

# Pediatric Heart Failure

A Guide for Patients  
and Families



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# Acknowledgements

The authors would like to thank the Children's Cardiomyopathy Foundation for recognizing the need to develop an educational resource for families whose children are affected by heart failure. Their assistance in the development and review of this booklet, as well as their financial support, was invaluable. We gratefully acknowledge their contribution to this project.

Special thanks to the patients who submitted artwork to be featured in the book and to Novartis who also supported the production of this booklet. ♦



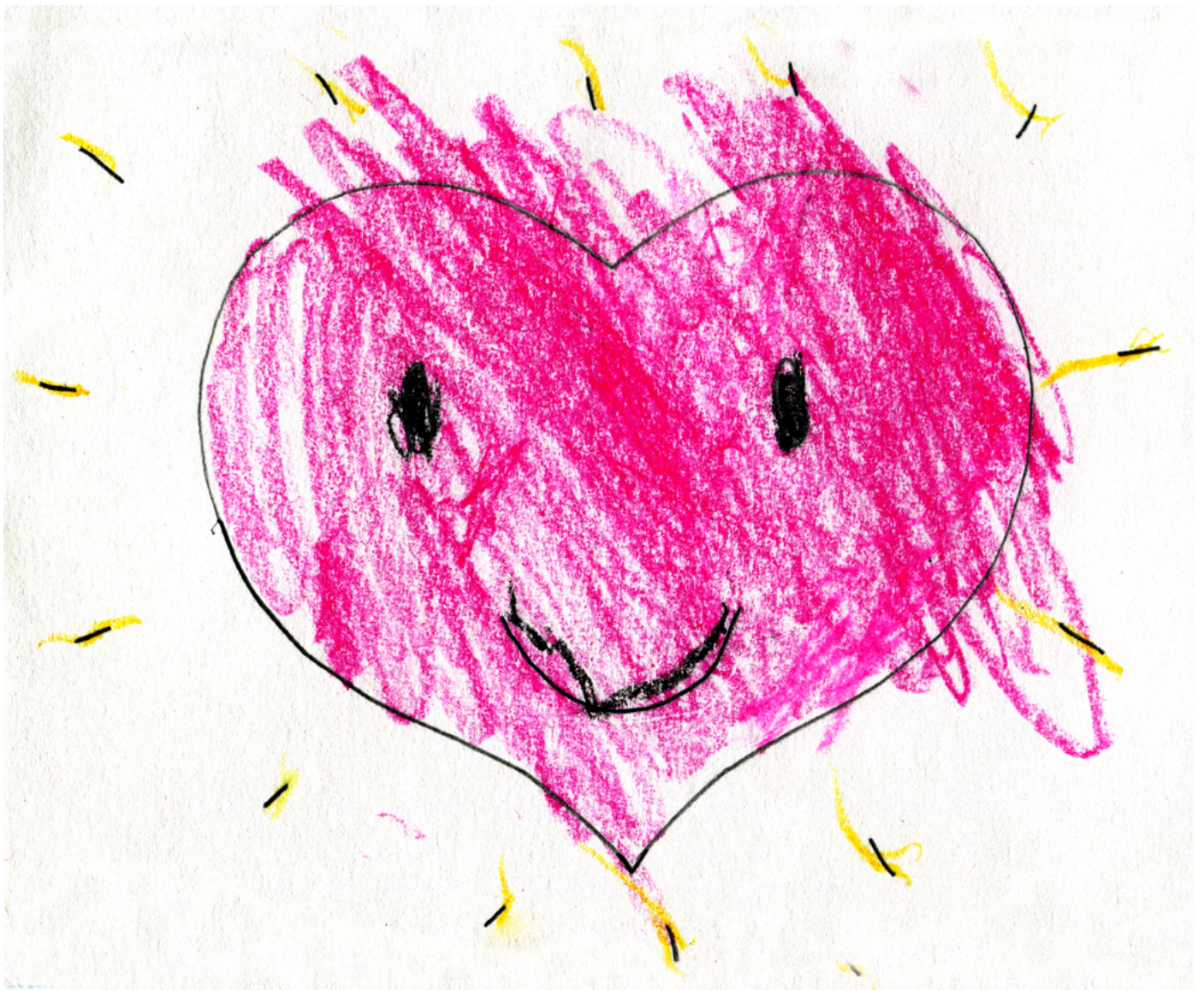
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# I. Normal Heart Function

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artist: gabriel, age 3



# I. Normal Heart Function

The heart plays an important role in keeping our body functioning. The job of the heart is to move blood around the body. The right side of the heart receives blood from the veins of the body and pumps it to the lungs. The oxygen-rich blood then returns to the left side of the heart to be pumped to the rest of the body. In addition to providing oxygen to the body, blood pumped by the heart provides essential fuel, vitamins, and minerals that allow our body to work well.

The heart is normally made of four chambers (right atrium, right ventricle, left atrium, and left ventricle) and has four valves. The atrium is the part of the heart that receives blood from veins. The job of the left and right atrium is to receive blood and move blood to the ventricles. The ventricles are made to pump blood - the right ventricle pumps blood to the lungs and the left ventricle pumps blood to the body. The four valves in the heart make sure that blood flows in the correct direction and does not flow backwards when the heart contracts and relaxes.

In the following sections, it will be explained what happens with a child has heart failure. There are many problems that can lead to heart failure and multiple treatment options. Not every aspect of treatment for heart failure can be covered in this guidebook. Therefore, it is important to have an in-depth conversation with your child's cardiologist. ♦

# II.

# Heart Failure

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## II. Heart Failure

### A. Types of Heart Failure

Heart failure occurs when the heart is not able to pump enough blood to the rest of the body. Heart failure is a general term and not all children have the same symptoms. Symptoms may be mild or severe. Some children may appear healthy and act “normal” on the outside, but the heart may not work well on the inside. Some children are born with heart problems and others develop heart problems later in life. The most common forms of heart disease causing heart failure in children are cardiomyopathies and congenital heart defects (CHD). A congenital heart defect is when the heart does not form correctly before a baby is born. There are many different forms of CHD. There are mild forms of CHD that are structural defects correctable with surgery and do not cause heart failure. Cardiomyopathy means “heart muscle disease” and is a general term used by doctors when the heart is not pumping properly. Your child’s cardiologist will discuss your child’s specific heart problem with you and let you know what to expect as your child grows and develops.

For most forms of heart failure, there is a problem with the heart function. Either the main pumping chamber of the heart fails to squeeze properly (systolic heart failure) or it fails to relax properly (diastolic heart failure). Infants and children with congenital heart defects may have more complex forms of heart failure and develop unique signs and symptoms that are different from children with cardiomyopathy.

### B. Systolic Heart Failure or Pump Failure

Weakening of the heart muscle can lead to what is typically called **systolic heart failure** or pump failure. In systolic heart failure, the heart muscle itself is weak and cannot squeeze enough blood out to the body to provide the energy that other organs need to work properly. When the heart is weak, the heart responds by becoming bigger (dilated), often with thinning of the heart walls. This form of cardiomyopathy is called **dilated cardiomyopathy (DCM)**. Your child’s doctor may describe the squeezing ability of the heart with an **ejection fraction (EF)**, which is the amount of blood pumped out to the body on each squeeze of the heart. The normal EF is 55-65%, although most children feel well even when the ejection fraction is lower than normal. When the heart is enlarged, the heart pumps harder to keep up with the needs of the body. Children with this heart enlargement can feel well for a period of time before developing symptoms of heart failure.

Children will eventually show signs of systolic heart failure by being more tired, and not being able to keep up with peers or perform activities like they had in the past. They may be less hungry, complain of stomach aches, be unable to keep food down, or have unexplained weight loss. Signs and symptoms of heart failure will be discussed in more detail in the next section of this guidebook. When the heart gets bigger this can lead to other problems, including abnormal heart beats (**arrhythmia**) or leaking heart valves.

Dilated cardiomyopathy and systolic heart failure can happen due to many causes. Some include viral infections in which the heart muscle is attacked by a common virus. When an infection injures the heart, it affects the pumping. This process, called **myocarditis**, is one of the more frequent causes for systolic heart failure in children. Affected children will need immediate medical evaluation and treatment, possibly in the hospital or the critical care unit. Sometimes myocarditis will improve, and the heart will heal back to its normal size with normal function (**systolic function**). Other times, heart function will not improve.



## II. Heart Failure ▶ B. Systolic Heart Failure or Pump Failure (Cont'd.)

Dilated cardiomyopathy can also happen due to genetic disease. Genetic diseases are ones that your child is born with, where the genes that instruct how the heart is supposed to form and work, are abnormal. These changes in the genes (**variants**) can lead to a number of different problems with the heart. Some genes that affect the heart are involved in using energy sources (such as sugar or fat that the heart muscle needs to work) while other variants lead to changes in the muscle proteins inside the heart. Sometimes heart failure shows up when the child is a baby, while other times it shows up later in life or even in adulthood. If your child's doctor is concerned about a genetic cause, they may ask you and your other children to get screened to make sure that your and your children's hearts are healthy.

