiac MRI?

- Detailed pictures of the heart are taken by scanning your baby's heart, using magnetic radio waves and a computer.
- Your baby cannot move during the scan, which means he or she will need to be **sedated** during the test.
- Before the cardiac MRI, a nurse will put an IV in your baby to give fluids and medicine that will help him or her stay calm, relaxed, and sleepy during the procedure.
- A specially trained cardiologist will be present during the test to guide imaging of the heart.

## Are there any risks?

- There are few risks associated with a cardiac MRI, including limited access to the baby and breathing problems associated with **sedation**. The sedation team will closely monitor your baby during the procedure to minimize this risk.

## How do I prepare my baby for the procedure?

- A nurse will call you to give you detailed instructions. He or she will let you know when to stop feeding your baby and what medications you can give or not give before the procedure.
- You and your baby will be screened for any metal in your bodies by a questionnaire, and you will be asked to remove any jewelry or clothing that has metal.
- Your baby is closely monitored after the procedure, often overnight in the hospital, to make sure he or she is safe to go home.

## Preoperative Surgery Consult

- Once the pre-Glenn testing has been completed and reviewed by the cardiologist, a consult is scheduled with the surgeon.
- You will meet with the surgeon before the surgery to discuss the Glenn surgery in detail. The benefits and risks will be explained, including the expected postoperative course. Consent for the surgery will be obtained.
- Feel free to ask your surgeon questions you may have about the surgery and recovery.

## Questions to Consider Asking the Surgeon

- How long does the surgery take?
- How long can I expect my baby to stay in the hospital after surgery?
- Will my baby still be intubated when he or she returns from the surgery?
- What complications can happen?
- Can we donate blood for the surgery?

## Is there anything that might delay or postpone the surgery?

- If your baby gets sick with an infection before the scheduled surgery date, this may delay the procedure.
- Depending on what kind of infection it is, surgery may be rescheduled up to 6 weeks later.
- Remember to use good hand washing to help prevent your baby from getting sick, and remind everyone coming in contact with your baby to practice good handwashing.
- Keep your baby away from sick people.
- To help prevent your baby from getting sick, your cardiologist will recommend giving a special injection called **Synagis (palivizumab)**. Synagis boosts the immune system to help protect the baby against a lung virus called **respiratory syncytial virus (RSV)** often seen in the winter. The doctor's office will give this shot to your baby once a month during RSV season.

## What if my baby is not ready for the Glenn surgery?

- If your baby has concerning results on the pre-Glenn tests, this may postpone or change the procedure.
- Depending on what the problem is, your baby may have to wait longer and take additional medication before the Glenn surgery.
- Another possibility is that your baby may need to undergo a different surgical procedure first and then undergo the Glenn surgery at a later date.

# 10

## Glenn Hospitalization and Discharge Planning

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The Glenn surgery is the second of the three surgeries for HLHS. This surgery routes blood from the upper part of the body directly to the lungs, decreasing the amount of work that the heart has to do. The surgery is usually done when your baby is about 4 to 6 months old and weighs about 5 kg (10-12 lb). This is an open-heart procedure performed under **general anesthesia**. Your baby will be on a heart-lung bypass machine that will take over the job of breathing and circulating blood through your baby's body during the operation. The superior vena cava (SVC) is the large vein that returns deoxygenated (blue) blood from the head, chest, and arms back to the heart. The surgery involves separating the SVC from the heart and reattaching it directly to the pulmonary artery. Blue blood from the inferior vena cava (IVC), the large vein carrying blue blood from the lower body back to the heart, will still drain directly into the heart. After this surgery, your baby's oxygen saturations will usually be between 75% and 85%.

For many infants, the hospital recovery from the Glenn surgery is typically about a week, generally much shorter than the hospital stay after the first surgery. This is due to many factors: infants are bigger and stronger, the complexity of the surgery is often less than the first one, and for many infants, the transition back to full feeds is much faster, whether by mouth or by G tube. For those having more complex surgery with the Glenn procedure, such as a comprehensive Glenn, the hospital recovery can be longer.

## After Surgery

After surgery, your baby will be cared for in a pediatric or cardiac intensive care unit (PICU or CICU). The breathing tube is usually removed after surgery in the OR or within the first 24 hours after surgery, in the PICU. Similar to the first surgery, there will be many lines and tubes, including lines in a major artery and vein for monitoring of **vital signs**, obtaining blood samples, and giving medicines. There may also be chest tubes to remove extra fluid from the lungs and a bladder catheter to drain urine. Feeds are started when it is safe to do so.

## Nutrition During the First Postoperative Days

During the first few days after the bidirectional Glenn procedure, your baby will have changes in his or her blood flow. This blood flow is more stable than the Norwood or shunt blood flow, but it may give your baby headaches. Your baby may not feed well in

the first couple of days following the bidirectional Glenn, but this will get better. If your baby required tube feedings before the bidirectional Glenn, he or she may continue to need them during the hospitalization and for several weeks after hospital discharge.

## Glenn Discharge Planning

Planning for home after surgery is very important and starts early during your hospital stay. Below are some topics to consider as you and your child prepare to come home:

### Nutrition

Getting good calories is important for incision healing and growth. If you have concerns that your child is not tolerating their feedings or formula/breastmilk, discuss this with your pediatrician and your cardiologist.

### Medications

If your baby requires medication at the time of discharge, prescriptions will be sent to your pharmacy. Some medications may need to be compounded (tablets are crushed and made into a solution). This may require extra time and a specialty pharmacy. Before discharge, work with your bedside nurse to make a home-friendly medication schedule. Ask your bedside nurse to show you how to give medications, and practice giving them before you go home.

### Surgical Incision Care

Your baby's chest incision is closed using special stitches on the inside that are later broken down and absorbed by the body and don't need to be removed. Skin glue is used to close the incision on the outside and provides a barrier against infection. In about 7 to 10 days, the skin glue will begin to flake and naturally come off the incision. Do not scratch, pick, or scrub off the skin glue.

There will be stitches where your child had chest tubes. These stitches will need to be removed 7 to 10 days after the chest tube was removed.

Your child can start having baths in the bathtub once the incisions are completely healed and all stitches have been removed. Until then, your child should only have sponge baths, making sure to keep the incisions and stitches dry. Avoid lotions and creams on the incisions and stitches until they are completely healed.

## Home Equipment

After your child's Norwood surgery, you were probably using a baby weight scale to do daily weight checks and a pulse oximeter to check blood oxygen levels. After the Glenn surgery, you will likely no longer need to do daily weight and oxygen level checks.

## Immunizations

Immunizations are important to keep your child healthy but should be not be given for 4 to 6 weeks after the Glenn surgery. RSV is a virus that causes "common colds" but can also cause lung infections or pneumonia in children less than 1 year of age and in children with certain types of heart disease such as HLHS. Your child should receive monthly RSV vaccines or Synagis during the RSV season (November-March). This should be organized through your pediatrician's office.

## Home Care

Be sure to complete CPR training while in the hospital if you have not done so previously. Ask questions about things to look out for that would prompt you to seek medical care for your child. Make sure you know places near your home where you can get emergency medical care for your child. Be sure to get a phone number to call after hours if you have any medical questions or concerns after going home. Try to maintain a normal routine despite the extra care your child may need. Getting outside and interacting with others is generally good. It is important to take care of yourself as well: practice good handwashing, make sure to get a flu shot and other recommended immunizations for yourself and other adults in the house, avoid sick family members and friends if possible, drink plenty of fluids, eat a balanced diet, and get enough sleep.

## Outpatient Follow-up

Your child should be seen by the pediatrician soon after you leave the hospital and by the primary cardiologist usually within 2 to 4 weeks. These appointments are usually made before you leave the hospital. Appointments with other healthcare providers, such as dietitians/nutritionists and physical therapists, may also be made before you leave the hospital.

# KASH



Kash aka “Kash Money” is celebrating his first birthday with a fist full of cake. From a garden gnome to a happy-hearted little king, this little guy is always ready for the next best thing to smile at.

He is courageous, brave and charming!





# 11

## After the Glenn Surgery

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

## What to Expect After Going Home

The time after discharge from the Glenn can be an adjustment for many families.

One of the biggest difficulties for families can be the sense of abandonment (feeling completely alone). Before the Glenn, your cardiology team is in close contact. During that time, communication and visits to the office are occurring frequently. This involvement of the cardiac team in the life of your child and family can become very reassuring. In many cases, families are very close to their cardiology team. They look forward to the frequent contact. After the Glenn however, most children are much more stable. They are also less prone to **adverse events** now. With this, the involvement of the cardiac team generally decreases. There will still be clinic visits, but they are spaced out to a scale based on months instead of weeks. Families are encouraged to contact the cardiology team to discuss concerns. Rather than the cardiology team, you will now take on more responsibility for the day-to-day medical support of your child. At that time, you may feel a new pressure to be completely in charge of your child's care and may even feel alone. Rest assured, this is completely normal.

Most children do better after the Glenn, requiring fewer medications and less monitoring. However, there are other possibilities that might arise at the time of discharge from the Glenn. It is possible that there will be changes to feeding. Some children that may have been able to feed completely by mouth before surgery will now require feeding by a tube. This is a new burden for families. This may involve increased work at home as well as increased specialist involvement and rehabilitation visits. Medications might be different from those required before surgery, which forces families to learn a new routine.

Overall, many children are stronger and with less risk of adverse events after the Glenn. Most will have improved growth and development over the next few years before the final surgical stage, the Fontan.

### Nutrition and Growth After the Bidirectional Glenn

Now that your baby's blood flow is less fragile, and as your baby's feeding pattern changes by age, he or she will also require fewer calories to grow. The average weight gain for your baby will now be about 15 to 20 grams/day. Studies have shown that

babies with single ventricle CHD tend to grow very well in the first few months after the bidirectional Glenn.

## Outpatient Feeding Therapists and Weaning Supplemental Feedings

After your baby has fully recovered from the bidirectional Glenn (usually 4-6 weeks), your baby's feeding team will support you and your baby to start the tube weaning plan, so that your baby will not need tube feedings any longer. They will work closely with your cardiology team so that your baby can start taking all feeds by mouth. Weaning the tube feedings is a process, but this is the main goal for most babies after the bidirectional Glenn.

ARIANNA



*Two days post Glenn*



*One month post Glenn*



Arianna was born full term with HLHS at John's Hopkins Hospital in Baltimore, Maryland. Her Norwood surgery occurred 3 days later. She remained at the hospital for 8 weeks after surgery, mostly due to feeding challenges. Her Glenn surgery occurred when she was 5-1/2 months old. This time around, she was at the hospital for 5 days. As a baby, she loved to observe her surroundings, interact with people, and take baths. She also learned to roll over and crawl, got her first teeth, and started to eat baby food before her first birthday. Now, at 2-1/2, she enjoys attending gymnastics class, playing outside with her sister and friends, coloring, playing with baby dolls, and making people laugh.



