Single Ventricle Survivorship Clinic

at the Heart Center

Intermountain
Primary Children's Hospital
The Child First and Always®

HEALTH
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Dear patients and families,

Medical advances, including the Fontan operation, have greatly improved the survival rate for children born with single ventricle heart disease. The majority of these children are now expected to survive into adulthood. However, there is still no cure for children born with single ventricle defects. As they age, these children will face many health issues and need lifelong specialized care and support to deal with rather than overcome these health issues and live as normal a life as possible.

To help single ventricle patients live a longer and healthier life, healthcare providers must focus on health issues affecting organ systems in addition to the heart. We now know that liver problems, poor bone health, growth issues, delayed physical maturation, blood clotting problems, and abnormal kidney function are common and sometimes inevitable after Fontan surgery. Patients also often face developmental and mental health issues, and lung and breathing problems. These health issues often begin and develop without any warning signs or symptoms. Sometimes, by the time patients have symptoms the disease has often progressed to an irreversible and advanced stage. Careful follow-ups including early, appropriate and periodic tests is the best way to identify, closely follow, and manage these health problems. This clinic embodies the principles of vigilance, innovation, and holistic care in improving the health of single ventricle patients.

The Heart Center at Primary Children's Hospital and the University of Utah offers many years of experience, expertise and innovation in treating patients with single ventricle heart disease. We were among the first centers in the world to develop a state-of-the-art multispecialty clinic for screening and evaluating Fontan patients. This clinic is comprised of multiple specialists, who provide comprehensive care during a single visit. The team members in this clinic are passionate about and committed to caring for Fontan patients.

We hope this guide helps you understand some of the major health concerns for Fontan and single ventricle patients and the reasons tests and evaluations are performed in the single ventricle clinic. Our healthcare providers recognize the burden of health issues you and your child face as well as your time and financial commitments. We regularly reassess and evaluate the need for these tests, based on emerging evidence, and are available to discuss all questions and concerns with you. *We are committed to working closely with you, your primary cardiologist, and your primary care provider to improve all single ventricle patients' health and well-being.*

Sincerely,

On behalf of the entire Heart Center Team
at Primary Children's Hospital, Utah.

Shaji Menon, MD, MS
Single Ventricle Survivorship Clinic Medical Director
Associate Professor of Pediatrics and Radiology
Pediatric Cardiology
University of Utah
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Heart Center

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What is a single ventricle heart defect?

Single ventricle heart defects are rare problems that affect the lower chambers of the heart. With these defects, one of the lower chambers (ventricles) may be smaller, underdeveloped, or missing.

The small lower chamber (ventricle) may not be able to handle pumping blood to either the lungs (to pick up oxygen) or to the body (supplying oxygen and nutrients to the body's tissues). They include defects like:

- **Hypoplastic left heart syndrome (HLHS):** Underdeveloped left side of the heart, including a too-small aorta, mitral valve and left ventricle.
- **Pulmonary atresia with intact ventricular septum:** The pulmonary valve failed to develop normally, and this results in a small (hypoplastic) right ventricle that can't pump sufficient blood into the lungs. This defect is often associated with abnormalities of the coronary arteries (arteries that supply blood to the heart).
- **Tricuspid atresia:** The tricuspid valve is underdeveloped or absent, and the right ventricle is small (hypoplastic). The pulmonary or aortic valve may be small or absent.

There are other heart defects that may qualify as single ventricle heart defects, like double inlet left ventricle, double outlet right ventricle, and other defects that cannot be fixed by other types of surgery.

Often, a doctor can diagnose a single ventricle heart defect when the mother has a fetal ultrasound. Otherwise, it may be diagnosed after the baby is born. After birth, the baby may appear blue or ashen, have rapid and labored breathing, and have difficulty feeding. This heart defect is usually fatal within weeks to months of life unless treated.

Understanding the Fontan operation

Children with a single ventricle heart defect are born with 1 effective pumping chamber (ventricle) instead of 2. This defect is one of the most serious congenital heart problems. It has no cure, but some babies can be treated with a series of operations or heart transplantation. Patients born with single ventricle heart defects require a lifetime of medical care.

The first challenge was survival

Forty years ago, most babies born with a single ventricle died in infancy. Now, thanks to advances in medical care and technology, most live to adulthood. To survive the first few years of life, these patients often have multiple open-heart surgeries and typically have a Fontan operation. This allows them to survive with only one pumping ventricle.