Systolic heart failure can also develop after heart surgery. Even if the surgery is done correctly, the heart may get injured and not pump the way it should. Other times, the heart was weakened prior to surgery, and the pump failure is discovered after the surgery. When systolic heart failure follows heart surgery, it is treated like pump failure with standard monitoring and medications.

Systolic heart failure or pump failure is usually treated with medication first. Usually medicines are taken by mouth, but when a child is in severe heart failure, these medicines may need to be given intravenously. Common medications used to treat heart failure are outlined in the section on medications. If your child is started on IV medicine, the goal will be to change them to oral medicine for convenience. However, if the heart is too weak, they may need to consider advanced heart failure therapies as explained in the advanced heart failure section.

## C. Diastolic Heart Failure or Relaxation Failure

Sometimes the squeezing or pumping function (systolic function) of the heart is normal but the heart does not relax well in between beats. This is called **diastolic heart failure** or heart failure with preserved ejection fraction (HFpEF). In this case, the heart pumps well, but when it fills with blood, the heart becomes very stiff, and a lot of pressure builds up inside the heart. Increased pressure in the heart can cause fluid to back up into other tissues or organs of the body. For instance, the lungs may take on water (**pulmonary edema**), the liver may become enlarged (**hepatomegaly**), or extra fluid may cause swelling in the face, hands, legs, or feet (**edema**).

Diastolic heart failure is less common than systolic heart failure in children and can also have many causes. It can happen when the heart muscle is abnormally thick or when the muscle tissues do not relax normally. Thickening of the heart with poor relaxation can be caused by **hypertrophic cardiomyopathy (HCM)**. When the muscle is not thick but still does not relax well, this may be **restrictive cardiomyopathy (RCM)**. Similar to dilated cardiomyopathy, both hypertrophic cardiomyopathy and restrictive cardiomyopathy are caused by abnormal heart muscle genes which cause the muscle to not act normally. Either way, your child's doctor may order multiple tests to determine the cause of these types of heart disease. Often, when there is a concern for either hypertrophic or restrictive cardiomyopathy, your child's doctor may want to do genetic testing on you and/or any other children to make sure that other people in the family do not have the same heart problems.

Diastolic heart failure is often treated with medicines to decrease fluid retention (**diuretics** or water pills) to make your child urinate more often. These medicines can decrease the symptoms felt by your child and are the most effective medications for this type of heart failure. Your child's doctor may also prescribe other medications to lower blood pressure, slow the heart rate, or prevent additional injury to the heart as described in the medications section. If your child continues to have symptoms, advanced heart failure therapies may need to be considered. ♦

III.

# Signs and Symptoms in Heart Failure

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artist: alyssa, age 13

# III. Signs and Symptoms in Heart Failure

## A. Introduction

Signs and symptoms of an illness are related to how your child looks, feels, or behaves when they are sick. The symptoms of heart failure differ by age and are described below. It is important to pay attention to symptoms based on your child's growth and development. Some symptoms of heart failure, such as abdominal pain and chest pain, are common in children without a heart condition. Therefore, it is very important to communicate any new complaints or problems that your child may have to his/her cardiologist.

## B. Infants — less than 1 year old

Since infants cannot tell us how they are feeling, how they act is very important. Infants with heart failure most often show up at the doctor's office because they are having difficulty feeding, breathing, or both. This means that the infant could be breathing faster than usual or working extra hard to breathe. Breathing fast with no extra effort for a very short period is normal for a newborn. However, breathing fast with extra effort at all times could be a symptom of heart failure or other condition.

Difficulty feeding could also be a sign of heart failure. The infant may not be able to take as much breastmilk or formula as usual, or they may take longer to feed or take breaks during feeding to catch their breath. They may spit up or vomit frequently and have trouble gaining weight.

When you go to the doctor's office, they will examine your child to determine if they are experiencing heart failure. When they weigh and measure your child, they will plot these numbers on a growth curve. Children in heart failure may have poor weight gain or be diagnosed with "failure to thrive." Your doctor may also tell you that your child has abnormal heart and lung sounds or a large liver. These exam findings may indicate heart failure, and further testing by a cardiologist is recommended.

## C. Pre-School — 1-5 years old

In this age group, doctors still cannot solely rely on the child to describe their symptoms. Toddlers and preschoolers are usually very active, and one of the symptoms of heart failure is a child who gets tired easily or cannot keep up with his/her friends when playing. If you notice that your child often takes breaks from playing to rest or catch his/her breath or takes more frequent naps, you should let your doctor know. Some children may complain of chest pains, stomach pain, feeling nauseous, or not wanting to eat for multiple days without fever or other signs of an infection (such as a virus or "stomach bug").

When the doctor examines your child, they may speak about your child having an enlarged liver, or an abnormal heart sound called a gallop. The doctor might also be concerned about how your child is breathing if it takes a lot of work or extra effort to take each breath. Preschool children can also experience poor weight gain or growth ("failure to thrive"). If your doctor notices any of these symptoms on exam, they will do more tests to evaluate further for heart failure.



## D. School-Age — 5-12 years old

School-aged children are able to describe their symptoms better than younger children. Similar to younger children, school aged children in heart failure may complain of chest pain, abdominal pain, have a decreased appetite, or have stomach pains after eating. They may also experience vomiting without diarrhea.

When the doctor examines your child, they may mention your child has an enlarged liver or an abnormal heart sound called a gallop. They might be concerned that your child has trouble breathing or abnormal lung sounds due to pulmonary edema or water in the lungs. If your doctor finds any of these signs during the exam, they will discuss it with you and do further testing.

## E. Teenagers and Young Adults

During the teenage years, symptoms of heart failure can be similar to those seen in younger children, as well as what adults may experience.

Chest pain is a common symptom that often causes teenagers to worry about their heart. While chest pain can occur in heart failure, most chest pain in teenagers is not related to the heart. To help distinguish heart related chest pain from other causes, it is important to describe the type of chest pain (a dull or crushing type pain versus sharp pain), the timing of chest pain, and the location of the pain. Typically, heart failure-related chest pain is described as a dull or crushing pain that is located in the middle or the left side of the chest and occurs during exercise. It is often associated with shortness of breath and resolves when the child rests. Therefore, when you speak to your doctor about chest pain, it is very important to be able to describe how it feels, what brings on the pain, when it happens, how long it lasts, what happens immediately before, during, or after the pain, and what makes the pain go away. This information enables the doctor to determine the likely cause of the chest pain.

Another common symptom for heart failure during the teenage years is shortness of breath. Shortness of breath from heart failure typically occurs during exercise or strenuous activity. In mild heart failure, shortness of breath may never occur, or it could occur with activity or during sports. In more severe heart failure, light activities such as walking up a flight of stairs may cause shortness of breath. It is important when talking with your child's cardiologist to describe what activities cause shortness of breath and whether it changes over time. While heart failure can cause shortness of breath, other lung problems, such as asthma, can also cause shortness of breath.

Other frequent symptoms in heart failure include loss of appetite, nausea, or vomiting. Often these symptoms last for multiple days to weeks and do not improve. If your teenager develops these symptoms without signs of infection (fever, diarrhea or contact with other sick children), you should call his/her doctor.

When the doctor examines your child, the doctor may say your child has crackles in the lungs, a gallop or hepatomegaly. Crackles can be caused by pulmonary edema. Pulmonary edema, or water in the lungs, is due to a build-up of fluid in the lungs because the heart cannot pump blood through the body well. Your doctor may prescribe diuretics ("water pill") to help get rid of this fluid. A gallop is an extra heart sound that can be heard in children with heart failure. Hepatomegaly means that the liver is enlarged. The liver is close to the heart and can get bigger from the back up of blood when the heart is not pumping properly. ♦

# IV. Exercise, Restrictions, and Nutrition in Pediatric Heart Failure

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artist: rexford, age 9

## IV. Exercise, Restrictions, and Nutrition in Pediatric Heart Failure

### A. Exercising with Heart Failure

Exercise and physical activity are important for everyone. For infants and young children, regular child play is okay. In older children with heart failure, regular exercise, physical education classes (PE or gym), or playing sports may be possible and even encouraged depending on your child's condition and heart disease. Exercise may improve heart failure symptoms and help children feel better. Before starting any exercise program or a new physical activity, it is very important to speak to a pediatric cardiologist because some activities are not safe for children with heart failure. Your child's cardiologist can design an exercise program that is safe and will provide health benefits for your child. To determine the type and level of exercise that is best for your child, a cardiologist may do an electrocardiogram or exercise stress test. During an exercise stress test, the child's heart rhythm, blood pressure, and breathing are closely monitored by a medical team while walking, running on a treadmill or riding a stationary bicycle. How the heart responds to exercise will help the cardiologist make decisions about how your child should exercise. Recommendations should include what type of exercise is safe, how often to safely exercise, how long each exercise session should be, and how intense each session should be. Your child's cardiologist should also provide recommendations on participation in PE or gym class, as well as recreational and school sports teams. While no test can assure 100% safe sports participation, these tests along with a discussion of your child's individual condition and desires can help to develop a healthy sports routine.