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*“The time leading up to the Glenn was a roller coaster of emotions for our family. We had feelings of anxiousness for the unknown, excitement for the possibilities, fear of risks, and hope for the future. Having gone through an open-heart surgery once with the Norwood, we had some insight into what to expect. This could be argued as both an advantage and a disadvantage. Although we experienced similar emotions leading to the first surgery, there were evident differences. We had developed our own sense of normalcy and comfort with Arianna at home, and this was about to change. Our bond with our daughter was much more real now that she was part of our everyday life, as opposed to before the Norwood. On the other hand, we recognized the opportunities the Glenn would allow for our daughter and our family. For us, the hospital stay for the Glenn came with increased familiarity, greater trust, shorter admission time, smoother recovery, less medications, and more enthusiasm for the impending return home.*

*After discharge from the Glenn, our family was able to carry on with life as we hoped. Soon after Arianna’s discharge, my husband and I were back to work, our older daughter returned to her life as she knew it, and our heart warrior was able to just be a baby, growing and developing with her peers. As a family, we attended more functions with our friends and family, went on adventures, and made plans for our future.”*

**Jodi**

MOTHER OF ARIANNA

# 12

## Neurodevelopment: Supporting Your Baby to Reach Milestones

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*With contributions from members of the NPC-QIC Neurodevelopment Learning Lab*

As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

## Will my baby's development be delayed?

- Your baby's development *may* be delayed.
- **Developmental delay** means that a baby is reaching milestones more slowly than is typical. Milestones include being able to reach for toys, sit up, hold a bottle, or talk.
- It is very common for babies with CHD, especially those with single ventricle CHD, to have some delays.
- Although delays are expected, it is **IMPORTANT** that babies with single ventricle CHD get the right help as early as possible. Therapies can help babies work on the areas where they need help. This improves outcomes. **EARLY = BETTER**

## Why is my baby at risk for developmental delay?

- Research shows that even before babies with CHD are born, there are differences in how their brains develop due to their circulation. This puts these babies at risk for developmental delay.
- Babies with single ventricle CHD are born with brains that are less developed compared to heart-healthy babies. At birth, full-term babies with SV have brains that are similar to a baby born at up to 1 month early (35 weeks).<sup>1</sup> Having a less mature brain at birth makes them more likely to be injured.  
<sup>1</sup> Licht DJ, Shera DM, Clancy RR, et al. Brain maturation is delayed in infants with complex congenital heart defects. *J Thorac Cardiovasc Surg* 2009;137:529-36; discussion 36-7.
- Oxygen is important for the developing brain. Babies with single ventricle CHD do not get as much oxygen to their brains as heart-healthy babies.
- While babies are recovering from surgery, they are sedated and immobile on their backs for long periods of time, which does not allow them to move and play normally. As a result, it is harder for them to build up their strength and exercise their muscles.
- They don't get enough tummy time early on.
- Babies with single ventricle CHD may not be able to learn to breast or bottle feed as newborns. See [\*Chapter 7 – Nutrition and Growth in Patients with Single Ventricle CHD from Birth to Fontan and Beyond\*](#) for more information about this topic.

## How will I know if my baby is delayed?

- Ask your baby's primary care doctor. Your baby's doctor should be doing regular screening tests to check if your baby's development is on target.
- Through NPC-QIC you can participate in a developmental screening program where you can fill out the Ages and Stages Questionnaires (ASQ) every few months. You will receive scores to find out if your baby's development is on target. For more information visit [npcqic.org](http://npcqic.org).
- Your cardiac program may have a specialized developmental clinic. At this clinic, a team of experts can monitor your child's progress and suggest ways to help.

## What should I do if my child has delays?

- You are not alone and there are lots of things you can do to help your baby or toddler!
- When children have delays, it is important for them to work with **therapists**. This includes Early Intervention specialists who are experts in helping children reach their milestones.
- Therapists are coaches who give children and their parents exercises to practice together. This helps children get stronger and learn new skills!
- Every state in the US has Early Intervention programs. These programs are usually run by the county where you live. Therapists can come to your home and work with your child from birth to 3 years of age.
- In addition to participating in Early Intervention programs, some children also benefit from private, outpatient therapies such as speech or physical therapy.

## What can I do at home to support my child's development?

- Talk to your child a lot. Sing to your child.
- Make sure your baby gets tummy time every day once your baby's doctor says it is okay.
- Get down on the floor and play with your child.
- Read stories or look at books with pictures with your child every day.
- Try to avoid screen time! Babies and toddlers don't need to watch any television or play with electronic games. They learn best when they can explore objects with their hands and interact with people.

## Types of Therapies Defined

- **Early Intervention (EI):** State-run program, usually run at the county level. They can come to your home and provide services for babies from birth to 3 years of age. They work with children who either have developmental delays or are at high risk for delays. Services can include a developmental interventionist who is a teacher specializing in early childhood development. Other types of therapy include physical therapy, occupational therapy, or speech/language therapy.
- **Physical Therapy (PT):** works on building gross motor skills such as rolling, sitting, crawling, walking, and improving large muscle strength and coordination.
- **Occupational Therapy (OT):** works on fine motor skills such as how a baby uses his or her hands as well as feeding and sensory processing, meaning they can help babies who are sensitive to sounds, lights, crowded places, textures, and touch sensations.
- **Speech/Language Therapy (SLT):** works on feeding and communication. This includes how a baby understands language or follows directions and how a baby communicates to others by pointing, speaking, or using sign language.

TYLER



Our son has had some challenges that he has worked very hard to overcome with outpatient therapies. He had a walker for a few years and wears leg braces, but his positive attitude keeps him progressing. Gross motor skill development has been the most difficult to watch as a parent as he was quite far behind his typically developing peers, but it was helpful to know that other children with single ventricle CHD were also having similar struggles, and that he was not alone.



## Resources

- NPC-QIC website: [npcqic.org](http://npcqic.org)
- Centers for Disease Control and Prevention (CDC): **Learn the Signs. Act Early.** Program website has excellent resources for parents about development, when to be concerned, and what to do if you are concerned about your baby's development: [www.cdc.gov/ncbddd/actearly/index.html](http://www.cdc.gov/ncbddd/actearly/index.html)
- We recommend downloading the CDC's developmental milestones tracker app for you to be able to monitor and follow your baby's milestones: [www.cdc.gov/ncbddd/actearly/milestones-app.html](http://www.cdc.gov/ncbddd/actearly/milestones-app.html)
- Check with your baby's primary care doctor or cardiac center about how to enroll in your state's Early Intervention (EI) program. In many states, parents can call the EI program directly to request an evaluation. However, in some states, babies have to be referred by a professional. Call your state's EI program to learn more. You can find the contact information for the EI program in your state at this website: [www.cdc.gov/ncbddd/actearly/parents/states.html](http://www.cdc.gov/ncbddd/actearly/parents/states.html)

## What to Expect After the First Year

- Children with single ventricle CHD continue to be at high risk for developmental and learning problems after the first year of life. This is true even if there have been no concerns about development in infancy.
- Delays in some areas such as speech are not usually detected until a child is 18 to 36 months of age. Learning and attention problems may not be detected until elementary school.
- Guidelines from The American Heart Association recommend developmental evaluations for children with SV. Evaluations will be scheduled at several time points during childhood and adolescence. Here is a link to the article: <http://circ.ahajournals.org/content/early/2012/07/30/CIR.0b013e318265ee8a>
- Children can receive EI services until age 36 months. Once they are 3 years or older they may be able to attend a local public preschool. Preschool provides enrichment and can provide therapies. Your child's EI team will help with the transition to the school system at age 3.

<sup>2</sup> Marino BS, Lipkin PH, Newburger JW, et al. Neurodevelopmental outcomes in children with CHD: evaluation and management: a scientific statement from the American Heart Association. *Circulation* 2012;126:1143-72.



*“Our family’s journey with single ventricle CHD and neurodevelopment started very early. Our son was not diagnosed until after he was born and transferred to his surgical center, where it was discovered that he was having seizures. It took a number of days to stabilize his brain so that he was able to have his Norwood. Once he had recovered enough to begin therapies as an inpatient, he had the benefit of having the neurology team also helping guide his medical decisions in conjunction with cardiology.*

*Due to his heart and brain injuries, he was automatically referred to Early Intervention and he began home-based therapies as soon as he was home from his Glenn at 5 months old. It was encouraging for us to have a local team of therapists who continued the therapies that he was already receiving during his extended stay in the hospital, and although they had never worked with a single ventricle CHD infant, we felt confident in their abilities because they did an excellent job in communicating with both his primary doctor and surgical center team. Due to the extent of his brain injuries, at 6 months old he was essentially at a newborn level in terms of gross motor skills and was unable to lift his head, move his arms with purpose, or put any weight on his legs despite daily hospital therapies.*

*Through Early Intervention, he received feeding therapy, infant development therapy, physical therapy, and occupational therapy, all weekly in our home, and when he was 11 months old, we added speech therapy as he was not yet making typical infant sounds. Our community also hosted a weekly playdate for infants and toddlers in Early Intervention, which he enjoyed immensely, and it was a nice place for us to connect with other parents with children who needed extra help. Progress was slow, but measurable, and by 18 months our son was able to sit up reliably and was reaching for toys and showing interest in extended*

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*time on his tummy. He was given a walking frame and leg braces to help him explore the world around him at age 2, and started in a toddler classroom similar to preschool through Early Intervention. He was highly motivated by sports and being outside, so we tried to give him as many natural exposures to these as possible, as well as regular playdates with neurotypical friends so that he could work towards goals. We had regular follow-ups with his cardiologist, neurologist, and pediatrician, as well as working with a High-Risk Infant development team to help check that he was progressing at a pace that was expected for his trajectory.*

*By the time he was 3 years old, he was walking independently and beginning to use words and sign language to communicate, and we know that neither of these would have been possible without the multiple therapies he was receiving each week. He began at a special education preschool at age 3, and while he did take a few weeks off to have his Fontan at age 4 from school, he has thrived on the routine and consistency of public school and school-based therapies in addition to outpatient therapies.*

*He is currently in first grade, and is in a mainstream classroom, with a few school therapies to help him progress. We are very fortunate that his cognitive skills seem to be on par with his friends; he is motivated to learn adaptive skills, and we have a long-term single ventricle CHD follow-up team that monitors every aspect of his growth that is not limited to just his heart, but his entire well-being and progress.”*

**Jennie**

MOTHER OF TYLER



# 13

## The Fontan: Surgery and Initial Postoperative Course

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This section reviews the third **palliative** surgery for patients with single ventricle CHD, the Fontan surgery, and what to expect at the time of surgery and in the months and years after the Fontan.

**TYLER**



Seeing our son standing with his portrait allows us to reflect on how far Tyler has come since birth. The photo was taken contrasting the day of his Norwood, to post-Fontan and standing tall.

## The Surgery

Fontan completion is the final surgery for single ventricle heart defects. The connection created at the Stage 2 Glenn surgery directed the **blue** blood from the upper body directly to the lungs. At the time of Fontan completion, the surgeon creates a special pathway for **blue** blood from the lower body to go directly to the lungs without passing through the heart (*see Figure 5*). With the Fontan completion, almost all of your child's **blue** blood flows directly to the lungs without the help of a pump, and your child's single ventricle pumps the **red** blood to the body. Because the pathways for the **blue** blood and **red** blood are separated, your child's oxygen saturations should be near normal (90s).

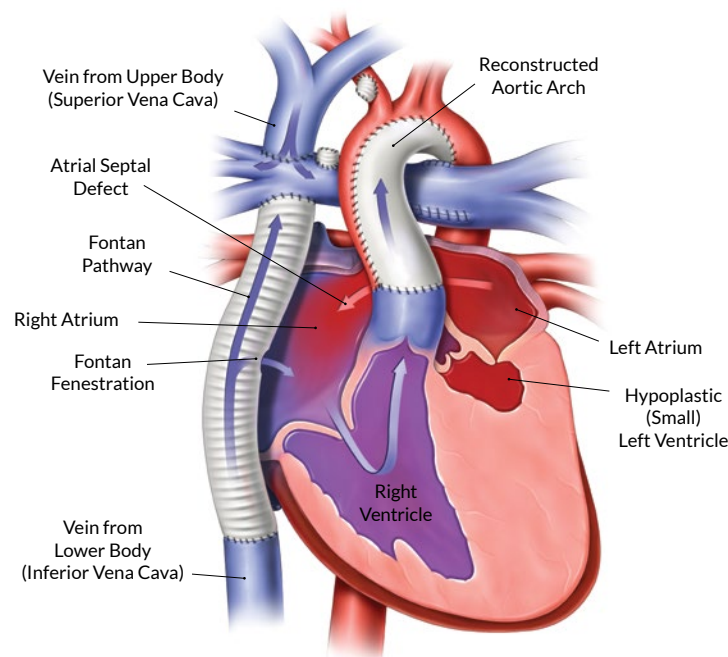
Before the Fontan surgery, almost all patients with single ventricle CHD will undergo a pre-Fontan cardiac catheterization to check for any problems that would make it difficult for blood to flow to the lungs without a pump. These would include any narrowings (stenosis) in the in the blood vessels carrying blood to the lungs (pulmonary arteries) or

in the blood vessels carrying blood back to the heart from the lungs (pulmonary veins). A catheterization also confirms that the pressure inside the heart chambers is low and that the blood vessels in the lungs are relaxed, so that there will be low resistance to flow through them after the Fontan.