The goal of the Fontan operation is to improve oxygen levels by diverting venous (blue) blood directly to the lungs without a pump. This separates venous (blue) and arterial (pink) circulations and reduces the workload on the heart. The one good ventricle is used as the “pump” for driving blood through the body circulation. However, this leads to increased pressure and backup of blood in the veins and less-efficient circulation.

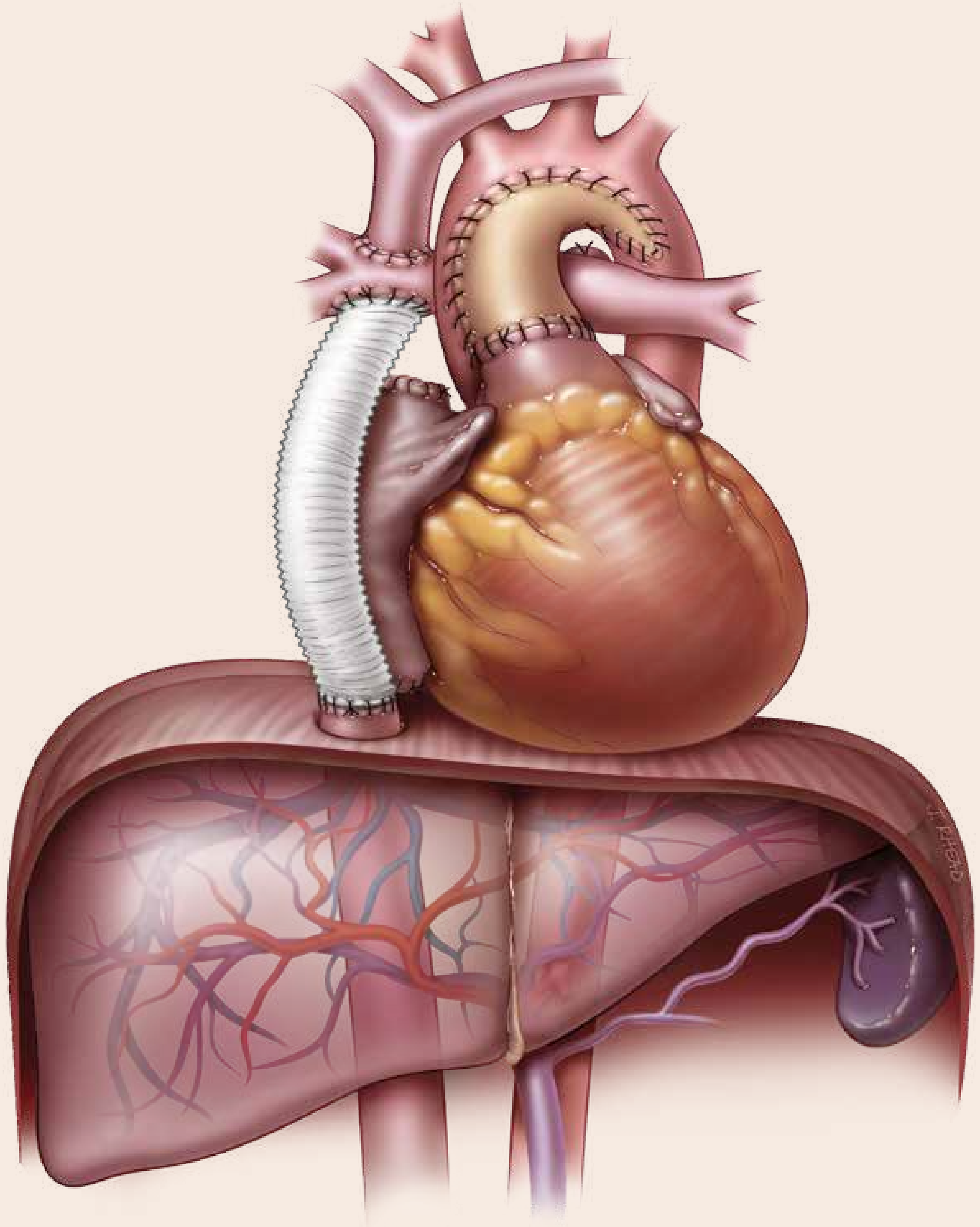
The Fontan operation can be done in several different ways. At Primary Children’s Hospital and other leading centers around the world, the most common Fontan operation is an Extracardiac Fontan. This surgery makes sure your child’s lungs have as much blood flow as possible.