Occasionally, the exercise stress test may show that there is a risk for an arrhythmia or abnormal heart rhythm. In these cases, your child's cardiologist may recommend an implantable cardiac defibrillator (ICD) if the arrhythmia is considered to be life threatening. ICDs are discussed in the section on advanced cardiac therapies.

### B. Competitive Sports

Participation in competitive sports typically involves frequent training with relatively intense levels of exercise. Furthermore, taking breaks and limiting activity is often discouraged during competitive training. In some cases of heart failure stress on the heart caused by the intense exercise or exertion may be dangerous. Even in patients who are encouraged to exercise, your child's cardiologist may restrict your child from more competitive sports activities such as soccer matches or high-intensity sports such as sprinting, swimming, football, basketball, or hockey because these sports are considered high-risk. In these cases, it is appropriate to discuss the specific restrictions in detail with your child's cardiologist, so everyone in the family understands exactly what is, and what is not a safe activity. Alternative activities should be discussed, as many children and teenagers enjoy participating in team activities with their friends, and sports often play a large role in their social lives.

### C. Eating and Drinking in Heart Failure

Good nutrition is a very important part of managing heart failure. The type of food and beverages a person consumes can either help or worsen symptoms of heart failure. Nutrition should be discussed with your child's cardiologist.



#### [IV. Exercise, Restrictions, and Nutrition in Pediatric Heart Failure ▶ C. Eating and Drinking in Heart Failure \(Cont'd.\)](#)

When food is eaten, the human body processes the food into energy. People, especially infants and young children with heart failure, often require extra calories to support their body and control heart failure. Infants may require increased calorie formulas or supplemental formula in addition to eating solid food. In some cases, infants or young children cannot take in enough formula or calories by mouth to grow and will need a feeding tube inserted. On the other hand, if too many calories are consumed, excess weight gain can worsen heart failure. For this reason, specific calorie goals and weight tracking are an important part of managing heart failure. Cardiologists, with support from nutritionists and primary care physicians, can help set these goals and review them with the family.

The amount of liquid that a child drinks may need to be closely monitored. In children with heart failure, too much water or other drinks absorbed by the body can lead to swelling, shortness of breath, stomach pain, or cause other heart failure symptoms. In some cases, even a “normal” amount of liquid per day can lead to symptoms. For this reason a cardiologist may set a fluid restriction — a maximum amount of liquid that an individual can drink per day. Fluid restrictions are put in place to control symptoms of heart failure and maintain the best possible health. Salt or sodium restrictions are also sometimes placed to prevent or manage symptoms. Eating foods that contain excess salt can cause the body to retain extra water. Similar to drinking excess liquids, salt increases the amount of water in the body, and extra water can lead to shortness of breath, stomach pain, swelling, or other heart failure symptoms. For teenagers, a cardiologist may advise limiting the amount of caffeine and to avoid coffee and energy drinks.

## D. Dietary Choices, Vitamins, and Supplements

While getting adequate calories to support the body is necessary, it is equally important to make healthy choices that will support heart health in the short and long term. A balanced diet containing lean proteins, whole grains, fruits, and vegetables will help to maintain heart health and overall body function. Consultation with your child’s cardiologist and/or a nutritionist can provide information about healthy diet choices.

A variety of vitamins, herbal supplements, and dietary supplements claim to enhance physical and/or heart health. However, not all of them are appropriate or safe for children in heart failure, and some may interact with heart medications to cause side effects. Patients with heart failure should treat vitamins and supplements as medications and discuss them with a cardiologist prior to starting them. ♦

V.

# Heart Failure: Diagnostic Testing

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artist: elizabeth, age 9

# V. Heart Failure: Diagnostic Testing

After your doctor has taken a medical history, reviewed your child’s symptoms, and performed a physical examination, diagnostic tests will be requested. All infants, children, and teenagers with a heart problem will undergo an electrocardiogram (EKG or ECG), echocardiogram (ECHO), and blood tests. Additional testing will be based on your child’s age and specific heart problem:

- **Blood Tests:** Tests will be done at the time of diagnosis to check for possible causes and to check the function of other organs that may be affected by heart failure. Some common blood tests are listed below, but the cardiologist may order additional tests based on your child’s specific heart disease. Blood tests will be repeated at most visits to monitor how the heart and body are doing.

COMMON DIAGNOSTIC LABS		
Test	Other Names Used	Purpose
<b>Complete Blood Count</b>	CBC	Checks for low blood counts (anemia) or signs of infection
<b>B-type Natriuretic Peptide*</b>	BNP, NT-proBNP	Checks for stretching of the heart muscle to determine if symptoms are caused by a heart problem  Monitors how the condition changes over time
<b>Basic Metabolic Panel</b>	BMP, Chemistry	Checks electrolytes in the blood and kidney function
<b>Liver Function Tests</b>	LFTs, albumin, INR, PT	Checks for inflammation and liver function
<b>Thyroid Function Tests</b>	TFTs, TSH, FT4	Checks for thyroid disease that may contribute to heart problems
Post Diagnosis Labs		
<b>Genetic Testing</b>	Cardiomyopathy Panel	Identifies if there is an underlying problem with the structure and function of the heart muscle cells. This is related to problems that are inherited and passed through families. If genetic testing identifies a genetic mutation, additional testing of family members will be recommended.

\* Newer labs besides BNP are being evaluated and may be helpful in the future



#### V. Heart Failure: Diagnostic Testing (Cont'd.)

- **Chest X-ray or Radiograph:** X-ray images give a “snapshot” of the condition of the lungs and heart, and may lead to additional imaging tests. In heart failure, the heart may appear enlarged and abnormal in structure. Also, fluid buildup may be visible in the lungs (pulmonary edema). An X-ray may give insight into other conditions that are causing symptoms.
- **Electrocardiogram (ECG or EKG):** Small stickers are placed on the chest, arms, and legs to record the electrical activity of the heart. This test screens for heart rhythm problems (arrhythmia), monitors the heart rate, and gives information about how the heart is beating.
- **Holter monitor:** If your child’s doctor is concerned about an abnormal heart rhythm, a Holter monitor may be ordered. This monitor is an extended EKG that is worn for 24-72 hours and continuously records the electrical activity of the heart.
- **Echocardiogram (ECHO):** The most common advanced imaging test used to diagnose and follow heart disease is an echocardiogram. A small camera and gel are placed on the chest and sound waves (ultrasound) capture moving images of the heart. An ECHO determines the structure or anatomy of the heart. It can also measure the size of the heart, determine how the heart is pumping, and detect any fluid build-up around the heart (pericardial effusion). Echocardiograms are done at the time of diagnosis and then repeated during regular cardiology visits to check the heart’s function.
- **Cardiac computerized tomography (CT) scan:** In a cardiac CT scan, your child lies on a table inside a doughnut-shaped machine while an X-ray tube rotates around his or her body and collects images of the heart and chest. These images provide a 3-dimensional (3D) view of the heart and surrounding structures. Your child will require an IV for a CT scan as it uses dye (contrast) to obtain pictures of the heart and blood vessels. Some children may require medication to stay calm during the test. Your child’s cardiologist will talk to you beforehand if sedation is needed. A CT scan does expose your child to radiation and will only be ordered if your cardiologist thinks it is necessary.
- **Cardiac magnetic resonance imaging (CMR or MRI):** In a cardiac MRI, your child lies on a table inside a long tube-like machine that produces detailed pictures of the heart and blood vessels. It can provide information about the heart’s function. Unlike echo or CT, cardiac MRI can show changes in the heart muscle due to inflammation, infection, or scarring. An MRI may require an IV for dye (contrast) and often requires sedation for young children who cannot lay still.
- **Stress test or exercise stress test:** A stress test measures how the heart and lungs respond during activity or exercise. Stress tests can be done in most children starting 6-8 years of age. Your child will walk or run on a treadmill or pedal a stationary bike while attached to an ECG machine. Occasionally, your child may receive medication through an IV that stimulates the heart like exercise would. Stress tests measure heart rate, heart rhythm, and blood pressure in response to exercise. This helps doctors understand how the heart responds to physical stress. With older children and teenagers, they may wear a special mask during the test to measure energy needs during exercise. Stress tests can be followed over time to check for improvement or worsening of a heart condition. If your child’s doctor also wants to see images of the heart while your child is exercising, they may order a stress echocardiogram, stress MRI, or nuclear stress test.