There are different kinds of Fontan connections. An **intracardiac tunnel** (also called a **lateral tunnel** or **intracardiac baffle**) passes through the heart and uses the wall of one of the top chambers of the heart (atrium) as part of the tunnel. The **extracardiac conduit Fontan** uses a tube that passes outside of the heart to send blood from the inferior vena cava (IVC) (blue blood from the lower body) to the lungs (main pulmonary artery). There are also Fontan tunnels which may be created at the time of cardiac catheterization. The choice of Fontan type for your child will be based on features of his or her heart anatomy and the judgment and experience of your child's cardiology and surgery teams.

The Fontan pathway may be a tube or a tunnel for blood to flow from the IVC to the lung blood vessels (pulmonary arteries). Many children will have a **fenestration**, or small hole or connection between the Fontan tunnel and the heart. This connection is a "pop-off," which may lower the pressure in the tunnel. Fenestrations may decrease the time your child has fluid collecting around the lungs (**pleural effusions**) after surgery. Because this

Figure 5 – Fontan Surgery



connection also allows a small amount of the [blue](#) blood to bypass the lungs and return directly to the heart, fenestrations lower the oxygen saturation. At some centers, the fenestrations are closed in later childhood during another cardiac catheterization. Other centers prefer to leave fenestrations open.

## Postoperative Course

During the time period following the Fontan surgery, your child will require medications, at least temporarily. There is an increased need for diuretics, which are medicines that help your child pee out extra fluid in their urine. This is: (1) because of the changes in your child's blood flow with more blood going to the lungs through the Fontan tunnel; and (2) because the heart-lung bypass machine was used for your child's Fontan surgery. In the weeks after surgery with the heart-lung bypass machine, children and adults tend to hold on to fluid.

Your child will likely have one or more chest tubes after the Fontan, just as they did after the Norwood and Glenn. Chest tubes help prevent fluid from collecting in the space around the lungs (pleural effusion). Diuretics and other medicines may decrease the time your child will need a chest tube after surgery). Rarely, the chest tube becomes milky in appearance and is high in fat. This is known as a chylous effusion. Children with chylous effusions may have a longer period of chest tube drainage and may be placed on a low-fat diet to help decrease the chest tube drainage.

The amount of medicines your child needs will usually decrease in the weeks and months after surgery.



# 14

## Long-Term Fontan/ Single Ventricle CHD Outcomes

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## Follow-Up After Discharge

Your child will continue to need life-long follow-up with their cardiologist after the Fontan. You may also meet other caregivers important in your child's care including psychologists, developmental specialists, social workers, and liver specialists.

When you visit the cardiologist, your child may have tests to assure that his or her heart is working well after surgery. In early childhood, your child's visits will include tests you already learned about at the time of your child's first and second surgeries: electrocardiograms, echocardiograms, and saturation checks. In later childhood and the teenage years, **exercise (stress) tests**, **Holter monitor** evaluations, MRI studies and CT scans may be added. Because the Fontan changes the way the blood from the lower body travels to the lungs, there can be changes in blood flow in other parts of the body, including the liver. In later childhood and as an adult, your child will also need imaging tests and blood tests to check the health of his or her liver.

WYLEY



This is Wyley. He is 8 years old and 5 years post-Fontan. He loves the outdoors, swimming, riding his bike, and video games.

Your child's follow-up visits will also include checking for problems in development and in learning. In addition to the developmental concerns in the previous chapter, children who have had heart surgery are also more likely to have attention-deficit/hyperactivity disorder (ADHD) and problems in school. Your child's care team may suggest testing and special therapies and services to help with development and learning, if needed. If your child needs extra help in school, your care team can help you find the people in your school district who can work with you to get your child the help and support he or she needs.

Your child's cardiologist will ensure that your child gets all the care needed during childhood and adulthood. Some centers have special Single Ventricle CHD Clinics to help with all the services needed in your child's care. Your child's care will likely be moved or transitioned to an **adult congenital cardiologist** during his or her teenage years. Adult congenital cardiologists have specialized training to care for adults born with abnormal hearts and are experts in ensuring that they continue to receive the specialized care they need in later life.

## Medications

The number and type of medicines each child needs after the Fontan may be very different. After the Fontan, your child will require a daily medicine to prevent blood clots from forming in the special pathways that direct blood to the lungs. This may be an **antiplatelet agent** like aspirin, or a stronger blood thinner or **anticoagulant** like **Coumadin (warfarin)**. Many children will remain on diuretics like **Lasix (furosemide)** or **hydrochlorothiazide** long-term. Some may be on medicines like **digoxin** to help with heart function. Some children may take **ACE inhibitors** or **spironolactone (Aldactone)**, which may be helpful to the heart in the long-term. A special type of medicine called a **pulmonary vasodilator (eg, sildenafil, tadalafil)** may also be helpful for some children after the Fontan. Some children may have abnormal heartbeat patterns called arrhythmias and may need special **antiarrhythmic medicines** for these. Your child's cardiologist will choose the best medicines for your child.

## Nutrition and Growth – Fontan and Beyond

After the Fontan, some children will grow and develop normally, while others may need additional support. At the time of the Fontan, some children may still require extra nutrition, such as higher calories and protein supplements or tube feedings. Oral motor

skills, and behavioral feeding therapy may be ongoing in the Fontan patient if feeding difficulty remains present. The goal after the Fontan procedure is to encourage and maintain healthy eating habits as these children grow into adults.

A well-balanced diet should include fiber and whole-grain foods, the right-sized servings of all food groups, including intake of fruits, vegetables, and proteins. Eating more high-fat foods than needed should be avoided, and added salt and high-salt foods should be limited (this may be particularly important for some children after Fontan). Several possible long-term complications can develop any time after the Fontan procedure that may require dietary changes in order to support the blood flow created by the Fontan surgery.

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**JACK**



Jack was born with double inlet left ventricle, transposition of the great arteries, a hypoplastic aorta, interrupted aortic arch, and a ventricular septal defect (VSD). He is now almost three years old and loves the going to the playground, pretending to be a fireman, and playing with his cousins. His favorite foods are pizza and grapes.

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## Exercise

After the Fontan, your child will not be able to exercise quite as well as children with two-pump hearts. This is because your child will not have a pump to help increase the flow of blood to the lungs when they exercise (they have passive blood flow to their lungs).

Staying active, with regular exercise, is important for everyone whether they have a two-pump heart or a Fontan. Children and adults who stay active after the Fontan have

better exercise ability. After the Fontan, your child will not have any limits in recreational activities. Your child can play as actively as he or she would like at home or school or in local recreational leagues where children can choose to play at their own pace. They may need to rest more often than others and may prefer sports like baseball or volleyball.

Your cardiologist may check your child's ability to exercise with an exercise (stress) test. Current guidelines allow some children after the Fontan to participate in competitive sports in middle school and high school after evaluation by their cardiologist. Your child may be able to participate in other competitive sports after testing and discussion with your cardiologist.

## Long-Term Medical Complications

Most children and young adults after the Fontan do well during the first 20 years after surgery. We have less information after this time, because we are only now caring for large numbers of adults who had Fontan surgeries as children. The surgeries done now for HLHS have only been performed for about 30 years. We do know that 20% to 30% of patients will have significant problems in the first 20 years after surgery that lead to death or the need for heart transplantation. As your cardiologist cares for your child, he or she will be checking for any of these important problems and the best way to treat them. These include:

- Decreases in heart function which may be treated with medications or heart transplantation.
- Abnormal heartbeat patterns (arrhythmias) which may need treatment with medicines, a pacemaker, or a procedure called **ablation**.
- Blood clots that form in Fontan pathways even when medicines are taken to prevent them.
- A rare infection of the heart or blood vessels called **endocarditis**. Your doctor will teach you and your child about how to prevent this with good dental care and how to watch for signs of this infection.
- Rare conditions called **protein-losing enteropathy** and **plastic bronchitis** that may occur months or years after Fontan. In protein-losing enteropathy, patients have difficulty absorbing protein from the gut. Plastic bronchitis is a condition where abnormal **lymphatic** flow in the lungs causes the patient to form plugs or "casts" in their airways, which they may cough up or swallow and which may interfere with their lung function. Treatments for these conditions continue to advance. Some patients will require heart transplantation.

- An increased risk of liver disease in adulthood. It is not clear how many will have important liver disease. This is an area of ongoing research.

There may be some differences in the likelihood of long-term problems depending on whether your child has a single left ventricle or single right ventricle.

## Heart Transplantation

Heart transplantation may be the best treatment for children and adults who are not doing well after the Fontan. **Advanced cardiac therapy (ACT) specialists** and members of the heart transplantation team may be asked to meet with children and families after the Fontan to help decide whether heart transplantation would be helpful for them. Children and adults who undergo heart transplantation require daily medicines to prevent the body's defenses from damaging the new heart (**rejection**), as well as other medicines to prevent infections. Heart transplantation is not a complete cure in that it comes with a different set of issues and problems than single ventricle procedures, but for some, a heart transplant is the best treatment option.

# 15

## Common Cardiac Tests

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The following is a list of common cardiac tests that be may performed on your child to assess their heart and medical progress. Some tests are done more frequently than others. Some tests may require your child to be put to sleep either with oral (by mouth) sedation or general anesthesia, while others may be quick and easy and completed while your child is awake. Medical tests help guide your child's team to provide the best medical care and treatment plan. If you have any questions about testing, be sure to ask your care team. They will be happy to answer any of your questions.

## Electrocardiogram

An ECG, EKG, or electrocardiogram is a quick test that records the electrical activity of the heart. An electrocardiogram helps identify heartbeat or rhythm abnormalities. It can also provide information about the size or thickness of the heart chambers and the position of the heart in the chest.

The ECG usually takes about 5 minutes and is painless. Typically, 12 stickers, also called leads, are placed on the chest and each arm and leg, and the electrical activity of the heart is recorded from each of those positions; the stickers are then removed.

## Chest X-ray

The chest x-ray is the most common radiologic procedure. X-rays (a form of radiation) are projected through the chest to show the heart, lungs, bones, and soft tissues, and an image of those body structures is created. The actual time of the average x-ray exposure is extremely short, often less than a 1/2 second and is very much like taking a picture with a camera. The chest x-ray in young children usually involves looking at two views. The first view is from the front, and the second is a side view.

A chest x-ray usually takes about 10 minutes. There is no pain involved, but your child may need to lie on a hard surface while the images are being taken.

## Echocardiogram

An echocardiogram or "echo" is an ultrasound test that uses high frequency sound waves that are non-radioactive to view the heart. It is a safe and painless procedure



that helps doctors diagnose heart problems and understand the anatomy of the heart. Pictures of your child's heart can be viewed on a small monitor while the procedure is being performed. It is a non-invasive test, meaning no probes or needles are used, and everything is done from the outside of the body. An echocardiogram is similar to a sonogram or ultrasound that many women have had before a child was born; however, the echocardiogram focuses specifically on the heart and blood vessels around the heart. The echocardiogram is usually performed without sedation, but sometimes if more detailed information is required, a sedated study may be necessary.

An un-sedated echo takes approximately 1 hour. If a child is sedated for the study, he or she will need to stay until the nurse or doctor feels the child is ready to leave the hospital.

**ADELINE**



Our heart warrior, sitting through an echo at 4 months old. She will continue with periodic echos after her third surgery, the Fontan.



Sleeping soundly after a heart catheterization, Adeline will repeat this procedure before each open-heart surgery.

## Cardiac Catheterization

Cardiac catheterization or "cath" is a procedure, that requires placing a needle into an artery or a vein, most often in the groin area. A small, flexible tube called a catheter is

then guided from the groin area into the heart and blood vessels. The catheter is guided with the aid of an x-ray machine and contrast dye that is injected through it. This test is done to identify heart problems or to confirm a cardiac diagnosis. During a cardiac catheterization, your child's medical team can also perform procedures that will help your child's heart. **Interventional catheterization** is a type of cardiac catheterization in which these procedures and treatments are completed. Catheters include balloons that can open up small valves or arteries. Your child's medical team can also use devices that may open or close holes in the heart or close extra vessels that are not needed. Often, babies are admitted for IV fluids the night before their catheterization or early the same day.

A cardiac catheterization may take up to a few hours to complete, not including the recovery phase. After the catheterization, your child may be taken to the cardiac intensive care unit (CICU) where he or she will be observed. Your child should go home the same day or the following day.