Patients living with the Fontan circulation will usually face a shorter life expectancy and are at risk for lifelong medical problems.
These *may* include:

- Liver problems
- Reduced exercise capacity
- Heart rhythm abnormalities
- Decreased heart function
- Softened bones
- Need for heart transplant
- Delayed sexual maturation
- Short stature
- Abnormal clotting
- Protein-losing enteropathy (losing proteins from the digestive tract)
- Plastic bronchitis (fluid that builds up and hardens in the airways and lungs)

The challenges don’t end with a Fontan operation. Although the Fontan operation is not a perfect operation, at this point it’s the best available solution for most patients born with single ventricle heart defects.

Extracardiac Fontan



Attending the single ventricle survivorship clinic

In the last four decades, survival rates of patients born with single ventricle heart defects have dramatically improved.

The majority of children born with single ventricle heart defects are now expected to survive into adulthood. However, with improved survival, we have noticed many other non-heart related health problems and complications involving other systems. The focus is now on improving the overall quality of life and health of survivors.

This requires careful proactive surveillance and testing to identify early warning signs of complications and health issues involving multiple organs.

The Heart Center at Primary Children's Hospital and the University of Utah was one of the first centers in the world to offer a single ventricle survivorship clinic. This clinic allows your child to see multiple specialists, who evaluate their overall health.

A cardiologist alone cannot evaluate and manage all the health issues of a Fontan patient. This requires a team of specialized doctors from different specialties.

The single ventricle survivorship clinic team is not intended to take over your cardiac care. We work closely with your primary cardiologist to make treatment changes that will improve your health.

Patients with single ventricle heart disease may face health problems involving many different organs and systems. The single ventricle survivorship clinic helps these patients manage all the health problems related to their diagnoses, achieve good health, and enjoy life. Since the health issues Fontan patients experience can vary, the single ventricle survivorship clinic involves specialists from different medical fields to provide patient-centered, collaborative evaluation and treatments.

The process

Patients can attend the single ventricle survivorship clinic every 2 to 3 years (or more frequently if needed), starting at 5 years of age, in addition to their regular cardiology visits. The clinic will work closely with each patient's primary cardiologist and primary care provider to discuss the results, recommendations, and treatments.

Cardiologists may refer a child to the clinic earlier than 5 years old if they develop problems like protein-losing enteropathy (disease of the intestine), plastic bronchitis, liver disease, or Fontan failure.

Why are so many tests needed when patients feel fine?

Because changes in organs may occur and progress without symptoms, patients with single ventricle heart disease often feel fine until it's too late. For example, all Fontan patients will have significant liver abnormalities (such as scarring or fibrosis), and the usual blood tests for liver function may not detect these defects. The single ventricle survivorship clinic has developed specialized liver tests to evaluate Fontan patients' liver changes. Instead of waiting until patients have symptoms, healthcare providers want to be proactive and do tests early so patients remain healthy.

We at Primary Children's Hospital are at the forefront of medical care for patients born with single ventricle heart defects. *Some of the tests may pick up abnormalities in different organs that do not have a cure or effective treatment at this time.* However, knowing that an organ is affected helps us perform a closer look before the disease advances too far or becomes untreatable. We may be able to propose novel therapies or consider heart transplantation earlier.

Helping patients get the most appropriate tests

Single ventricle and Fontan treatment has changed a lot in the last decade. Instead of only focusing on heart issues, the clinic evaluates all body systems to improve patients' health and quality of care. Unfortunately, this involves expensive but necessary specialized testing.

Our center works to ensure Fontan patients receive appropriate but not excessive testing. We constantly compare our testing and surveillance strategies to other centers of excellence in the world. Our goal is to be a conservative center and recommend tests that are necessary, reasonable, and appropriate.