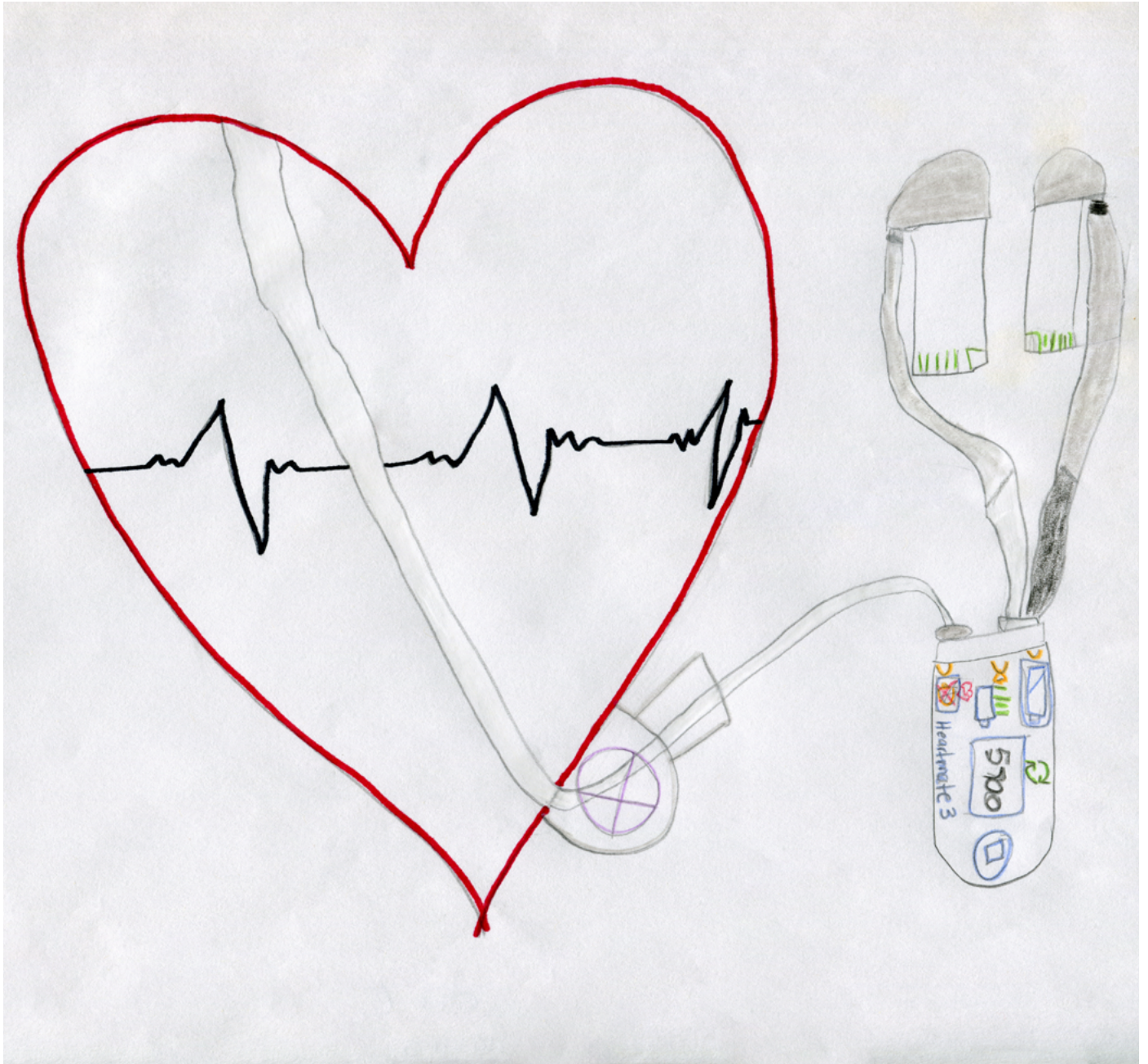
#### V. Heart Failure: Diagnostic Testing (Cont'd.)

- **Cardiac catheterization (Heart Cath):** A cardiac catheterization is an invasive procedure that requires anesthesia and a special IV. A thin, flexible tube (catheter) is inserted into a blood vessel in your child's groin, neck, or arm and guided to the heart to measure oxygen levels, pressures, and blood flow inside the heart. Pictures using dye (contrast) may be taken to see certain structures. Some heart problems may be addressed with additional procedures during the catheterization. Your doctor may also obtain small pieces of heart muscle (biopsy) to determine if an infection or certain types of heart muscle disease are causing heart failure. In patients with congenital heart defects, there may be additional testing or procedures that your child's cardiologist or the cath doctor may recommend.
- **Electrophysiology study (EP Study):** If there are concerns about a heart rhythm problem contributing to heart failure symptoms, your child's cardiologist may recommend an EP study. Similar to a cardiac catheterization, thin flexible tubes and wires (catheters) are inserted through veins in the groin or neck and are guided to the heart. Stimulated with electricity, these wires map out the heart's electrical system and test for abnormal electrical pathways, risk of heart rhythm problems, and response to medication through electrical activity. Some rhythm abnormalities may be resolved with an additional procedure done during an EP study. ♦

# VI.

# Medications

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# VI. Medications

## A. Heart Failure Medications

There are several medications used to treat heart failure in infants and children. It is important to familiarize yourself with these medications and understand their purpose and possible side effects.

Regular cardiology appointments are needed to monitor how your child is tolerating the medications. Some medications may require frequent changes in dosage. Other medications may require blood work to be drawn to monitor for side effects.

If your child is too young or cannot take pills, some medications can be made into liquids. This may require the use of special pharmacies called compounding pharmacies. Since only some pharmacies can make liquid compounds, it is important to locate a pharmacy near you that will make these compounds for your child on a regular basis.

The following pages lists many of the common medications used specifically to treat and manage the symptoms of heart failure. Your child may not require all medications listed below. Your child also may be prescribed additional medications or supplements.

## B. Guidelines for Taking Medications

Medication should be given as instructed by your child's cardiologist. It is best to give medicines at the same time every day and in the same way (with or without food), including weekends. If your child misses a dose, give your child the missed dose as soon as you remember. If it is almost time for the next dose, skip the missed dose, and give your child the next dose at the regular time. If you are not sure what to do, call your child's doctor for instructions. If your child spits out or vomits the medication, also call your child's doctor for instructions on what to do.

If your child has reactions or side effects from any medication, your child's cardiologist should be notified immediately. Before you stop or change any of your child's medications, you should consult with your child's health care team.

## C. Angiotensin Converting Enzyme Inhibitors (ACE-inhibitors or ACE-I)

These medications lower blood pressure by relaxing the blood vessels. By relaxing the blood vessels, the heart does not have to work as hard to pump blood to the body. These medicines can help improve heart function over time (called reverse remodeling) by blocking stress hormones.

VI. Medications ▶ C. Angiotensin Converting Enzyme Inhibitors (ACE-inhibitors or ACE-I) (Cont'd.)

<b>Generic Name</b>	<b>Captopril</b>
<b>Brand Name</b>	<b>Capoten</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Lowers blood pressure to improve heart function</li> <li>• Reduces the amount of work the heart needs to beat</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Three or four times daily, same time, every day</li> <li>• Take 1 hour before or 2 hours after meals on empty stomach</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Dry Cough, often persistent</li> <li>• Headache</li> <li>• Tiredness</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Feeling dizzy or lightheaded is common, especially after the first dose. Be prepared to sit down or lie down after taking the medicine.</li> <li>• If your child becomes pregnant, she should stop this medication and speak to her cardiologist as soon as possible as it could injure the fetus.</li> </ul>

<b>Generic Name</b>	<b>Enalapril</b>
<b>Brand Name</b>	<b>Vasotec or Epaned (liquid)</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Lowers blood pressure to improve heart function</li> <li>• Reduces the amount of work the heart needs to beat</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Twice daily</li> <li>• May be given with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Dry cough, often persistent</li> <li>• Headache</li> <li>• Low blood pressure</li> <li>• Feeling dizzy or lightheaded</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Feeling dizzy or lightheaded is common, especially after the first dose. Be prepared to sit down or lie down after taking the medicine.</li> <li>• If your child becomes pregnant, she should stop this medication and speak to her cardiologist as soon as possible as it could injure the fetus.</li> </ul>

VI. Medications ▶ C. Angiotensin Converting Enzyme Inhibitors (ACE-inhibitors or ACE-I) (Cont'd.)

<b>Generic Name</b>	<b>Lisinopril</b>
<b>Brand Name</b>	<b>Prinivil, Zestril</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Lowers blood pressure to improve heart function</li> <li>• Reduces the amount of work the heart needs to beat</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Once daily</li> <li>• May be given with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Dry cough, often persistent</li> <li>• Headache</li> <li>• Feeling dizzy or lightheaded</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Feeling dizzy or lightheaded is common, especially after the first dose. Be prepared to sit down or lie down after taking the medicine.</li> <li>• If your child becomes pregnant, she should stop this medication and speak to her cardiologist as soon as possible as it could injure the fetus.</li> </ul>

<b>Generic Name</b>	<b>Ramipril</b>
<b>Brand Name</b>	<b>Altace</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Lowers blood pressure to improve heart function</li> <li>• Reduces the amount of work the heart needs to beat</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Once daily</li> <li>• May be given with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Dry cough, often persistent</li> <li>• Headache</li> <li>• Feeling dizzy or lightheaded</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Feeling dizzy or lightheaded is common, especially after the first dose. Be prepared to sit down or lie down after taking the medicine.</li> <li>• If your child becomes pregnant, she should stop this medication and speak to her cardiologist as soon as possible as it could injure the fetus.</li> </ul>



VI. Medications (Cont'd.)

## D. Angiotensin Receptor Blockers (ARBs)

These medications are similar to ACE-inhibitors and help blood vessels relax, which decreases the heart’s workload. ARBs may be an alternative for those who cannot tolerate or have side effects from ACE inhibitors.