## Cardiac MRI

MRI stands for magnetic resonance imaging. An MRI machine uses a powerful magnet, radio waves, and a computer to acquire pictures of just about any body part, including the heart. An MRI study uses no radiation. An MRI is a non-invasive way to evaluate the heart's function and structure. Sometimes a contrast agent, which is a liquid material, is injected before MRI pictures are taken, to make the blood vessels and heart easier to see.

To secure good pictures or images, it is important for the patient to lie very still during the scan. For younger children or infants, it may be best to use sedation or anesthesia. If sedation is required for your child, IV medications or general anesthesia may be used, and he or she will need to stay until the nurse or doctor feels the child is ready to leave the hospital. If your child is an infant, it may be possible to get the images while they are sleeping. Because the MRI does make loud sounds, such as clicks and bangs while the images are being taken, earphones may be helpful.

A cardiac MRI may take up to 1 to 2 hours to complete. If a cardiac MRI is completed as an outpatient procedure and no sedation is used, your child may go home after the scan is complete. If sedation is used, your child will need to be monitored closely after the procedure, and may need to stay overnight. You will also be asked to avoid having any metal near the child in the MRI suite because it is a large magnet that can attract the metal.

## CT Scan

A **computed tomography (CT) scan**, also called a CAT scan, uses x-rays and a large camera to take pictures of the inside of the body. For some scans, the doctor may ask for contrast. A contrast dye is given either by mouth or through an IV line. The scanner has a large donut-shaped opening and a bed the patient lies on that can move in and out. The CT scan makes a humming noise as it scans the patient. The CT scan does not hurt, nor does it touch the patient. To secure good pictures or images, it is important for the patient to lie very still during the scan. For younger children or infants, it may be best to use sedation or anesthesia.

A CT scan may take 10 to 30 minutes. Some CT scans may even be done at a patient's bedside. If a CT scan is completed as an outpatient procedure and no sedation is used, your child may go home after the scan is complete. If sedation is used, your child will need to be monitored closely after the procedure.

## Holter Monitor

A Holter monitor or heart monitor is a device that records the heart rate continuously. It can be applied for 24 or 48 hours. It is generally attached to your child during an outpatient visit. Your child will have three or four small leads or stickers applied on the chest which are connected to the monitor by thin flexible plastic cords. The monitor is a small-sized handheld device. While the monitor is applied, your child may be asked to not take a bath. The nurse in the clinic will explain in detail about the monitor and how to take it off. After 24 to 48 hours, you will take the monitor off and return it to your cardiologist's office. There are no side effects of the monitor. It is painless and does not deliver shock or current. It helps your cardiologist identify abnormal heart rhythms. It is generally performed at 6-month to 1-year intervals, depending upon your child's age and heart condition. If your child requires monitoring for more than 24 or 48 hours, your doctor may suggest a Zio Monitor, which is similar to a Holter monitor but is smaller, has no leads or wires, and can be worn for up to 14 days.

## Cardiac Stress Test

A stress test is designed to evaluate performance of the heart during activity, to look for an abnormal heart rhythm during exercise, to know baseline exercise capacity, to guide exercise rehabilitation, to know lung function during the exercise, and to rule out

exercise-induced asthma. It can be combined with an echocardiogram or other advanced imaging of the heart to look for heart wall motion problems and evaluate blood supply to the heart muscle during exercise.

Different forms of stressors can be used. In children, the majority of the time, mechanical stressors like a bicycle or treadmill is used. Rarely, medications are used to do stress tests when a child cannot otherwise perform mechanical stress testing. A mechanical cardiac stress test is usually performed as an outpatient procedure. It takes about 2 hours. Your child may be asked to avoid heavy meals before the test and to bring sports shoes. During the test, you may not be allowed to be with your child. However, your child will be constantly monitored by a cardiologist, certified registered nurse, and stress technician throughout the test. If the stress test is performed specifically to evaluate an abnormal heart rhythm, your child may be required to get an IV line.

During the stress test, your child will be connected to a heart monitor that shows the heart's electrical activity as well as your child's blood pressure and oxygen saturation levels. If lung function is being tested, then a mouthpiece will be given to your child to breath into throughout the test. The speed of the treadmill or bicycle is gradually increased until your child can no longer continue. After the test, your child will be monitored during a recovery period of 15 minutes.

Minor risks include fall, injury, dizziness, and fainting. Rarely, a stress test may result in an abnormal heart rhythm. That is why it is done in a controlled setting with all measures of care available. Your child's cardiologist would decide what type of stress test is required and how often. For children who have had the Fontan surgery, additional specialized test may be needed, and your cardiologist will also decide if and when these may be necessary.

## Alpha-1 Antitrypsin (stool test)

With Fontan circulation, blood from the body returns to the lungs passively, without going through the heart. Over a period of time, this has shown to increase pressure and fluid in the liver and gut. In some patients, this can cause the body to start losing protein. Those patients develop limb swelling, diarrhea, and fluid collection in the belly. **Alpha-1 antitrypsin** is one such protein that starts showing up in the stool when the body starts losing protein. Thus, its presence and level help your cardiologist start and maintain treatment of protein loss. The test is generally done on a yearly basis.

## Liver Testing

**Liver function tests** (measured through a blood sample) and liver **biopsy** (taking a sample of the liver with a needle): As noted above, with Fontan circulation, blood from the body returns to the lungs passively, without going through the heart. Over a period of time, this has shown to increase pressure and fluid in the liver and gut. This may increase the blood level of liver **enzymes** and decrease the liver's production of proteins. Liver function tests are done yearly for surveillance. Additionally, after a long period of time, Fontan circulation may cause changes in liver tissue. So, a liver biopsy may also be done to monitor liver tissue every 4 to 5 years. It is generally done along with cardiac catheterization under sedation. A small needle is introduced into the liver and a tiny piece of tissue is taken out to be examined microscopically. It takes 3 to 4 weeks for the results to come back.



*“I’m used to appointments and don’t get nervous. I like having echos because it feels neat. I used to watch a movie during it but now that I’m older I like to look at the screen and see my heart.”*

**Avery**

AGE NINE

# 16

## Medications

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

Your child may be sent home on daily medications; there are various types of medications used in CHD, and for different reasons. It is important to understand your child's medications. We have described some of the medications and their effects below. In general, when your child is started on a new medicine, learn from your medical team what the medication does, how it works, what side effects may happen, and how much your child should take (liquid amount, number of times per day). It will be helpful to keep track of all of your child's medications. You can write them out on paper or you can use an app on your phone; you may ask your medical team for help. Please make every effort to not miss the medication dose.

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**ARIANNA**



Arianna's sister  
Joslynn giving her  
"medicine" during  
the first few days  
post-Glenn

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For children, some prescriptions need to be compounded (made into liquid); and not every pharmacy can do this for all medicines, so plan ahead and call to make sure they can provide your child's specific medications. The pharmacist can help explain how to store and give the medications properly. For refills or questions about the medications, contact your child's primary cardiologist. If you ever run out of medication, call the primary cardiologist's clinic for the prescription to be sent to the pharmacy. Please do not wait to schedule a clinic visit for medication refills. In case of a medication overdose, bring your child to the nearest emergency room and inform your child's primary cardiologist.



## Aspirin

### How does it work?

Aspirin is an antiplatelet agent. **Platelets** are parts of our blood that are important for forming clots at times of bleeding. Aspirin is one of several drugs called “blood thinner” medications. It prevents platelets in the body from sticking together to form clots in the blood vessels, shunts, or stents placed during procedures or surgery. By preventing clots, blood can flow freely through the blood vessels, shunts, and stents to the rest of the body.

### What dose of medication will my child take?

The dose of the medication will depend on your child’s body weight. Your cardiologist will tell you the right dose for your child. Aspirin is available as an 81 mg tablet and is given once daily. Depending on the size of your child, you will give  $\frac{1}{4}$  or  $\frac{1}{2}$  tablet. The tablets may be crushed and mixed with a small amount of liquid to give to your child.

### What are common side effects?

Stomach upset, heartburn, blood in stool, bruising

## Digoxin (e.g., Lanoxin, Digitek)

### How does it work?

**Digoxin** is a medication that affects how the heart pumps. By controlling how fast and how hard the heart beats, it allows the heart to pump more blood to the rest of the body with each heartbeat. Digoxin is made from the foxglove plant and is an old drug that has been used for many years for **congestive heart failure** as well as control of heart arrhythmias (abnormal heartbeats).

### What dose of medication will my child take?

The dose of the medication will depend on your child’s weight. Your cardiologist will tell you the right dose for your child. Digoxin is given once or twice daily. It is available as a 0.05 mg per mL solution and is available at all pharmacies. Careful attention to dosing is required as overdose of digoxin is possible, usually due to a toddler or child finding and swallowing an adult’s pill.

### What are common side effects?

Dizziness, drowsiness, blurred vision, irregular heartbeat, stomach upset

## ACE Inhibitors – Captopril, Enalapril, Lisinopril (e.g., Capoten, Vasotec, Zestril)

### How does it work?

These three related medications are blood pressure lowering medications and differ mostly in the way of dose frequency (*captopril* is given three times daily for young infants, *enalapril* twice daily for older infants and children, and *lisinopril* once daily for adolescents and young adults). These medications widen the blood vessels in the body. This decreases the resistance the heart is pumping against, lowering the blood pressure and making it easier for the heart to deliver blood to the rest of the body.

### What dose of medication will my child take?

The dose of the medication will depend on your child's blood pressure and weight. Your cardiologist will tell you the right dose for your child. Enalapril is given two times daily. It is available as a 1 mg per mL suspension made by your pharmacist.

### What are common side effects?

Cough, dizziness, drowsiness, difficulty breathing or swallowing

## Enoxaparin (e.g., Lovenox, Clexane)

### How does it work?

*Enoxaparin* is a blood thinner or anticoagulant. It is a form of *heparin* that is used to help the body break down any blood clots (*thrombus*) that have formed in the circulation. These medicines also help prevent other blood clots from forming in blood vessels that are at high risk for forming blood clots, such as artificial shunts (like a BTT shunt).

### What dose of medication will my child take?

The dose of the medication will depend on your child's body weight and other factors. Your cardiologist will tell you the right dose for your child, and measuring the activity of the drug on your child's clotting system is often necessary to ensure the correct dose is

being given. Enoxaparin is usually given two times daily as an injection under the skin (*subcutaneous injection*).

### What are common side effects?

Pain due to multiple injections, increased bruising, unwanted bleeding

## Furosemide (*Lasix*)

### How does it work?

**Furosemide (*Lasix*)** is a **diuretic** medication, often called a “water pill.” It is used to decrease fluid that builds up in your child’s body by acting on the kidneys to increase the amount of urine your child pees out. Sometimes fluid can build up around the heart and lungs, which makes it more difficult for your child to breathe. Furosemide prevents this from happening by reducing fluid in the body.

### What dose of medication will my child take?

The dose of the medication will depend on your child’s blood pressure, amount of fluid present, and body weight. Your cardiologist will tell you the right dose for your child. Furosemide is given one to four times daily. It is available as a pre-mixed 10 mg per mL solution and is available at most pharmacies.

### What are common side effects?

Abnormal electrolyte levels in the blood, increased urination, thirst, weakness, muscle aches or cramps

## Palivizumab (*Synagis*)

### How does it work?

Palivizumab (*Synagis*) is an injection of **antibodies** given monthly to help protect high-risk infants from severe respiratory syncytial virus (RSV) infection during RSV season. RSV is a harmful common cold virus that infects a child’s lungs and breathing passages, and most children will have been exposed to RSV by 2 years of age. Although palivizumab will not prevent RSV, it can help decrease the how sick your child becomes if he or she does contract RSV.

### What dose of medication will my child take?

Palivizumab is a shot, usually given in the thigh once a month during RSV season (winter months, usually starting in November and continuing through March). Your child should receive the shot every month, even if they have breathing symptoms or an infection. You should contact your doctor with any breathing symptoms, such as a cough, cold, runny nose, or difficulty breathing.