To help patients:

1. Healthcare providers tell patients about the tests they've planned when they're scheduled to attend the clinic. If their primary cardiologist or other healthcare provider has recently done any of the tests, the clinic won't repeat them unless absolutely necessary.
2. The single ventricle team will help with the preapproval process, but they won't know what out-of-pocket costs will be. Patients and their families must ask their insurance about copays and deductibles.
3. Experts discuss tests with each other before choosing the right ones for each patient.
4. Primary cardiologists and members of the single ventricle team help patients understand why they need specific tests.

Note: If patients skip a specific test or tests, the clinic may not be able to provide a comprehensive evaluation and treatment plan.

What to expect on the day of clinic

Healthcare providers are committed to making the clinic visits as quick as possible. However, patients meet with 3 to 5 specialists, so the clinic lasts 3 to 5 hours. This allows each specialist to evaluate the patient, review all the test results, discuss the results with the team, and give detailed recommendations about the patient's condition and test results. Patients don't have to make separate appointments and visits with each specialty, and they receive a comprehensive evaluation in a single visit.

Asking questions after clinic visits

Although healthcare providers will try to answer all questions during the clinic visit, patients can sometimes be overwhelmed with information. A coordinator will provide a dedicated phone number to call when patients have questions or concerns.

Your child's team

The single ventricle clinical team members may include these professionals:

- Cardiologist (medical director)
- Liver and gastrointestinal disease specialist
- An endocrinologist
- Lung disease specialist
- Nurse coordinator
- Social worker
- Hematologist
- Radiologist
- Cardiothoracic surgeon

Assessing your child

The single ventricle survivorship clinic focuses on health issues that affect other organs in addition to the heart. Your child may need liver, bone, blood clotting, and kidney tests at the clinic.

Healthcare providers may also evaluate your child's physical and sexual maturation, along with developmental and behavioral health issues. These health problems often begin without signs or symptoms, so it's important to do early, frequent tests to identify and manage them.

> *"As parents, this was a no-brainer. He will not be tested for anything unnecessary, and we have top doctors monitoring Scott's advancement. Scott is very optimistic, not only for his future, but in laying the data and foundation for little ones who may have to go through the same thing he did."*
>
> *– Parent, Single ventricle survivorship clinic*