<b>Generic Name</b>	<b>Losartan</b>
<b>Brand Name</b>	<b>Cozaar</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Reduces the amount of work the heart needs to beat</li> <li>• Lowers blood pressure to improve heart function</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Once daily</li> <li>• May be given with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Diarrhea</li> <li>• Tiredness</li> <li>• Muscle pain</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Feeling dizzy or lightheaded is common, especially after the first dose. Be prepared to sit down or lie down after taking the medicine.</li> <li>• If your child becomes pregnant, she should stop this medication and speak to her cardiologist as soon as possible as it could injure the fetus.</li> </ul>

<b>Generic Name</b>	<b>Valsartan</b>
<b>Brand Name</b>	<b>Diovan</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Reduces the amount of work the heart needs to beat</li> <li>• Lowers blood pressure to improve heart function</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Once daily</li> <li>• May be given with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Tiredness</li> <li>• Headache</li> <li>• Diarrhea</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Feeling dizzy or lightheaded is common, especially after the first dose. Be prepared to sit down or lie down after taking the medicine.</li> <li>• If your child becomes pregnant, she should stop this medication and speak to her cardiologist as soon as possible as it could injure the fetus.</li> </ul>

VI. Medications ▶ D. Angiotensin Receptor Blockers (ARBs) (Cont'd.)

<b>Generic Name</b>	<b>Valsartan + Saubitril</b>
<b>Brand Name</b>	<b>Entresto</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Reduces the amount of work the heart needs to beat</li> <li>• Lowers blood pressure to improve heart function</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Twice a day</li> <li>• May be given with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Tiredness</li> <li>• Headache</li> <li>• Facial swelling</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• If you are switching from an ACE-inhibitor to Entresto, consult your child’s cardiologist before stopping the ACE-inhibitor and starting Entresto.</li> <li>• If your child becomes pregnant, she should stop this medication and speak to her cardiologist as soon as possible as it could injure the fetus.</li> </ul>

## E. Beta Blockers

Beta blockers block specific beta receptors on blood vessels and heart muscle, causing the heart to beat slower and with less force. They are commonly used to lower blood pressure or to control the heart rate when the heart rate is too high. These medications can also improve heart function by blocking the effects of stress hormones on the heart and encouraging a process known as reverse-remodeling.

<b>Generic Name</b>	<b>Carvedilol</b>
<b>Brand Name</b>	<b>Coreg; Coreg CR</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Lowers blood pressure to help improve heart function</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Give twice a day, same time, every day</li> <li>• Give with food or milk</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Dizziness</li> <li>• Drowsiness</li> <li>• Slow heart rates (bradycardia)</li> <li>• Nausea</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Inform your child’s cardiologist if your child has a history of asthma or any problems breathing.</li> <li>• Inform your child’s cardiologist if your child has diabetes, as carvedilol may cause changes in blood sugar levels.</li> </ul>

VI. Medications ▶ E. Beta Blockers (Cont'd.)

<b>Generic Name</b>	<b>Metoprolol</b>
<b>Brand Name</b>	<b>Lopressor, Toprol XL</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Lowers blood pressure to lower heart rate</li> <li>• Controls uneven or fast heartbeats, chest pain, or migraines</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Given once or twice daily</li> <li>• May be given with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Dizziness or lightheadedness</li> <li>• Tiredness</li> <li>• Trouble concentrating, changes in mood, or depression</li> <li>• Nightmares or problems sleeping</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Inform your child’s cardiologist if your child has a history of asthma or any problems breathing.</li> <li>• Inform your child’s cardiologist if your child has diabetes, as metoprolol may cause changes in blood or urine sugar levels. Metoprolol may also mask signs of low blood sugar, such as changes in heart rate.</li> </ul>

<b>Generic Name</b>	<b>Ivabradine</b>
<b>Brand Name</b>	<b>Corlanor</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Slows down the heart rate; is not a true beta blocker</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Twice daily</li> <li>• May be given with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Low heart rate</li> <li>• Diarrhea</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Feeling dizzy or lightheaded is common especially after the first dose. Be prepared to sit down or lie down for some time after taking the medicine.</li> </ul>



VI. Medications (Cont'd.)

## F. Digitalis

Digitalis, or digoxin, is used to treat patients with heart failure. It is sometimes used to treat certain abnormal fast heart rhythms. Digoxin is not used frequently due to newer and safer medications available today, and its use in children is not as common as adults. Slow heart rate leading to heart block is a side effect when levels of digoxin in the blood are too high. It is important to only use digoxin when prescribed by your child’s cardiologist and never change the dose.

<b>Generic Name</b>	<b>Digoxin</b>
<b>Brand Name</b>	<b>Lanoxin</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Improves heart function in symptomatic patients</li> <li>• Controls and slows certain irregular or fast heartbeats</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Twice daily at least 12 hours apart</li> <li>• Give with full glass of water</li> <li>• May be given with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Mild diarrhea</li> <li>• Loss of appetite or upset stomach</li> <li>• Drowsiness</li> <li>• Dizziness</li> <li>• Unusual tiredness or weakness</li> <li>• Headache</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Electrocardiograms (EKG) or blood tests may be requested to ensure appropriate dosing.</li> <li>• If your child’s heart rate seems irregular or slower than usual, contact your child’s cardiologist immediately to see if further evaluation is needed. Other signs of toxicity include loss of appetite nausea, vomiting, and visual changes.</li> </ul>

VI. Medications (Cont'd.)

## G. Diuretics

Diuretics, known as “water pills” are used to help the body get rid of excess salt and water. This decreases the amount of fluid flowing through blood vessels and reduces pressure on the vessel walls. Getting rid of extra fluid build-up in the lungs makes breathing easier and reduces swelling.

There are three common types of diuretics: potassium sparing, thiazide, and loop diuretics. Each type affects a different part of a child’s kidneys and has different uses, side effects, and precautions. All will cause increased urination, but some cause more than others.

<b>Generic Name</b>	<b>Spironolactone or Eplerenone</b>
<b>Brand Name</b>	<b>Aldactone or Inspra</b>
<b>Type</b>	Potassium sparing
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Removes salt and water from the body</li> <li>• Lowers blood pressure and helps retain potassium in the blood</li> <li>• Promotes healing of heart tissue with long term use</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Once or twice daily, same time every day</li> <li>• Give with food to reduce the chance of upset stomach</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Headache</li> <li>• Nausea or vomiting</li> <li>• Increase in serum potassium level</li> <li>• Swelling of the breasts or breast soreness (males and females)</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Spironolactone and eplerenone can raise the level of potassium in your child’s blood to dangerous levels. Talk with your child’s cardiologist if increased potassium is a concern.</li> </ul>

VI. Medications ▶ G. Diuretics (Cont'd.)

<b>Generic Name</b>	<b>Furosemide</b>
<b>Brand Name</b>	<b>Lasix</b>
<b>Type</b>	Loop diuretic
<b>What it does</b>	<ul style="list-style-type: none"> <li>Removes salt and water from the body</li> <li>Lowers blood pressure</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>One to four times daily, same time every day</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>Dizziness when getting up from a lying or sitting position</li> <li>Stomach cramps</li> <li>Headache</li> <li>Dehydration</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>May cause the body to lose potassium.</li> <li>May make your child’s skin more likely to sunburn.</li> <li>Your child’s cardiologist may want to stop this medicine during times of illness if vomiting or diarrhea occur.</li> </ul>

<b>Generic Name</b>	<b>Bumetanide</b>
<b>Brand Name</b>	<b>Bumex</b>
<b>Type</b>	Loop diuretic
<b>What it does</b>	<ul style="list-style-type: none"> <li>Removes salt and water from the body</li> <li>Lowers blood pressure</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>Daily or every other day as prescribed by your child’s cardiologist.</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>Low potassium</li> <li>Headache</li> <li>Muscle pain</li> <li>Dehydration</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>Your child’s cardiologist may want to stop this medicine during times of illness if vomiting or diarrhea occur.</li> <li>Call your child’s cardiologist before stopping this medicine.</li> </ul>

VI. Medications ▶ G. Diuretics (Cont'd.)

<b>Generic Name</b>	<b>Chlorothiazide</b>
<b>Brand Name</b>	<b>Diuril</b>
<b>Type</b>	Thiazide diuretic
<b>What it does</b>	<ul style="list-style-type: none"> <li>Removes salt and water from the body</li> <li>Lowers blood pressure</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>Daily or every other day as prescribed</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>Low potassium</li> <li>Headache</li> <li>Muscle pain</li> <li>Dizziness</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>Your child’s cardiologist may want your child to stop taking this medicine during times of illness when vomiting or diarrhea occur. Call your child’s cardiologist before stopping this medicine.</li> </ul>

## H. Inotropes

These are intravenous (IV) medications given through a central line to improve heart pumping and maintain appropriate blood pressure.