### What are possible side effects?

Low fever, cold-like symptoms, pain, redness or swelling at the site where the shot was given

## Beta-Blockers – Propranolol, Metoprolol, Nadolol, Atenolol (*e.g., Inderal, Lopressor, Corgard, Tenormin*)

### How does it work?

This group of medications (called **beta-blockers**) is used to control how fast the heart beats. Sometimes the heart beats too fast or the beats become irregular. A beta-blocker prevents this from happening.

### What dose of medication will my child take?

The dose of the medication will depend on your child's blood pressure, heart rate, and weight. Your cardiologist will tell you the right dose for your child. Propranolol is given two to four times daily.

### What are possible side effects?

Slower heart rate, drowsiness, weakness, sleep disturbance, cold hands or feet

## Pulmonary Vasodilators – Sildenafil, Tadalafil (*e.g., Revatio, Adcirca*)

### How does it work?

These medications (called **pulmonary vasodilators**) are commonly used in children when lung pressures are high. They work by relaxing the muscles of the lung arteries which,

reduces the resistance that the heart needs to pump against so that blood flow can more easily pass through the lungs.

### What dose of medication will my child take?

The dose of the medication will depend on your child's blood pressure, weight, and degree of increased lung pressures. Your cardiologist will tell you the right dose for your child. Sildenafil is given three to four times daily. It is available as either 2.5 mg per mL or 10 mg per mL solution and is available at all pharmacies.

### What are possible side effects?

Flushing, dizziness, diarrhea, blurred vision, headache



Pumps help staff to give various medications and flushes that need to be delivered through IVs. Patients are often on various medications to help with heart pumping, blood pressure, and pain.

## Warfarin (Coumadin)

### How does it work?

**Warfarin** is an anticoagulant or blood thinner. As part of the normal blood clotting process, the liver makes proteins called **clotting factors** that are released into the blood. Some of these clotting factors need vitamin K to work properly. Warfarin reduces these vitamin K-dependent clotting factors in the blood and keeps the blood “thin” and less likely to clot.

### What dose of medication will my child take?

The dose of the medication will depend on your child’s heart condition. Different degrees of blood thinning are required in different heart conditions. Degree of blood thinning is measured by a blood test called the **INR** or **international normalized ratio**. Your cardiologist will tell you the right dose for your child depending upon what the range that his or her INR should be in. Frequent blood testing of INR will be required while being on warfarin. Warfarin is given once daily. It is available as 1, 2, 3, 4, and 5 mg tablets at all pharmacies.

### What are possible side effects?

Bleeding and bruising. Intake of vitamin K should be avoided at all cost while being on warfarin. Certain types of food like green leafy vegetables, brussels sprouts, cabbage, spring onions, etc. contains large amounts of vitamin K. Please discuss with your cardiologist which foods should be avoided.

# 17

## Pediatric Palliative Care

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As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

## What is pediatric palliative care?

The goal of pediatric palliative care is to improve the quality of life for patients and their families by helping with physical, social, psychological, and spiritual needs. This program works directly with your child's doctors to create a team that supports you, your child, and your family on this special journey.

## What does a palliative care team do?

This partnership of care can be provided during hospital stays and/or used in everyday life. For example, we can provide pain management for your child or tips on reducing stress for you and the rest of your family. We can suggest ways to talk with your child, your child's brothers or sisters, family members and friends about your child's diagnosis, treatment, and other sensitive topics. We can also help you talk with your child's doctors about important decisions. We can help you be confident in your knowledge and comfortable with the future.

## About Your Child

We consider your child's role as a patient, but also his or her role outside of the hospital setting. Your child has meaningful roles as a loved child, sibling, grandchild, and an overall vibrant being! We can share suggestions for coming together as a strong, resilient family. We want to know your hopes for your child and explore your family's concerns about the underlying diagnosis and treatment plan. Also, we want to know your larger hopes and goals, and we want to help you get there. And some day, we hope your child will tell us about his or her own hopes and dreams!

## About You

It has been proven that when parents and family caregivers take care of themselves, they can better care for their child. Palliative care teams can help families design family care plans such as walks within the hospital, journaling, adjusting to emotions, accepting



support, or developing relaxation techniques. By taking care of YOU, you are helping your child. Remember: be gentle and take good care of yourself and each other.

## **Now what?**

Please talk to your child's doctor to learn more about palliative care or to meet the palliative care team. We are available to support your child, your family, and your medical team as an additional layer of support.



“Avery is the youngest of four girls. Our first family vacation was Avery’s Make-A-Wish trip to Florida when they were 6, 11, 15, and 16 years old. Family time has always been very important to us, but the time and experiences we had together that week, away from home, were more than we could have ever imagined. We returned home the day before Thanksgiving and 2 days before the busiest shopping day of the year. It was hard to get into the holiday spirit because we ALL knew we’d NEVER open another present that would compare to what we had just experienced. A few weeks later, the six of us decided not to buy gifts and start saving for a vacation instead! We made saving a priority, and for the next 3 years, they each got a small gift in their stockings and a small gift on their birthdays. This photo is from our trip to Mexico where we said Merry Christmas and Happy Birthday to each other every day of the week we spent there. The day after we came home, the girls came to us and thanked us, said they didn’t miss receiving gifts, and were already excited for our next vacation even though they knew it would take years to save for. I don’t think anything will ever compare to our first family vacation, but we are all committed to the bond we strengthen while we are away; and that is greater than any gift ever wrapped.”

### Anne Dee

MOTHER OF AVERY

# 18

Single Ventricle

Q & A: A Guide to

Communicating with

Your Child's Care Team

*Prepared by NPC-QIC and Sisters by Heart*

## Suggested Items to Bring to the Hospital

This information, prepared by NPC-QIC and Sisters by Heart, is available in booklet form online via this link: [https://npcqic.org/sites/default/files/resources/resources/SV\\_Q%26A\\_Booklet.pdf](https://npcqic.org/sites/default/files/resources/resources/SV_Q%26A_Booklet.pdf)

### MOM & DAD

- SBH Tote Bag/Care Package
- Journal/Pen (*might want to bring two— one for you to journal thoughts/draft blog posts and one to document everything for our HLHSer*)
- File Folder for important documents and handouts
- Electronic chargers & USB cords (phone, camera, laptop, iPad, e-reader...)
- Books to read/e-reader
- Camera
- Headphones
- Magazines
- Crossword Puzzles
- Scrapbook kit (papers, glue, scissors, tape)
- Change of clothes
- Flip flops/slippers
- Sweatshirt (*it can be cold in the hospital!*)
- Sports Bras/Nursing Bras
- Lasinoh (*nipple cream for pumping moms*)
- Personal Breast Pump
- Toiletries
- Dry Shampoo
- Extra sanitary pads
- Tylenol/Ibuprofen
- Chapstick
- Hand Lotion (*Recommendation: Gold Bond Hand Sanitizer – the constant use of antibacterial hand gel can dry out your skin so it's nice to have an alternative*)
- Insulated Cup (*keep hydrated!*)
- Tea Bags, Instant coffee or Starbucks VIA sticks
- Cash for coffee, vending machines...
- Takeout menus/phone numbers for nearby restaurants
- Non-perishable snacks/Gum/Mints
- Pillow or your own pillow cases (*hospitals usually provide pillows/blankets*)
- Cord blood collecting kit

### PATIENT

- Boppy Pillow (*and extra covers*)
- Pictures/decor (*to hang on warming bed/crib*)
- Baby legs (*leg warmers*)
- Side snap onesies
- Hats/beanies
- Baby socks/booties
- Baby nail clippers or baby mittens
- Lovie/Wooby/special toys that can comfort them & be taken to surgery (*recommended to have mom sleep with these items a few times so they smell like her*)
- Soothing music player (*toy or CD player*)
- Wubbanub binky
- Crib sheets/blankets/swaddlers (*hospitals supply these but it's nice to have your own too to make it extra cozy for your baby*)
- A bag to keep by their bed for infant items (*and dirty clothes/blankets*)
- A book/poster board/autograph frame for nurses, docs & visitors to leave well wishes
- Healing Helpers (*healinghelpers.net*) check out the "Zip-Flight" option where you can send your own plush in "for surgery"!

### SIBLINGS

- Books
- Drawing/Coloring supplies (*they will love making things for their baby sibling's room!*)
- iPad/iPhone (*get some iTunes gift cards to give them once you get to the hospital and throughout your time there. Nice way to reward "good behavior" and keep them from getting bored with their existing apps*)
- Snacks/Lollipops/Drinks that don't need to be refrigerated
- A special gift for them (*new teddy bear/doll*)
- Extra clothes/slippers/pj's
- Blankets
- Sweatshirt

We hope these questions prepare you for the medical journey ahead and help foster a good, working relationship with your child's medical team.

## Discovering Your Child Has a Heart Defect is Shocking... Words of Wisdom We'd Like to Share

- Sometimes you have time to research, sometimes you don't. Remember, not everything you read on the internet is accurate. Discuss findings and concerns with medical professionals.
- Each child is different, each family is different; only you can decide what is best for you and your baby.
- The suggested questions in this booklet may be overwhelming – don't feel pressured to ask them all at once, and don't feel you need to ask them all.
- Information regarding your child's care can be confusing, especially numbers and data. Ask your center to explain their numbers and data in a way that you can understand.
- Communication with your child's care team is important. Be open and honest about your feelings and expectations for your child.
- Remember that your child's medical team is there to help you and your child; they care about your family's well-being.
- If you're concerned about the care your child is receiving, you may get a second opinion from another doctor or cardiac center. Ask your child's medical team or Sisters by Heart for guidance on how to obtain a second opinion.

## Suggested Questions to Ask the Cardiac Team About Your Hospital/Cardiac Center

Can you tell me more about your center? Why is your center the best option for the care of my child?

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How many Norwoods/Hybrids has your program done in the last year? Over the last five years?

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What are your surgeon's expected survival rates from birth through the second surgery (Glenn)? What are your center's overall survival rates from birth through Glenn? How do your survival rates compare with other centers' survival rates?

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What are the most common complications following a Norwood/Hybrid? At your center, what percentage of babies need re-**intervention** (cardiac catheterization or surgery) in the 1st year of life?

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Do your surgeons hold **sub-specialty certification** in **congenital cardiac surgery**? When can I meet with the surgeon who will perform my child's surgeries?

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Does your program participate in the National Pediatric Cardiology Quality Improvement Collaborative (**NPC-QIC**), **Society of Thoracic Surgeons (STS) Database**, and/or **Impact Registry**?

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How am I included in decision-making regarding my child? How can I communicate with the surgical team before, during, and after surgery? Who is on my child's care team, and what are their roles (i.e., **primary cardiologist, surgeon, ICU team, outpatient team**, etc.)?

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How do you decide when my child will have each surgery – Norwood, Glenn, and Fontan? Do you have certain criteria (weight, age, etc.) my child must meet prior to each surgery?

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## **Suggested Questions to Ask the Cardiac Team About Your Hospital Stay**

How long can I expect my child to be in the hospital after the Norwood? Glenn? May I participate in daily clinical "rounds" when my child's health is discussed with the care team?

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What are your visiting policies? Am I allowed to stay with my baby overnight? Can you help prepare siblings for hospital visits?

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What are my options for when, where, and how to deliver my baby? When and how do you communicate with my **OB** regarding delivery and **perinatal care**?

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When and how do you communicate with my child's pediatrician following birth, procedures, and surgeries? When do I need to identify a pediatrician for my child, and can you provide guidance in my search?

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Who will care for my baby between birth and his or her first procedure? After the procedure? Do you have a dedicated Cardiac Intensive Care Unit (CICU)? Can I tour your heart center? Are physicians in the CICU **board certified** in both **cardiology** and **critical care**?

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Will I be able to hold my baby before and after his or her Norwood? If so, when and how?

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How will I feed my baby before and after the first surgery? Can I breastfeed? If so, what support do you have for breastfeeding moms? What percentage of babies eat by mouth when they are released from the hospital after the Norwood?

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Will my baby be sent home after his or her Norwood recovery? How will you prepare me to care for my baby at home? Does your center require Interstage monitoring? If so, who is responsible for my baby's care during the Interstage period?

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What support is available for me and my family? Do you provide any professional or peer support? Do you provide financial, nutritional, mental health, and developmental guidance?