Heart tests

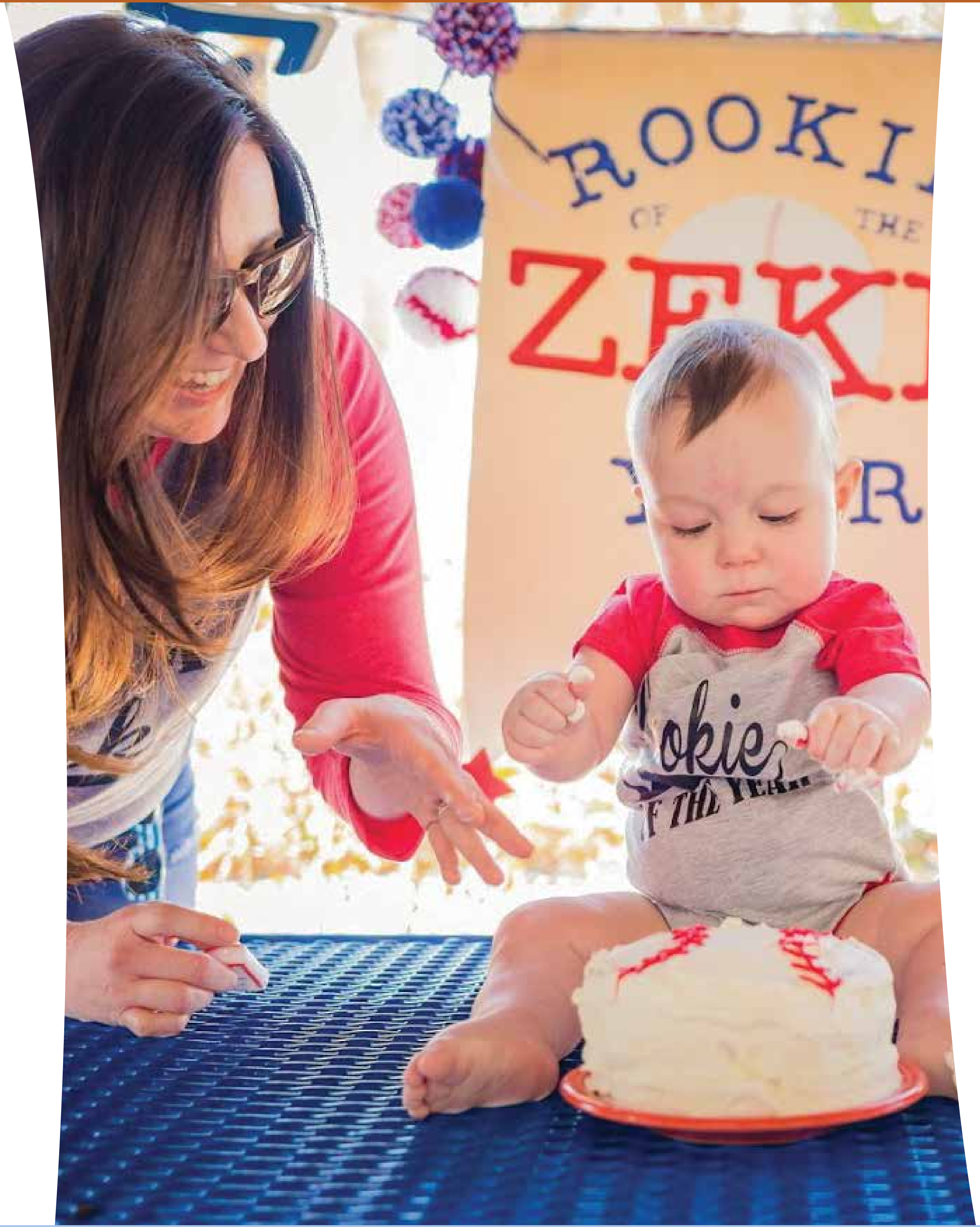
Electrical activity heart tests

Multiple open-heart surgeries for single ventricle heart disease can lead to scarring within the heart. The scarring can cause rhythm and electrical abnormalities in Fontan patients. Some patients will need a pacemaker that generates a low-energy electrical pulse to help the heart beat normally. The clinic will evaluate the heart's rhythm using these screening tests:

- **An electrocardiogram (ECG):** An ECG translates the heart's electrical activity into line tracings on paper to measure heart rhythm and condition.
- **24-hour Holter monitor:** A patient wears electrodes (small electricity-conducting patches) on their chest that are attached to a small recording monitor. They carry the battery-operated monitor in their pocket or a small pouch around their neck or waist.
  - Here's how the Holter monitor works:
    - The monitor records the heart's electrical activity.
    - Patients keep a diary of activities they do while wearing the monitor, noting how they feel.
    - After 24 to 48 hours, patients return the monitor to the clinic.
    - A pediatric cardiologist, who is an expert in evaluating and treating heart rhythm abnormalities, looks at the recordings for any abnormal heart rhythms.
- Pacemaker testing (when indicated).

Heart failure test

Patients will have their B-type natriuretic peptide (BNP) checked with a blood test. BNP is a substance secreted from the heart ventricles if heart failure is developing or getting worse.



Exercise stress test

An exercise stress test is a screening tool that measures how the heart responds to exercise (stress). A patient walks on a treadmill or rides a stationary exercise bicycle. They begin slowly and then walk or pedal faster or on an incline. During the test, an exercise technician monitors the patient's breathing, exhaled gases, electrocardiogram, blood pressure, and oxygen saturation. An exercise stress test can monitor and record changes in a patient's heart performance over many years.

Cardiac catheterization

During a cardiac catheterization, a pediatric cardiologist who specializes in these procedures passes a catheter (thin, flexible tube) into either side of the heart through a blood vessel in the groin, neck, or arm. Most Fontan patients have had catheterization as part of other evaluations. Cardiac catheterization is the only way to precisely measure Fontan circulation and lung artery pressure. High Fontan pressures may indicate problems with the lung or heart function, and patients may need more tests and treatment. Doctors may also be able to improve blood flow or heart function without surgery during cardiac catheterization.

Cardiac MRI and CT scans

Cardiac MRI (magnetic resonance imaging) uses radio waves, magnets, and a computer to create both still and moving pictures of the heart and blood vessels. A cardiac MRI provides more detailed pictures of the beating heart and blood vessels than possible with an Echo (echocardiogram). It is the only noninvasive imaging test that can show healthy heart muscle tissue and help providers see whether the arteries and veins are narrowing.