Inotropes are generally used for patients who continue to have symptoms on oral medications and require more intense therapies to support their heart function.

<b>Generic Name</b>	<b>Milrinone</b>
<b>Brand Name</b>	<b>Primacor</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>Lowers blood pressure to improve heart function</li> <li>Reduces the amount of work the heart needs to beat</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>Intravenously through a peripheral IV or central line</li> <li>Used inpatient, and in some cases as an outpatient via pump or PICC line</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>Headache</li> <li>Chest pain</li> <li>Low blood pressure</li> <li>Rapid heart rate</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>If your child is receiving milrinone through an IV, and the IV becomes damaged (leaking, disconnected, starts to bleed back), you should contact your local hospital immediately.</li> <li>Your child’s cardiologist should be alerted if there is redness, tenderness, or drainage from the central line site or your child experiences chest pain, fast or uneven heart beats, lightheadedness or fainting, trouble breathing, hives, or a severe rash.</li> </ul>



VI. Medications ▶ H. Inotropes (Cont'd.)

<b>Generic Name</b>	<b>Dopamine</b>
<b>Brand Name</b>	<b>Intropin, Dopastat</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• At low doses, makes the heart pump better and increases heart rate</li> <li>• At higher doses, causes blood vessels to constrict, and increases blood pressure</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Intravenously through a central line</li> <li>• Typically, inpatient use only</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Headache</li> <li>• Nausea or vomiting</li> <li>• Tingling feeling, goosebumps or “adrenaline rush”</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Your child’s cardiologist should be alerted if your child experiences chest pain, fast or uneven heart beats, lightheadedness or fainting, trouble breathing, hives, or severe rash.</li> <li>• Your child’s cardiologist will be monitoring your child’s reaction to the medicine and will be titrating to the dose that is right for your child.</li> </ul>

<b>Generic Name</b>	<b>Dobutamine</b>
<b>Brand Name</b>	<b>Dobutrex</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Lowers blood pressure to improve heart function</li> <li>• Reduces the amount of work the heart needs to beat</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Intravenously through a central line</li> <li>• Typically, inpatient use only</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Headache</li> <li>• Nausea or upset stomach</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Your child’s cardiologist should be alerted if you child experiences slow, fast, or irregular heartbeats, chest pain, trouble breathing, hives, or severe rash.</li> <li>• Your child’s cardiologist will be monitoring your child’s reaction to the medicine and will be titrating to the dose that is right for your child.</li> </ul>

VI. Medications ▶ H. Inotropes (Cont'd.)

<b>Generic Name</b>	<b>Epinephrine</b>
<b>Brand Name</b>	<b>Adrenalin</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Makes the heart pump better and increases heart rate</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Intravenously through a central line</li> <li>• Typically, inpatient use only.</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Feeling anxious, nervous, scared, or weak</li> <li>• Headache or dizziness</li> <li>• Pale skin</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Your child’s cardiologist should be alerted if your child experiences chest pain, fast, pounding or uneven heartbeat, difficulty breathing, heavy sweating, nausea or vomiting, pain or, redness or warmth at the injection site.</li> <li>• Your child’s cardiologist will be monitoring your child’s reaction to the medicine and will be titrating to the dose that is right for your child.</li> </ul>

## I. Anticoagulation

These medications are used to thin the blood and to reduce the risk for clots which can block the blood flow to other organs such as the heart and brain.

Anticoagulation medications will cause your child to bruise and bleed more easily, and it may become difficult to control bleeding. When your child is taking these medications special caution is needed to prevent bleeding and injury.

<b>Generic Name</b>	<b>Aspirin or Acetylsalicylic Acid (ASA)</b>
<b>Brand Name</b>	<b>Various</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Treats or prevents blood clots from forming. In rare cases, a stroke may occur if these blood clots break loose and travel to the brain vessels.</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Give with food or milk to prevent upset stomach</li> <li>• Enteric-coated ASA should be swallowed whole, not crushed or chewed</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Rash or itchy skin</li> <li>• Nausea or vomiting</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Contact your child’s cardiologist immediately if your child experiences itching, swelling chest tightness or difficulty breathing, bloody or black stool, bloody vomit or vomit that looks like coffee grounds, excessive bleeding, bruising or weakness, severe stomach pain, or ringing in the ears.</li> </ul>

VI. Medications ▶ I. Anticoagulation (Cont'd.)

<b>Generic Name</b>	<b>Warfarin</b>
<b>Brand Name</b>	<b>Coumadin</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Treats or prevents blood clots from forming. In rare cases, a stroke may occur if these blood clots break loose and travel to the brain vessels.</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Take with or without food</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Bleeding or bruising more easily</li> <li>• Headache</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Warfarin begins to work within 24 hours after the first dose, but the full effect may take 3-4 days to occur. The amount of warfarin your child needs depends on how his or her body responds to the drug and the results of a blood test called an INR.</li> <li>• Vitamin K can reverse the effects of warfarin and can be found in common foods, multivitamins, and supplements.</li> <li>• Avoid contact sports.</li> <li>• Contact your child’s cardiologist if your child experiences itching, swelling, chest tightness or difficulty breathing, excessive or uncontrolled bleeding, black or bloody stools, weakness on one side of the body, chest pain, dizziness, fainting, lightheadedness, severe headache, or change in skin color.</li> </ul>

<b>Generic Name</b>	<b>Enoxaparin</b>
<b>Brand Name</b>	<b>Lovenox</b>
<b>What it does</b>	<ul style="list-style-type: none"> <li>• Treats or prevents blood clots from forming. In rare cases, a stroke may occur if these blood clots break loose and travel to the brain vessels.</li> </ul>
<b>How it is given</b>	<ul style="list-style-type: none"> <li>• Given by injection under the skin</li> </ul>
<b>Most common side effects</b>	<ul style="list-style-type: none"> <li>• Bleeding or bruising more easily</li> <li>• Nausea</li> <li>• Diarrhea</li> <li>• Pain, redness, or irritation at the site of medicine injection</li> </ul>
<b>Other important information</b>	<ul style="list-style-type: none"> <li>• Avoid contact sports.</li> <li>• Contact your child’s cardiologist if your child experiences itching, swelling, chest tightness or difficulty breathing, excessive or uncontrolled bleeding, black or bloody stools, weakness on one side of the body, chest pain, dizziness, fainting, lightheadedness, severe headache, or change in skin color.</li> </ul>

# VII.

# Advanced Heart Failure Therapies

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## VII. Advanced Heart Failure Therapies

Advanced therapies refer to treatments for heart failure other than medications or corrective surgery. Advanced therapies are used when standard medical therapies are no longer working to lessen or control the symptoms of heart failure. Commonly, these are machines or devices which help the heart function. They can be broken into two broad categories: machines that increase or take over the job of pumping blood, known as “mechanical circulatory support”, and those that address electrical or rhythm problems of the heart. If your child still has symptoms of heart failure that cannot be managed with medications, then he or she may be referred for a heart transplant evaluation. Often, cardiologists that manage children with heart failure also take care of children who need or have had a heart transplant. They will be able to discuss the transplantation process in more detail with you and your child.

### A. Mechanical Circulatory Support

Mechanical circulatory support refers to the use of a machine that assists your child’s heart in pumping blood. These are used when the heart is no longer able to supply adequate blood flow by itself. This may happen because of an abnormality in part of the heart or a disease of the heart muscle itself, causing it to become too weak to pump adequately. These circulatory assist machines work to either supplement or completely replace blood flowing out of the heart.

There are a number of devices currently being utilized, and more devices are being developed. Some are completely outside the body while others are contained within the chest with minimal external components. The below provides a brief overview of the different types of devices.