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## Suggested Questions to Ask the Cardiac Team Looking Ahead

What can I expect for the next few years as my child develops and grows? Can he or she play sports? Are there possible life-long complications my child may experience?

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Developmentally and socially, what can I expect for my child as a preschooler, school-age child, teenager, and as an adult? Do you have a neurodevelopmental program my child can participate in?

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Do you have a plan for transitioning my child from pediatric to adult care?

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# 19

## Research and Family Partnership

*James Cnota, MD, Cincinnati Children's Hospital Medical Center*

**PARENT PARTNER**

*Jillian Hintz, Cincinnati Children's Hospital Medical Center*

As is true for all sections of our *Single Ventricle Guide*, the information provided is general, as each child is different and will follow their own path through treatment and recovery.

## Improving Outcomes

The goal of the NPC-QIC is to improve the outcomes for children with complex CHD. This challenge must be approached from multiple angles. The NPC-QIC's two-pronged quality improvement approach of sharing best practices and standardizing care has led to increased survival, improved growth, and fewer hospital re-admissions. However, creating the next set of best practices will require new ideas and new approaches. We need an even deeper understanding of the problems our patients and families face, and we need to develop new treatments that can then be shared using quality improvement tools.

**Innovation** is critical. A better understanding of any problem requires study. New treatments must be monitored to be sure they provide enough benefit without too much risk. This is clinical research, and like **improvement science**, it requires a partnership between the family and research team. Without such a relationship, the work of identifying and testing new and improved treatments cannot be accomplished. And because many of the problems facing children with CHD are unique, we cannot assume that information from other patient groups will apply.

As the caretaker of a child with complex CHD, you can expect to be approached with requests to partner in research. A research partnership means that patients, families, and scientists work together to find the best answers to difficult questions. In addition to participating in research projects, we also need your input about what unanswered questions are important to you. Our hope and our goal are to improve the outcomes, not only for our current patients, but future patients as well.

## A Parent's Perspective

### Dear Patients, Parents, and Caregivers:

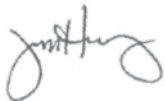
My son, Joey, was born in February 2017 with several congenital heart defects. Joey underwent his first open-heart surgery at 3 days old. His second surgery occurred the day he turned 7 months. Today, Joey is thriving. Joey would not have the bright future that he has today had it not been for research studies conducted in the past. Advancements in medicine gave our child a chance at life. Throughout your journey,

you may be presented with research opportunities. Since your reason for being at the hospital may be stressful, emotional, and exhausting, it can become overwhelming to receive requests for participation at the same time. I'd like to share with you a few tips for navigating this part of your journey:

- **It is a personal choice.** It is up to you to decide if participating in research studies is right for you or your child. Your decision will not impact the level of care you receive, and feelings won't be hurt. Every patient and family has the right to follow their personal beliefs. Do not feel badly if you choose to pass on a study, for whatever reason.
- **Ask for time.** Hospital visits and inpatient stays can be busy, to say the least. Your reason for being at the hospital may require you to meet with many medical professionals, and your time may be stretched thin. It's OK to ask the research coordinator, who will present the opportunity to you, to come back at another time. I would also recommend setting a time for re-connecting so that both parties are able to focus on the discussion.
- **Ask questions.** If you need more explanation about the opportunity being presented, don't hesitate to ask. Part of deciding whether research studies are right for you and your child is being completely informed. If you would like to speak to the professional conducting the research, or another credentialed member of the study, just ask. They'll be happy to talk with you.
- **Use your team.** I do not mean that you should ask your medical team to communicate with research coordinators on your behalf. If, however, you are unable to get in contact with a research coordinator or would prefer not to receive information about research opportunities, your team can help you to make sure the message is received. It takes a village to ensure that you and your child are getting the best care possible, and working together in this area is part of it.

I hope you find this information helpful. I wish you all the best on your journey.

Sincerely,



*Jillian Hintz*

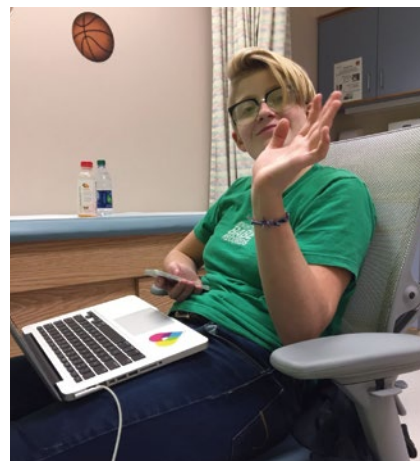
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## Lucy

MOUNT HOLYOKE CLASS OF 2022, HLHS



Doing an exercise (stress) test at a study follow-up.



Staying entertained (and hydrated) during a research study visit!

# 20

## Resources

## Beads of Courage

Through the use of strings of beads representing medical procedures, helps children with serious illness record, tell and own their stories of survival.

[more info](#)

## Congenital Heart Information Network

National organization that provides reliable information, advocacy, support services, financial assistance and resources to people affected by CHDs. Also includes information on Spencer's Fund, which provides help to families affected by CHD's by providing financial assistance with expenses incurred during extended or far from home clinic appointments and hospital stays.

[more info](#)

## Icing Smiles

Provides custom celebration cakes and other treats to families impacted by critical illness of a child (provides cakes to both the affected child and siblings).

[more info](#)

## Little Hearts

A national organization dedicated to providing support, education, resources, networking, and hope to families affected by congenital heart defects. Membership consists of families nationwide who have or are expecting a child with a congenital heart defect.

[more info](#)

## Mended Little Hearts

A support program for parents of children with heart defects and heart disease. Offers resources and a supportive network of families.

[more info](#)



## Midwest Heart Connection

Midwest Heart Connection is a 501(c)3 nonprofit organization that supports families affected by all types of CHD. Midwest Heart Connection's mission is to support individuals affected CHD by providing access to others with CHD, raise awareness of CHD in the community, and raise and provide funds for pediatric cardiology research, education, and treatment resources so that more children with heart defects will survive, live longer, and lead an improved quality of life.

[more info](#)

## National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC)

The NPC-QIC is a network of 60 pediatric cardiology care centers across the United States and the District of Columbia. With our parent partner organization, Sisters by Heart, we work together with families, clinicians, researchers, and patients to dramatically improve the outcomes for children with **cardiovascular** disease.

- [NPC-QIC](#) website
- [HLHS Book of Hope](#)  
This book features stories of hope and inspiration from parents and patients of HLHS. These are encouraging messages to let families know someone else has been there too and you are not alone.
- [Parent's Guides to HLHS](#)  
The guides offer support, information and resources to families during their first year with HLHS. Co-developed by parents and clinicians, they are in bulletin format.
- [Questions to Ask When Selecting Your Pediatrician](#)  
NPC-QIC parents suggest that you ask these questions when you are selecting a pediatrician.
- [Research Explained](#)  
HLHS research articles summarized for comprehension by families.
- [Sisters by Heart \(SBH\)](#)  
Sisters by Heart provides support, education, and empowerment to families affected by HLHS – from initial diagnosis and beyond.

- **All About HLHS**

*Many links to informative and educational sites which will help you understand HLHS from leading medical establishments.*

- **Article Downloads**

*Full length articles for printing*

- **Brochure Downloads**

*Brochures designed to be printed*

- **Linked by Heart**

Advances the mission of Sisters by Heart by providing additional regional and local support and resources to families affected by HLHS. Linked by Heart also provides an extensive password-protected secure database for HLHS families to connect and network nationally.

- **Video: Transitioning to Home for the Interstage**

Preparing for Interstage with your HLHS infant. Made for parents and caregivers, by parents and caregivers.

# A

## Glossary/Terminology

**Ablation**

A procedure in which a long, flexible tube is inserted into a vein or artery and is passed to the heart where it delivers heat or extreme cold to destroy the tiny area of tissue that is causing an irregular heartbeat

**ACE inhibitors**

Group of medications that relax veins and arteries to reduce blood pressure

**Advanced cardiac therapy (ACT) specialists**

Members of the heart transplantation team who help families decide if heart transplantation is right for their child

**Adcirca**

see **Tadalafil**

**Adult congenital cardiologist**

Doctors who are specially trained to care for adults who were born with abnormal hearts

**Adverse event**

An unexpected medical problem that happens during treatment with a drug or other therapy. Adverse events may be mild, moderate, or severe, and may be caused by something other than the drug or therapy being given. Also called *adverse effect*.

**Aldactone**

see **Spironolactone**

**Alpha-1 antitrypsin**

A protein that can be measured in the stool when there is concern for protein-losing enteropathy

**Amniocentesis**

Test performed on a pregnant mother's belly in which a thin needle is used to sample a small amount of amniotic fluid from the sac that surrounds the baby in the womb

**Anesthesia**

Medication given before surgery that puts a person unconscious (sleeping) and unable to move or feel pain or pressure

**Anesthesiologist**

Doctor who specializes in anesthesia care and who monitors patients' vital signs during surgery

**Angiography**

X-ray testing of blood vessels after the injection of contrast dye

**Anticoagulant**

Type of medication that prevents the blood from clotting by affecting clotting factors (proteins) in the blood

**Antiplatelet agent**

Type of medication that prevents blood from clotting by affecting platelets (cells in the blood that stick together to form clots)

**Antibodies**

Proteins in the blood that help to recognize bacteria, viruses, and foreign substances to help fight infection

**Aorta**

The great artery that carries blood from the heart to be carried to branch arteries throughout the body

**Aortic valve**

This heart valve separates the aorta from the left ventricle (lower heart chamber) and prevents blood from flowing back into the left ventricle

**Arrhythmia**

An alteration in rhythm of the heartbeat either in time or force

**Arterial line**

see **Intra-arterial line**

**Artery**

Blood vessel that carries blood away from the heart and to the rest of the body

**Aspiration**

Drawing fluid such as feedings into the lungs

**Aspirin**

A medication that is used to prevent blood from clotting easily by making platelets “less sticky”

**Atresia**

Absence or abnormal narrowing of an opening or passage in the body, e.g., a heart valve

**Atrial lines**

Special thin IV lines that are placed through the skin in the top of the belly that lead to the top chambers of the heart (atria)

**Atrial septal defect (ASD)**

A hole in the wall between the two upper chambers of the heart

**Atrium (atria, pl.)**

Each of the two upper chambers of the heart from which blood is passed to the ventricles (lower chambers of the heart). The right atrium receives **blue** blood from the veins of the body; the left atrium receives **red** blood from the pulmonary vein.

**Balloon catheter**

A special long, thin tube with a small inflatable tip that is guided through the blood vessels to a place of blockage and is inflated to stretch or open the blockage

**Barium swallow test**

see **Video fluoroscopic swallow study**

**Beta-blockers**

A class of medications that slow the heart rate and decrease blood pressure

**Bidirectional Glenn procedure**

see **Glenn procedure**

**Bilateral**

Both sides of the body

**Biopsy**

A procedure that involves removing a sample of tissue from the body to examine it under the microscope and perform test to identify disease

**Blalock-Thomas-Taussig (BTT) shunt**

A small connection between the neo-aorta and arteries to the lungs that is created during the Norwood procedure; also called a BT shunt

**Blood transfusion**

Giving donated blood to a person who has lost blood or is not making enough blood cells; all donated blood is carefully screened to ensure that it is safe

**Blue (deoxygenated) blood**

Blood without enough oxygen

**Board certified**

Describes a doctor who has had extra training and has taken an extra step to show that he or she understands the latest advancement in their specialty and has the skills needed to provide the highest quality of care

**Bonding**

The formation of a mutual emotional and psychological closeness between parents (or primary caregivers) and their newborn child. Babies usually bond with their parents in the minutes, hours, or days following birth.

**Breathing tube (endotracheal tube)**

Endotracheal intubation is a medical procedure in which a tube is placed into the windpipe (trachea) through the mouth or nose. In most emergency situations, it is placed through the mouth.

**BTT shunt**

see **Blalock-Thomas-Taussig shunt**

**Capoten**

see **Captopril**

**Captopril**

A type of ACE inhibitor

**Cardiac catheterization (“cath”)**

A procedure that involves passing a thin flexible tube (catheter) into the right or left side of the heart. The catheter is most often performed by inserting a needle into the groin.