Unlike CT (computed tomography) scans, an MRI doesn't use ionizing radiation, so it doesn't increase the risk for cancer. Patients who have a pacemaker, ICD (implantable cardioverter defibrillator), or other metal implants may not be able to have an MRI. They can have a CT scan instead.

Liver tests

Liver disease blood tests

Fontan patients can develop liver disease because of multiple surgeries, medicines, veins overfilling with blood, and liver overload because of a missing ventricle. The clinic uses a combination of less invasive blood tests and imaging studies to see if patients need a liver biopsy, which is an invasive test.

These tests include:

- **Liver function/enzyme tests:** These blood tests help to determine liver health by measuring the levels of certain proteins and enzymes in the blood.
- **FibroSure:** This noninvasive liver fibrosis test analyzes biochemicals in combination with age, gender, height, and weight.
- **Complete blood count (CBC):** CBC is one of the most commonly ordered blood tests, because liver damage can affect the blood count.
- **Viral hepatitis and serology tests:** Some Fontan patients need to receive more than 1 hepatitis B vaccination. Viral hepatitis (liver inflammation) can cause liver dysfunction in a liver already affected by Fontan. The clinic does tests to make sure the hepatitis B immunization worked and there isn't another viral infection that could harm the liver.
- **Alpha-fetoprotein (AFP) blood test:** AFP is a protein normally produced by the liver and is used to screen for liver disease and protein-losing intestinal diseases.

Liver biopsy

Liver biopsy is the best way to test for liver disease. Your doctor uses a needle to remove a small piece of the liver and looks at it under a microscope for signs of damage or disease. Patients have this test in a hospital as an outpatient procedure and are given medicines for sedation and pain.
During this test, your child will lie on his or her back as still as possible. The doctor will:

- Clean and numb an area of the abdomen.
- Insert a biopsy needle, usually using an ultrasound to guide it.
- Take a tiny sample of the liver, quickly removing the needle afterwards.
- Bandage the area.

A provider can also do a liver biopsy by inserting a needle into the jugular vein (in the neck) during cardiac catheterization. X-rays are used to guide the needle when doing a liver biopsy this way.

Liver MRI and elastography

MRIs provide detailed images of soft tissues in the body by using radio waves and strong magnets instead of x-rays. When providers use an MRI to look at the liver, they may take several sets of images.

After the first set is done, a provider injects the contrast material called gadolinium [gad-oh-LIN-ee-um] into a vein through an IV (a small tube placed into the vein) before taking the next set of images. This is called a dynamic contrast-enhanced MRI, and it helps providers see liver problems like fibrosis and cirrhosis. It can also help doctors see the difference between benign and malignant tumors and see blood vessels in and around the liver. Liver elastography measures the stiffness of the liver and can evaluate the amount of fibrosis.

Liver CT scan

These x-ray tests produce detailed cross-sectional images of the body. Similar to MRIs, CT scans help providers look for liver problems like fibrosis, cirrhosis, benign and malignant tumors, and nearby blood vessels.

Lung tests

After the Fontan procedure, blood has to flow passively through the lungs without a pump. Since a Fontan circuit's function depends on healthy lungs, patients will also need pulmonary (lung) function tests.

Spirometry

Providers assess the lungs' condition using spirometry, which measures airflow. Patients inhale and exhale after deep breaths, breathe normally and then quietly, and inhale a medicine to see how it changes airflow.

Plastic bronchitis

Plastic bronchitis is a rare but serious lung problem that may develop after the Fontan operation. In plastic bronchitis, the lung can't clear the lymphatic secretions (thick, sticky mucus in the lungs). Because of this, large, rubbery, cheese-like mucus forms within the airways, obstructing the airflow. Patients can have severe respiratory problems, including trouble breathing, low oxygen saturation, and coughing episodes. A pulmonologist at the clinic will evaluate and treat patients with plastic bronchitis. With newer treatment options, some patients with plastic bronchitis may improve without needing a heart transplant.

Bone tests

Bone health blood test

Fontan patients may have weak bones and muscles from too little calcium, phosphorus, and parathyroid hormone (PTH), reduced exercise capacity, and delayed puberty. The clinic will check blood levels of vitamin D and recommend vitamin D supplements if needed.