**Extracorporeal Membrane Oxygenation or “ECMO”:** This is a large machine that sits next to the patient and takes blood out of the big veins and pumps it back into the large arteries through large tubes (catheters) inserted into the patient. Another function of this machine is that it can put oxygen into the blood and is useful for patients who also have lung problems. This machine acts the same way as a heart-lung bypass used during open heart surgery. The attachment to the ECMO system requires a small operation, as does the disconnection.

**Ventricular Assist Device or “VAD”:** This is a machine which takes blood from the heart and pushes it out to the body. One way in which a VAD is different than ECMO is that a VAD does not include a way for oxygen to be put into the blood. A patient on a VAD will need their lungs to provide oxygen. When a VAD is used to support the left side of the heart (left ventricle), it can be referred to as an LVAD. When used to support the right side of the heart (right ventricle), it is called an RVAD. When two devices are simultaneously used to support both sides of the heart, it may be referred to as a BiVAD. A VAD may pump blood continually (continuous flow device) or it may fill and empty rhythmically (pulsatile flow device).

There are a number of devices made by a variety of manufacturers. The choice of which device to use depends on a number of factors, including your child’s weight, exact reason for heart failure, and local expertise. Some patients have the device outside the body with only the thin tubes inserted into a vein or body (cannulas) internalized, while other patients have devices that are contained within the chest with only the power line externalized.

## VII. Advanced Heart Failure Therapies ▶ A. Mechanical Circulatory Support (Cont'd.)

A benefit of using a VAD is that patients are allowed to be mobile. The level of mobility is dependent on the size of the device, how it is placed, and how strong a child is.

There are two goals in using mechanical circulatory support. They are used to help the patient get stronger for some type of treatment that will help the heart recover. In this case, it is considered a “bridge to recovery.” For patients who are waiting for a heart transplant and their heart is too weak to support them, a VAD may be used until a donor heart becomes available. In this case, the purpose of mechanical support is a “bridge to transplant.”

## B. Electrical Therapies

**Implantable Cardioverter Defibrillator (ICD).** In some patients, the heart muscle is so weak that it can begin to beat abnormally. This may lead to a variety of arrhythmias, which can cause decreased ability to pump blood, or sudden death. An implantable cardioverter defibrillator (ICD) is a device in which leads are directly attached to the heart allowing it to constantly monitor the heart rhythm and deliver an electrical shock if needed. The most commonly used devices are slightly smaller than a deck of cards and are implanted under the skin. Most of these devices can also act as a pacemaker, stimulating the heart to beat if the heart rate gets too slow or there is a problem with electricity passing from the top to the bottom of the heart.

The placement of this device will vary depending on your child’s age. In adults and larger children, the ICD is placed just beneath the skin right under the collarbone with the wires running through a vein and into the heart. In smaller children, the device is placed under the skin at the abdomen with the wires track under the skin to the chest and are sewn to the outer surface of the heart.

A newer type of ICD is known as a subcutaneous array. This device and its wires are placed just beneath the skin and the wires wrap around the heart. This device can deliver shocks to stop an abnormal heart rhythm, but it cannot act as a pacemaker.

**Resynchronization Therapy.** When the heart is caused to beat by external devices, the normal conduction system or internal wiring is not utilized. The heart muscle may become so weak that the normal electrical pathways do not work properly. This causes different parts of the ventricle to squeeze at different times leading to less blood being pumped forward with each heartbeat.

Resynchronization therapy involves using a pacemaker to electrically stimulate both sides of the heart simultaneously, so that both ventricles contract together and increase the volume of blood pumped. ♦

# VIII.

# Additional Resources

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# Terms and Definitions

**ACE-inhibitor (angiotensin converting enzyme inhibitor):** A class of medications that help to relax blood vessels and lower blood pressure, making it easier for the heart to pump blood. Examples include enalapril, lisinopril, and captopril.

**Automated external defibrillator (AED):** A machine used to shock the heart into a normal rhythm in the event of a sudden cardiac arrest. AEDs are located in the majority of public places and schools.

**Antibiotic/antimicrobial:** Medications that help treat infections.

**Aorta:** The main artery coming from the heart that supplies oxygen-rich blood to the body. It normally comes off the left ventricle of the heart.

**Aortic valve:** The valve between the left ventricle and the aorta.

**Arrhythmia:** An abnormal heart rhythm or heartbeat.

**Arrhythmogenic cardiomyopathy:** A rare form of cardiomyopathy in which the normal heart muscle gets replaced by fat and scar tissue. This can lead to dangerous heart rhythms, dilation of the ventricles, and poor heart function. It is known as arrhythmogenic right ventricular dysplasia (ARVD) or cardiomyopathy (ARVC) because it more commonly involves the right ventricle.

**Arterial line:** An IV catheter that is placed in an artery rather than a vein to continuously measure blood pressure and to draw blood for lab tests.

**Artery:** A blood vessel that carries blood away from the heart.

**Atresia:** Blocked or missing, most commonly in reference to blood vessels.

**Atrial septum:** The wall dividing the two upper collecting chambers of the heart (atria).

**Atrioventricular node (AV Node):** Part of the electrical pathway of the heart, which transmits a spontaneous electrical impulse from the atria to the ventricles with each heartbeat.

**Atrium (plural = atria):** The upper chamber of the heart that collects oxygen-poor (blue) blood from the body and delivers it to the heart's right ventricle. The left atrium collects oxygen-rich (red) blood from the lungs and delivers it to the heart's left ventricle.

**Beta Blocker:** A class of medications that lowers the heart rate and blood pressure by blocking the heartbeat effects of the hormone adrenaline. Examples include carvedilol, metoprolol, atenolol, and nadolol.

**BiPAP/CPAP:** Pressurized air and oxygen that is administered through a mask, which is fitted over the nose or nose and mouth, to assist in breathing.

**Blood clot/thrombosis:** A clump of blood that forms inappropriately in an artery or vein that reduces blood flow.

**Blood pressure:** The force of the blood in the arteries. Systolic blood pressure is the top number when the heart is contracted. Diastolic blood pressure is the bottom number when the heart is relaxed.

**Blood thinners:** Medications that thin the blood to prevent blood clots. Examples include aspirin, warfarin, enoxaparin, clopidigrel, and dipyridamole.

**Bradycardia:** An abnormally slow heart rate.



### Terms and Definitions (Cont'd.)

**Cannula:** A thin tube inserted into a vein or body cavity to administer medicine, drain off fluid, or insert a surgical instrument.

**Cardiac catheterization:** A test used to diagnose and/or treat some cardiac abnormalities. Long catheters are inserted through veins and arteries (usually in the groin or neck) into the heart and large blood vessels to take pictures (angiography), and measure pressures and oxygen levels (hemodynamics).

**Cardiopulmonary Stress Test:** A test typically done on a stationary bike or treadmill that determines how well the heart and lungs work during exercise.

**Catheter:** A small plastic tube that is inserted through veins and arteries. See **cardiac catheterization** definition above.

**Cardiac Magnetic Resonance Imaging (MRI):** A type of non-invasive diagnostic test which provides three dimensional pictures of the heart and blood vessels, as well as information about the function of the heart chambers and valves. It often requires special dye to be injected through an IV catheter.

**Cardiac resynchronization (CRT):** A procedure that involves placement of a special pacemaker with leads on several parts of the heart to improve the way the heart squeezes and contracts.

**Cardiologist:** A doctor that is trained to take care of patients with heart problems.

**Cardiomyopathy:** A condition in which the heart muscle is abnormal and the pumping function is affected. There are a variety of types, causes, symptoms, and treatments.

**Cardiothoracic (CT) Surgeon:** A surgeon that operates on the structures in the chest, including the heart and blood vessels. Pediatric CT surgeons have special training to operate on infants and children with heart problems.

**Central line:** An intravenous (IV) catheter that enters a large vein leading to the heart. It is used to administer medications and may be used to measure heart pressure and to draw blood.

**Congenital heart defect/congenital heart disease (CHD):** An abnormality of the heart and/or blood vessels around the heart that is present at birth. Some defects are mild and can be repaired with surgery. More complex and single ventricle defects cannot be repaired and are more likely to cause heart failure.

**Congestive heart failure (CHF):** A condition in which the heart cannot pump well enough or relax enough to allow filling. This creates a backup of blood and congestion in the veins, the liver, and/or the lungs.

**Chest x-ray (CXR):** A picture that shows an image of the heart, lungs, blood vessels, and diaphragm as well as the bones of the chest.

**Chest tube:** A tube that is placed in the chest to drain fluid from outside the lungs or heart.

**Cyanosis:** Blue color of the skin and mucous membranes caused by a low level of oxygen in the blood.

**Dilated cardiomyopathy (DCM):** A type of cardiomyopathy in which the heart muscle becomes enlarged (dilated) or stretched, ultimately making the heart weak and unable to pump blood well.