**Cardiac computed tomography (CT) scan**

An imaging method that uses x-rays to create detailed pictures of the heart and its blood vessels

**Cardiac intensive care unit (CICU)**

A hospital ward that specializes in the care of heart patients requiring more advanced support of lung and heart function, more frequent monitoring and more intensive nursing care usually before and/or after heart surgery. Also known as the Cardiothoracic intensive care unit (CTICU).

**Cardiac magnetic resonance imaging (MRI)**

A painless imaging test that uses radio waves, magnets, and a computer to create detailed pictures of the heart

**Cardiac transplantation**

A surgical procedure in which a diseased heart is replaced with a healthy heart from a person who has died

**Cardiology**

The medical specialty that focuses on treating heart disease

***Cardiopulmonary (heart-lung) bypass (machine)***

A technique that uses a machine to temporarily take over the function of the heart and lungs during surgery, maintaining the circulation of blood and oxygen to brain and rest of the body

***Cardiovascular***

Involving the heart and blood vessels

***Catheter***

A long, thin, flexible tube inserted into the body; e.g., cardiac catheterization, Foley catheter

***Cavopulmonary shunt***

A bidirectional shunt placed as the first stage in the treatment of HLHS (Glenn surgery)

***Cell-free fetal DNA testing***

Screening test that uses a sample of a pregnant mother's blood to look for copies of fetal (baby) DNA.

***Central venous line***

A thin tube placed into the neck or groin that is passed into the vena cava to measure pressures directly, take blood samples, and give medicines or nutrition

***Cesarean section***

A surgical procedure involving incision of the walls of the abdomen and uterus (womb) for delivery of a baby

***CHD***

see ***Congenital heart disease***

***Chest tube***

A tube that drains fluid or air from around the lungs or sac around the heart into a collection box

***CHF***

see ***Congestive heart failure***

***Chromosomal defect***

A problem with one's genes where all or part of a gene is missing, extra, or irregular

***Chromosomes***

Structures in every cell that contain DNA, which holds specific codes that define individual characteristics that are unique to a person

***Chylous effusion***

Milky, high-fat drainage from a chest tube

***Circulation***

Movement of blood through the arteries and veins; delivers oxygen and nutrients to and removes waste from the body's cells; requires the action of the heart to pump the blood

***Clexane***

see ***Enoxaparin***

***Clotting factors***

Proteins in the blood that help the blood to clot

***Colostrum***

Milk released from a mother's breasts for a few days after childbirth that is high in protein and antibodies (proteins that fight off infection)

**Compounded**

Combining, crushing, or suspending medication in liquid to make it easier for patients to take

**Computed tomography (CT, CAT) scan**

A non-invasive test that uses x-rays and sometimes contrast dye to create images of the inside of the body; involves a large, donut-shaped scanner

**Congenital cardiac surgery**

Heart surgery to treat patients born with an abnormal heart or large blood vessels connected to the heart

**Congenital heart disease (CHD)**

Condition where an infant is born with an abnormal heart or large blood vessels connected to the heart

**Congestive heart failure (CHF)**

Condition in which the heart is not pumping well, which leads to build-up of fluid in the lungs or lower part of the body

**Coumadin**

see **Warfarin**

**CPR**

Cardiopulmonary resuscitation (a process in which someone tries to revive a person without a pulse by repeatedly compressing the chest and giving breaths through the mouth)

**Credentialed**

Having undertaken the education and training needed to show that a professional has the knowledge, skills, and ability to treat patients in a specific area of medicine

**Critical care**

The specialized care of patients who have life-threatening conditions and need constant monitoring

**Deletion**

A change in a gene in which a part of a chromosome is missing

**Deoxygenated**

see **Blue blood**

**Developmental delay**

Not reaching the same level of growth either mentally or physically as children of the same age

**Developmental milestones**

Behaviors, abilities, and skills that are seen at specific ages during infancy and early childhood

**Digitek**

see **Digoxin**

**Digoxin**

A medication that helps the heart pump more effectively

**Diuretics**

A group of medicines that cause the kidneys to make more urine



**Duplication**

An extra copy of a section of a chromosome

**Early Intervention (EI)**

State-run program that can send healthcare team members to your home to provide services for babies from birth to 3 years of age to help with or help to prevent developmental delays.

**Endocarditis**

An infection of the inner lining of the heart that usually results from the spread of germs from another area of the body, such as the mouth.

**ECG**

see **Electrocardiogram**

**ECG/EKG leads**

Stickers placed on the chest and belly that detect the heart rate and rhythm and breathing rate to be displayed on the monitor

**Echocardiogram/echocardiography**

A test that uses sound waves to view the heart

**EI**

see **Early Intervention**

**EKG**

see **Electrocardiogram**

**Electrocardiogram (ECG, EKG)**

Non-invasive test in which leads that detect electricity are placed on the chest and arms and legs; shows the rate and rhythm of a person's heartbeat

**Enalapril**

A type of ACE inhibitor

**Endotracheal tube**

see **Breathing tube**

**Enoxaparin**

A form of heparin

**ENT**

see **Otolaryngologist**

**Enteral feeds**

Nutrition given through a tube that goes directly to the stomach or small intestine

**Environment**

The surroundings in which a person lives

**Enzymes**

Substances produced by the body that help chemical reactions occur more easily

**Esophagus**

The tube that connects the throat to the stomach

**Exercise (stress) test**

A test in which a patient walks on a treadmill or rides a stationary bike while the heart is monitored closely

**Exposures**

Risks that an unborn infant might have come in contact with through connection while in the womb

**Extracardiac conduit**

As part of the Fontan surgery, a tube that passes outside the heart to send blood from the lower part of the body (from the inferior vena cava) to the pulmonary (lung) artery

**Extubate/Extubation**

Removing a breathing tube to allow a person to breathe on their own

**Feeding tube**

A small tube that typically goes through a person's nose or mouth and into the stomach or small intestine to provide nutrition

**Fenestration**

A small hole or connection between the Fontan tunnel and the heart

**Fetal**

Relating to an unborn child

**Fetal cardiologist**

Pediatric heart doctors who specialize in fetal cardiology, that is, diagnosing and treating unborn babies with heart problems

**Fetal echocardiogram/echocardiography**

A non-invasive test that uses sound waves to view a baby's heart while still in the womb

**Fetal intervention**

Surgical procedure performed on an infant while still in the womb

**Fiberoptic Endoscopic Evaluation of Swallowing (FEES)**

A procedure in which a therapist passes a thin, flexible camera through the nose to look at the parts of the throat as the patient swallows

**Foley catheter**

A flexible tube that is passed through the urethra (tube inside penis or vagina) to the bladder to drain urine

**Fontan surgery/tunnel**

Third surgery for babies with HLHS or other forms of single ventricle CHD; usually performed between 2 and 4 years old; re-directs blue blood from the lower body to the pulmonary arteries, either through the heart or using a tube along the outside of the heart

**Furosemide**

A diuretic medicine that works by causing the kidneys to make more urine

**Gastroesophageal reflux**

When stomach contents frequently flow back up into the esophagus (tube connecting the mouth to the stomach)

**Gastrojejunal tube (GJ tube)**

Tube that leads from the mouth to the second part of the small intestine (jejunum); used for nutrition

**Gastrostomy tube (G tube)**

A tube inserted through the belly that is used to give nutrition directly to the stomach

**Genes**

The building blocks that make up chromosomes which pass on genetic information from parents to children

**Genetic**

Relating to a person's genes, which are inherited from each parent; problems or defects can be passed down from a parent or can be triggered by one's environment when in the womb

**General anesthesia**

A combination of medicines to put a person into an unconscious (sleep-like) state in which one cannot feel pain during a surgery

**Genetic syndrome**

A combination of defects or abnormalities in one's genes that leads to a group of symptoms; can be passed down from a parent or can be triggered by one's environment when in the womb

**G tube**

see **Gastrostomy tube**

**GJ tube**

see **Gastrojejunal tube**

**Glenn surgery**

Second surgery for babies with HLHS; usually at 4 to 6 months of age; re-routes **blue** blood from the upper body directly to the left and right pulmonary arteries so that it does not need to pass through the heart; also known as a *bidirectional Glenn* or *cavopulmonary shunt* surgery

**Heart-lung bypass machine**

A mechanical pump that maintains circulation (blood flow) during heart surgery by taking over the work of the heart and lungs; it draws blood away from the heart, puts it through an artificial machine that filters oxygen into the blood, and returns it to and pumps it through the body

**Heparin**

A medication that prevents blood clots from enlarging or from forming in the first place

**Holter monitor**

A device used to record the heart rate continuously can be worn at home, usually for 24 to 48 hours

**Hydrochlorothiazide**

A type of diuretic; also called Microzide

**Hypoplastic**

A condition in which an organ or part remains below the normal size or in an immature state

**HLHS (Hypoplastic Left Heart Syndrome)**

A congenital malformation of the heart in which the left side is underdeveloped resulting in not enough blood flow to support the brain and the rest of the body

**IL**

see **Intralipids**

**Improvement science**

Research that focuses on studying ways to improve patient care to ensure that it is as safe and effective as possible

**Individualized**

Created especially for a person based on the exact needs of that person

**Innovation**

New ideas or methods

**Inferior vena cava (IVC)**

The largest vein in the human body; this great vein returns blood to the right atrium (upper chamber) of the heart from the lower body

**Impact Registry**

A database of information that helps hospitals and other centers provide measurable, better-quality care based on strong evidence and real-world patient experiences

**INR**

see **International normalized ratio**

**Intensive care specialist**

Doctor who specializes in caring for critically ill patients in the ICU; also called intensivist

**International normalized ratio**

A blood test that helps determine how well a patient's blood is being prevented from clotting or "how thin" it is

**Interstage**

Time between the Norwood or Stage 1 surgery and Glenn surgery

**Intervention**

Treatment or procedure

**Interventional catheterization**

Treatment or repair of a heart condition during a heart catheterization (see **Cardiac catheterization**)

**Intra-arterial line**

A thin catheter inserted into an artery, usually used to measure blood pressure directly rather than with a blood pressure cuff; also called an "A-line" or "art-line"

**Intracardiac baffle**

see **Intracardiac tunnel**

**Intracardiac tunnel (also called a lateral tunnel or intracardiac baffle)**

As part of the Fontan surgery, this tunnel passes through the heart and uses the wall of one of the upper chambers of the heart (atrium) as part of the tunnel; it allows blood from the lower body to go directly to the lungs rather than needing to be pumped by the heart

**Intralipids (IL)**

Brand name of fat mixture that is given as a form of nutrition directly into the bloodstream

**Intravenous (IV)**

To give fluids or medications directly in to a vein (e.g., medication, blood, or nutrition); IV or IV pump also refers to the device used

***Intubated/Intubation***

The placement of a tube into the trachea (windpipe) to help a patient to breathe; can be used with a bag and mask to give oxygen or can be connected to a ventilator (breathing machine)

***IV***

see ***Intravenous***

***IVC***

see ***Inferior vena cava***

***Kangaroo care***

A method of caring for a baby, especially if premature, in which the bare skin of an infant is held against the bare skin of a parent for as long as possible each day

***Karyotype***

Number and appearance of chromosomes in a cell; a normal human number is two copies of each of 22 chromosomes, plus two sex chromosomes (46XX is a normal female karyotype, 46XY for male)

***Laryngoscopy***

A test in which a doctor can look directly at a person's vocal cords with a scope

***Lasix***

see ***Furosemide***

***LFTs***

see ***Liver function tests***

***Lipids***

Substances that dissolve in alcohol but not in water, e.g., fats, oils, waxes (see ***Intralipid [IL]***); often used to provide fat to those receiving intravenous nutrition (see ***total parenteral nutrition [TPN]***).