DXA scan to measure bone health

Patients with the Fontan circulation often have low bone density. Doctors scan with something called DXA, which measures bone density using a small amount of radiation. If patients have low bone density, they're referred to a specialist for more evaluation and treatment.

Bone age and sexual maturity test

Like other chronic diseases, chronic heart disease can delay a child's growth and maturation.

A bone age study estimates a child's skeletal system maturity, which can help doctors diagnose delayed growth and development. To do a bone age study, a healthcare provider takes a low-dose x-ray of the wrist, hand, and fingers.

An endocrinologist assesses sexual maturity and asks patients questions about their body's development. Delayed growth and sexual maturation can be treated with medicine, if needed.

Other tests

Blood clotting

Patients with a single ventricle may have blood clotting problems. They may have clotting and blood component tests at the clinic.

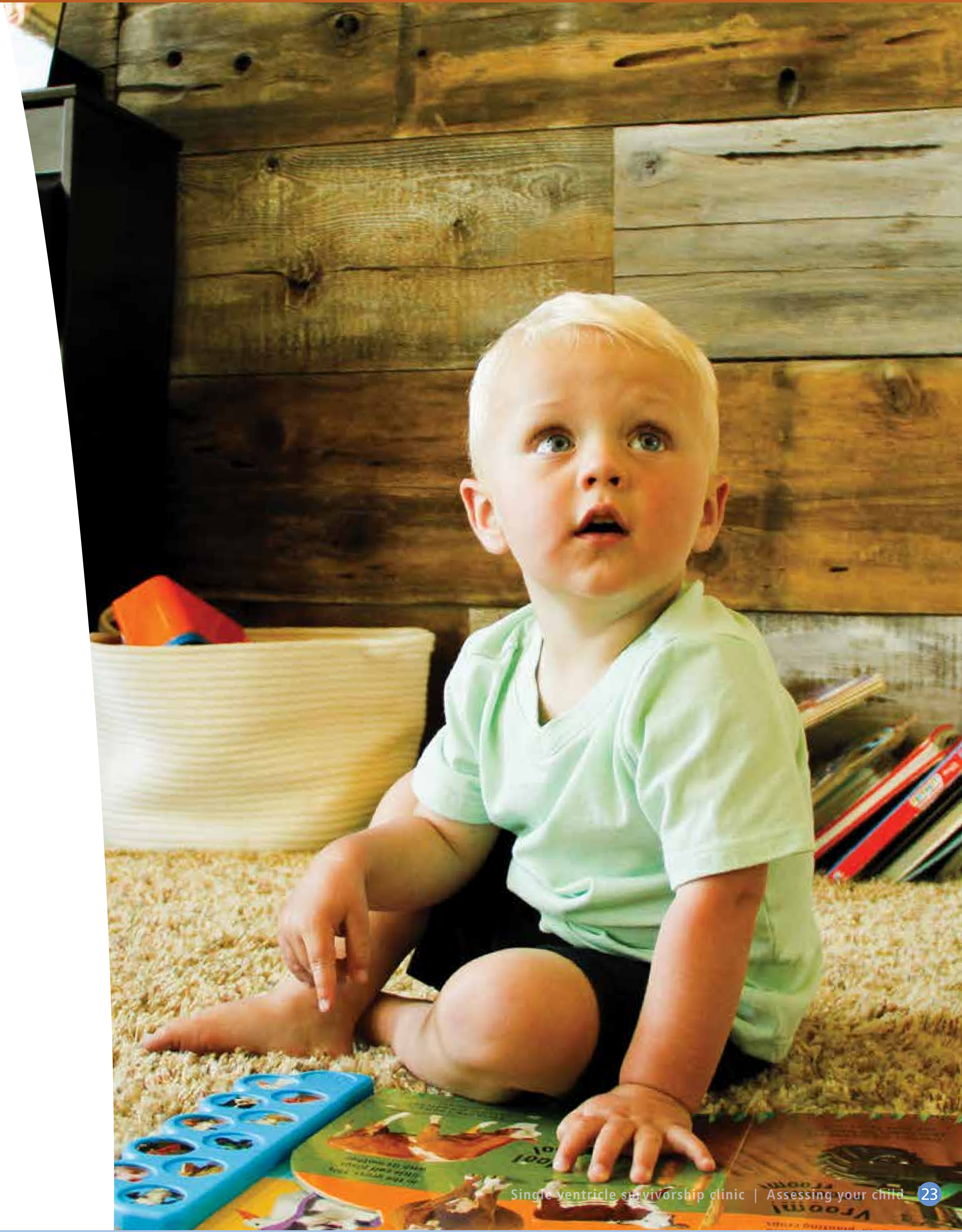
Kidney function blood tests

Having multiple open-heart surgeries is a known risk factor for kidney damage.