**Dialysis:** A process of removing waste products and excess fluids from the body when the kidneys are not working well.

**Diuretic:** Medications (“water pills”) that help the body eliminate excess fluid by increasing the amount of urine the kidneys produce. Examples include furosemide, chlorothiazide, spironolactone, torsemide, and bumetanide.

**Electrocardiogram (ECG or EKG):** A test that records the electrical activity of the heart. It is used to detect heart problems and monitor the heart’s status.

### Terms and Definitions (Cont'd.)

**Electrolytes:** Substances in the blood and body tissues that help regulate how the cells in our bodies function. Examples of electrolytes are potassium, sodium, chloride, magnesium and calcium.

**Echocardiogram (ECHO):** A diagnostic test that uses sound waves (ultrasound) to evaluate the structure and function of the heart and blood vessels.

**Extracorporeal membrane oxygenation (ECMO):** A heart and lung life support machine which temporarily pumps blood and regulates the oxygen and carbon dioxide content of the blood when the heart and lungs are not functioning well.

**Failure to thrive (FTT):** Poor growth and poor weight gain that is below what is expected for a person's age. FTT can sometimes be seen with significant heart problems.

**Feeding Tube:** A soft plastic tube through which breast milk or a continuous feeding formula can be administered to provide nutrition when a person is unable to eat on their own.

- **Nasogastric Tube (NG tube):** a tube that is placed through the nose into the stomach to allow extra air or fluid to be removed from the stomach or for breast milk or formula to be given.
- **Naso-Duodenal or Transpyloric Tube (ND or TP tube):** a tube that is placed through the nose into the duodenum, the part of the intestines that is immediately after the stomach. This allows administration of continuous formula feeding for infants and children who are not tolerating feeding into their stomach.
- **Gastrostomy tube (G-tube):** A feeding tube inserted surgically directly into the stomach for longer term use to deliver breast milk or formula, as well as medications.

**Fontan procedure:** a surgical procedure used for children who were born with only one functioning ventricle. It involves redirecting blood flow from the lower body to the lungs without having to go to the heart.

**Fontan failure:** A term used when patients develop significant complications after the Fontan procedure. Examples include congestive heart failure, protein losing enteropathy (PLE), and plastic bronchitis.

**Heart failure:** See **Congestive heart failure**

**Hypertension (HTN):** High blood pressure in the arteries that carry blood from the heart to the body.

**Hypertrophic cardiomyopathy (HCM):** A type of cardiomyopathy in which the heart muscle is thickened and stiff, making it difficult for the heart to relax and fill. It may lead to abnormal and life-threatening heart rhythms.

**Implantable cardioverter defibrillator (ICD):** A surgically implanted device that can deliver an electric shock to the heart if a dangerous abnormal heart rhythm is detected. Some ICD's can function as pacemakers as well.

**Inotropes:** Medications administered continuously through an IV to help the heart work better and support the blood pressure when it is too low. Examples include epinephrine, dopamine, dobutamine, and milrinone.

**Insufficiency/regurgitation:** The abnormal flowing of blood backwards through a heart valve.

**Left Ventricle (LV):** One of the two lower pumping chambers of the heart. The LV receives oxygenated blood from the left atrium via the mitral valve, then pumps blood through the aortic valve to the aorta which carries blood to the body.

### Terms and Definitions (Cont'd.)

**Left Ventricular Non-Compaction cardiomyopathy (LVNC):** A type of cardiomyopathy in which the left ventricle (the main heart pumping chamber) does not form correctly into a compact layer of muscle but instead is thicker than usual and has finger-like muscle projections known as trabeculations. The abnormally formed muscle may not squeeze normally. LVNC can be seen in combination with dilated cardiomyopathy, hypertrophic cardiomyopathy or in combination with some forms of congenital heart defects.

**Mechanical ventilation:** A machine (ventilator) that mechanically delivers oxygen and air through an endotracheal (breathing) tube. Breathing tubes are inserted through the mouth or nose into the airway to assist breathing in sick or sedated patients.

**Mitral Valve:** The valve between the left atrium and left ventricle, which opens to allow blood to flow forward from the atrium to the ventricle. As the ventricle squeezes to pump blood forward, the mitral valve closes to prevent blood from flowing backwards. The valve has two moving parts which are called leaflets.

**Magnetic resonance imaging (MRI):** A test that uses a special powerful magnet, rather than radiation, to provide images (pictures) of a portion of the body. These images allow doctors to evaluate the structure and function of the body part.

**Murmur:** A sound heard when listening to the heart. Some murmurs are normal and others are caused by abnormal blood flow in the heart. Normal murmurs sound different from abnormal murmurs.

**Myocarditis:** Inflammation of the heart muscle, often caused by a virus or bacteria. It can lead to dilated cardiomyopathy.

**Pacemaker:** A device which senses the heart rhythm and can stimulate the upper and/or lower chambers of the heart to beat normally. A pacemaker may be connected to the heart chambers during heart surgery by wires that exit the skin (temporary pacemaker). They may also be implanted under the skin (permanent pacemaker).

**Pericarditis:** Inflammation of the thin sac that surrounds the heart.

**Pericardial effusion:** Abnormal amount of fluid within the sac (pericardium) that surrounds the heart. A large pericardial effusion may impact the function of the heart and therefore may need to be drained.

**Peripherally inserted central catheter (PICC) line:** A long, thin catheter placed in a vein in the foot, hand or arm that ends in the large blood vessels near the heart. It can be left in place for weeks to months for long term infusions, IV nutrition, pressure monitoring, and lab draws.

**Plastic bronchitis:** A rare complication following Fontan surgery for single ventricle heart disease in which abnormal lymphatic flow causes buildup of lymph fluid in the airways creating “casts.” This blocks normal air flow causing difficulty in breathing and may, at times, require a ventilator and/or a procedure to remove a cast.

**Protein Losing Enteropathy (PLE):** A rare complication, usually in single ventricle patients following Fontan operation, in which protein is lost in the stools. Protein loss can cause a number of problems in the body including swelling, clotting and immune problems.

**Pulmonary edema:** “Fluid in the lungs” caused by the backup of blood into the lungs from high pressures in the heart or when the heart does not pump well. It is treated with diuretics.

**Pulmonary hypertension:** High blood pressure in the vessels (pulmonary arteries) that carry blood from the heart to the lungs.

**Pulmonary valve:** The valve between the right ventricle and the main pulmonary artery.

**Pulmonary veins:** Four blood vessels (usually two from each lung) that return oxygen-rich blood from the lungs to the left atrium of the heart.

### Terms and Definitions (Cont'd.)

**Restrictive cardiomyopathy (RCM):** A type of cardiomyopathy in which the walls of the ventricles (lower chambers of the heart) are abnormally stiff and the heart cannot relax well after pumping to allow the heart to fill back up with enough blood.

**Right Ventricle (RV):** One of the two lower pumping chambers of the heart. The RV receives blood from the right atrium via the tricuspid valve and then pumps blood into the pulmonary valve through the artery to the lungs.

**Single Ventricle (SV):** A complex form of congenital heart disease in which only one of the ventricles is completely formed and functions as the main pumping chamber to get blood to the body. Single ventricle heart disease requires open heart surgery.

**Single Ventricle Palliation:** A series of palliative surgeries for patients with a single ventricle that can improve quality and length of life. The number and type of surgeries depends on the specific heart disease of the child.

**Sonographer:** A medical professional who performs ultrasounds.

**Stent:** A small metallic mesh tube used to enlarge a narrowed structure or blood vessel and hold it open. Stents are usually placed using special catheters via a cardiac catheterization procedure.

**Stenosis:** A narrowing of a blood vessel or valve, which causes problems with blood flow.

**Tachycardia:** An abnormally fast heart rate.

**TPN (total parenteral nutrition):** A solution of sugar, protein, electrolytes and sometimes fats (lipids) that is administered through a central line to provide nutrition for patients not tolerating feedings into their stomach or intestines.

**Tricuspid valve:** The valve between the right atrium and the right ventricle.

**Ventricle:** The lower pumping chambers of the heart. The right ventricle pumps oxygen-poor blood to the lungs, and the left ventricle pumps oxygen-rich blood to the body.

**Ventricular assist device (VAD):** A device that is inserted inside or outside the chest and connected to the heart and major blood vessels. The device can be used short or long term to assist a damaged or weakened heart in pumping blood until the heart recovers or a heart transplantation is performed. There are different types of ventricular assist devices that may be used or considered based on a patient's size and anatomy.

- LVAD (left ventricular assist device): a device is used only to support the failing left ventricle
- RVAD (right ventricular assist device): a device used to support a failing right ventricle.
- BiVAD (bi-ventricular assist device): two devices are used, one for each failing ventricle.

**Ventricular septum:** The wall dividing the lower two chambers of the heart (right ventricle and left ventricle). ♦

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