***Lisinopril***

A type of ACE inhibitor

***Liver function tests (LFTs)***

A set of blood tests that measures enzymes and proteins made by the liver; shows how well the liver is working

***Lovenox***

see ***Enoxaparin***

***Lung maturity***

The ability of an infant's lungs to function (take in oxygen and breathe out waste) once outside the womb

***Lymphatic***

Related to the lymphatic system, which transports lymph, a colorless fluid that contains white blood cells and drains into the bloodstream

***Magnetic resonance imaging (MRI) scan***

Non-invasive test that uses a strong magnet, radio waves, and sometimes contrast dye to take images of the inside of one's body; uses no radiation; machine makes loud sounds; infants usually require sedation

***Maternal-fetal medicine (MFM) specialist***

A doctor who specialized in maternal-fetal medicine, who takes care of women having high-risk or complicated pregnancies

***MFM specialist***  
see ***Maternal-fetal medicine specialist***

***Microarray***  
A detailed genetic DNA test that specifically maps chromosomes or pieces of chromosomes

***Mindfulness***  
The practice of maintaining a nonjudgmental state of focused or complete awareness of one's thoughts, emotions, or experiences on a moment-to-moment basis

***Mitral valve***  
A heart valve between the left atrium and left ventricle that opens to allow blood into the ventricle and closes to keep the blood from flowing back into the atrium

***Modified barium swallow test***  
A special x-ray of the throat that shows how well a person can swallow

***MRI***  
see ***Magnetic resonance imaging scan***

***Nasoduodenal (ND) tube***  
A tube placed in the nose and down into the first part of the small intestine (duodenum) used to give breastmilk, formula, or other nutrition

***Nasogastric (NG) tube***  
A tube placed in the nose and down into the stomach to keep the stomach empty and decrease the chance of vomiting; can be used to give breastmilk, formula, or other nutrition

***Nasojejunal (NJ) tube***  
A tube placed in the nose and down into the second part of the small intestine (jejunum) used to give breastmilk, formula, or other nutrition

***Neonatal***  
Relating to the first few weeks of life of a newborn child

***Neonatologist***  
Pediatrician who specializes in neonatology, that is, caring for newborn babies, especially those who are ill or premature

***Neonatal intensive care unit (NICU)***  
Specialized hospital unit where critically ill newborns receive care under constant supervision and monitoring

***NG tube***  
see ***Nasogastric tube***

***Non-invasive***  
No probes or needles are used, and everything is done from the outside of the body

***Non-invasive prenatal testing (NIPT)***  
see ***Cell-free fetal DNA testing***

**Non-stress test**

A common non-invasive prenatal test to check on an unborn baby's health. A stretchy belt monitor is placed around a pregnant mom's belly that monitors the baby's heart rate, while mom keeps track of the baby's movements that she feels. The "non-stress" part means that the baby does not receive any stressful stimulation during this test

**Norwood surgery**

First surgery for babies with HLHS; usually within the first few weeks of life; includes multiple steps that re-direct blood flow so that the body is getting enough blood from the heart pump and there is enough blood going to the lungs to get oxygen

**NPC-QIC**

National Pediatric Cardiology Quality Improvement Collaborative

**Nurse Practitioner**

A registered nurse (RN) who has completed additional training and can provide medical care and prescribe medications without direct supervision of a physician

**Obstetrician (OB)**

Doctor who cares for pregnant mothers

**Obstructive lesions**

Blockage within the heart or blood vessels

**Occupational therapist/therapy (OT)**

Healthcare team member who works with children, helping them function as well as possible in their environment; OTs help children develop fine motor skills, hand-eye coordination, and feeding skills

**OG tube**

see **Orogastric tube**

**Operating room (OR)**

Room in a hospital where a surgery (operation) takes place

**OR**

see **Operating room**

**Oral aversion**

Fear, lack of interest in, or resistance to eating, drinking, or accepting sensation around the mouth

**Oral motor skills**

Movement of the muscles of the face to include the cheeks, jaw, tongue, and lips; usually refers to skills used in eating and drinking

**Orogastric (OG) tube**

Feeding tube that goes from the mouth to the stomach

**Otolaryngologist**

Doctor who specializes in disorders of the ears, nose, and throat; also called ENT or ORL

**Outcomes**

Results that occur because of treatment by the healthcare team

**Oxygen saturations (O<sub>2</sub> sats)**

A measurement of the amount of oxygen in the blood that is delivered to the body; measured by a pulse oximeter

**Oxygenated blood**

see **Red blood**

**Pacemaker**

A machine that can be used to treat arrhythmias (heart rhythm abnormalities). A temporary pacemaker is a small, box-shaped device kept outside of the body, which is used in the few days immediately following an operation. A permanent pacemaker is a small device, about the size of a silver dollar coin, which can be placed into the abdomen (stomach area) or thorax (chest); these are used to treat long-standing or permanent arrhythmias

**Pacemaker leads (or wires)**

Wires connecting the surface of the heart to a pacemaker; temporary leads are small and placed through the through the skin, usually at the top of the belly; permanent leads are usually slightly larger and placed internally

**Palivizumab**

An injected medication with virus-fighting antibodies that helps protect high-risk babies from a potentially serious lung infection called RSV disease; also called Synagis

**Palliative**

Refers to treating pain or discomfort without treating the cause of the condition

**Patent**

Open (not blocked or narrow), when referring to a blood vessel or part of the heart

**Patent ductus arteriosus (PDA)**

The *ductus arteriosus* is temporary normal blood vessel in an unborn baby that connects the pulmonary artery to the aorta. In the womb, it allows red blood from the umbilical cord to bypass the lungs and be delivered to the rest of the baby's body. When the ductus arteriosus fails to close after birth, it is called a PDA. In babies with CHD, doctors use medicine to keep the PDA so that it can red blood can continue to be delivered to the baby's brain and body.

**PDA**

see **Patent ductus arteriosus**

**Pediatric cardiologist**

A doctor who specializes in treating children with heart problems

**Pediatric cardiothoracic (heart) surgeon**

A doctor who specializes in surgeries for patients of all ages with congenital heart defects as well as other conditions of the chest

**Pediatric intensive care unit (PICU)**

An area of the hospital where specialized one-to-one care are monitoring are given to critically ill children



**Perfusionist**

Technician who operates the heart-lung bypass machine; works closely with the pediatric cardiothoracic surgeon during an operation.

**Perinatal care**

Care given to a woman and her child before, during, and after delivery from the 28th week of pregnancy through the 7th day after delivery

**Perinatologist**

Doctor who specializes in perinatal care (see **Perinatal care**)

**Physical therapist/therapy (PT)**

Specialist who helps patients with movement problems; for patients who are in the hospital, they also help to prevent problems with movement and strength

**Plastic bronchitis**

Condition in which lymphatic fluid builds up in the airways and forms casts (rubbery collections in the shape of the air passages) that can cause blockages, making it difficult to breathe

**Platelets**

Cells in the blood that, when activated, become sticky and clump together with proteins to form clots

**Pleural effusions**

A buildup of fluid around the lungs

**po**

Stands for per os, a Latin term that means by mouth; usually referring to taking medicine or nutrition by mouth

**Postnatal, postnatally**

Occurring after birth

**Prenatal, prenatally**

Occurring before birth

**Prostaglandins**

Medicine used to keep the patent ductus arteriosus (PDA) open, thereby maintaining blood flow to the brain and body, until the Stage 1 Norwood or hybrid Norwood is performed

**Protein-losing enteropathy**

Disease of the intestines in which protein leaks into the gut; can occur in children who have had their Fontan surgery

**Psychological**

Having to do with the mind; related to the mental or emotional state

**Pulmonary artery**

Blood vessel that receives blood from the right ventricle and divides into the left and right pulmonary arteries

**Pulmonary valve**

Heart valve between the right ventricle and main pulmonary (lung) artery; prevents blood from leaking backwards into the right ventricle

***Pulmonary vasodilator***

A medication that relaxes the muscles of the lung arteries, making it easier for the heart to pump blood to the lungs so that blood can flow more easily through the lungs

***Pulmonary veins***

Blood vessels that carry red (oxygenated) blood returning from the lungs to the left atrium of the heart

***Pulse oximeter/oximetry***

Machine that measures oxygen saturation (amount of oxygen in the blood)

***Recurrent laryngeal nerve***

Nerve that coordinates normal breathing and swallowing

***Red (oxygenated) blood***

Blood that contains high amounts of oxygen

***Rejection***

A condition in which a person's immune system does not recognize and attacks transplanted tissue (e.g., a heart)

***Renal***

Related to the kidneys

***Respiratory***

Related to breathing

***Respiratory syncytial virus (RSV)***

A common virus that causes mild cold symptoms in adults but that can cause serious illness in infants, particularly those with serious congenital heart disease

***Revatio***

see ***Sildenafil***

***Rooming in***

Arrangement in a hospital in which a newborn infant is kept in the parents' hospital room

***RSV***

see ***Respiratory syncytial virus***

***Sano shunt***

As part of the Norwood surgery, a shunt that is placed to allow blood from the right ventricle to be pumped to the pulmonary artery and then the lungs

***Sedated/sedation***

Giving a drug that helps a person relax or fall asleep

***Sensory processing***

The way a person's nervous system receives messages from the senses and turns them into responses

***Shunt***

A surgically created tube that allows movement of fluid (e.g., blood) from one part of the body to another

***Sildenafil***

A type of pulmonary vasodilator

***Social workers***

Healthcare team members who help you and your team communicate and help you cope with your child's journey

***Society of Thoracic Surgeons (STS)******Database***

Method by which records of heart surgery records are tracked in an effort to continually improve quality and patient safety

***Speech therapist, Speech/Language Therapy (SLT)***

Specialists who work with patients in the areas of speech and swallowing

***Spirolactone***

A type of diuretic

***Stenosis***

Narrowing of a passage in the body, e.g., a blood vessel or heart valve

***Stent***

A short, narrow metal or plastic tube, often in the form of mesh, that is inserted into a vessel (such as an artery), usually to keep a blocked passageway open

***Step-down unit***

A section of the hospital where a patient's heart is monitored but where closer intensive care is not required

***Stridor***

Harsh, hoarse wheezing sound when breathing

***Subcutaneous injection***

A type of shot in which a short needle is used to inject a drug into the tissue layer between the skin and the muscle

***Sub-specialty certification***

Certificate that shows a doctor has completed specialized training in a particular area of medicine or surgery and continues to pass tests that show an understanding of knowledge and skills in that area

***Superior vena cava (SVC)***

The large vein that brings blue (deoxygenated) blood from the head and upper part of the body back to the heart

***SVC***

see ***Superior vena cava***

***Synagis***

see ***Palivizumab***

***Tadalafil***

A type of pulmonary vasodilator

***Telemetry***

Continuous monitoring of heart rhythm (electrocardiogram or ECG), respiratory rate, and oxygen saturation

***Therapists***

Healthcare personnel specially trained in a specific area (e.g., physical therapist)

***Thoracic surgery nurses***

Operating room nurses with special training in the care of patients undergoing heart or chest surgery

***Thrombus***

Blood clot

***Trachea***

Tube that leads from the throat to the lungs; also called windpipe

**Tricuspid valve**

The heart valve between the right atrium (upper heart chamber) and right ventricle (lower heart chamber); prevents blood from flowing back into the atrium from the ventricle

**Total parenteral nutrition (TPN)**

A method of feeding in which nutrition is given directly into the bloodstream; when receiving TPN, the gut (stomach and intestines) are permitted to rest and recover

**TPN**

see **Total parenteral nutrition**

**Turner's syndrome**

A genetic condition in which a girl has only one X chromosome (normally, girls have two); girls with Turner syndrome are often short, not able to have children, and may have heart, kidney, and skeleton defects

**Ultrasound**

A test that uses sound waves to obtain live images of an organ, e.g., the heart (echocardiogram); also called sonogram

**Umbilical line**

A thin tube that is inserted into one of the arteries or the vein of the umbilical cord; used to take blood samples and provide fluids and medications

**Vein**

Blood vessel that carries blood from the body to the heart

**Ventilator**

A machine that provides artificial breathing to a patient who is unable to breathe or who needs assistance in breathing while the body heals

**Ventricle**

Pumping chamber of the heart; usually there is a right ventricle, which pumps blood into the lungs and a left ventricle, which pumps blood into the body.

**Video fluoroscopic swallow study (VSS)**

X-ray test to watch how a person swallows; also called barium swallow test

**Vital signs**

Temperature, respiratory (breathing) rate, heart rate, and blood pressure

**VSS**

see **Video fluoroscopic swallow study**

**Warfarin**

An anticoagulant medication commonly used to prevent blood clots

**Zestril**

see **Lisinopril**



## MORE INFORMATION

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