- **BUN and creatinine levels:** Patients often have blood tests of blood urea nitrogen (BUN) and creatinine levels to assess how well the kidneys are working.
- **Urine analysis:** Healthcare providers evaluate urine for protein loss.

Loss of protein

One of the most concerning problems after the Fontan surgery is an excessive loss of protein from the gut (called protein-losing enteropathy [en·ter·op·a·thy] or PLE. Patients may need to have their stool tested to see if they are losing needed protein.



Living healthy

After Fontan surgery, many patients struggle with physical, developmental, social, and behavioral health issues. The single ventricle survivorship clinic can help you or your child manage these problems and offer resources for your family.

> *"What happens in the beginning is not the only experience you will have together. Allow things to progress within your relationship with your child and this diagnosis. The personal experiences you will share with each other will expand and become far more important than the diagnosis itself. The rewards of discovering who your child is as they grow far outweigh any of the fears."*
>
> *– Parent, Single ventricle survivorship clinic*

People with congenital (present from birth) heart defects can also get acquired heart disease. To protect their heart, patients should eat a healthy diet rich in fruits, vegetables, and whole grains and low in saturated fats. They should also monitor their weight, cholesterol, and blood pressure and avoid smoking, using drugs, and drinking alcohol in excess. Your primary cardiologist will discuss risks for acquired heart disease. Heart problems can start without symptoms, and early treatment can protect the heart and body.

Activity and exercise

Patients with single ventricle heart diseases may have lower exercise endurance compared to their peers. A personalized exercise program is best planned with your doctor so that all factors can be included. Although patients with single ventricle heart defects are generally restricted from participating in vigorous or competitive sports, they should be encouraged to be active, exercise regularly, and to participate in recreational sports. It is important for them to always self-limit their activity and rest whenever they feel the need to do so.

Patients should do aerobic activity, which increases heart rate. This includes brisk walking, swimming, biking, jogging, rowing, cross-country skiing, hiking, or stair climbing. A good target is at least 30 minutes of aerobic activity a day for 5 or more days a week. Patients should avoid strenuous activities, like lifting heavy weights or doing sit-ups and push-ups. They should also avoid physical sports, like football, boxing, or hockey, which may increase the risk of injury, bleeding, and unnecessary heart strain. If a patient's heart isn't doing well, the cardiologist can recommend safe exercise.

Dental and oral health

Children with single ventricle heart disease have a higher risk of dental disease because of poor enamel, medicines, and nutritional issues. Bacteria can enter the bloodstream during dental procedures and cause a heart infection (infective endocarditis).

The American Dental Association recommends keeping the mouth as clean and healthy as possible to limit bacteria exposure without antibiotic use. Regular brushing, flossing, and dental visits starting before 18 months old are important for preventing and treating dental disease. Patients should also take antibiotics before dental procedures to help prevent heart infections (recommended by the American Heart Association). Cardiologists can provide more information.

School

Single ventricle patients may need extra help with school or personal care in regular or special education classes. They should have a written individualized healthcare plan (IHP) that lists their specific health needs and healthcare actions during school. For example, your child may need to rest or take medicine, have unlimited access to the restroom, or attend a shorter school day.

There should be a discussion with your child's physical eduction teachers about the importance of letting them self-limit their activity. Your cardiologist can provide a letter stating what your child can and cannot do.

Parents should tell teachers and school leaders about patients' specific needs. The pediatrician, primary cardiologist, and social worker can help with these plans.

Some students qualify for other aid. They may need a 504 plan (written educational need plan) or individualized education plan (IEP) for special education services. Talk to a teacher, school counselor, or principal for a special services evaluation.

Social work

A dedicated social worker can share hospital and community resources to help patients and their families cope with medical challenges and how your child's illness impacts the family.

A social worker will contact patients to discuss:

- Emotional support to cope with medical fears, anxiety, stress, and grief
- Decision making and talking with the care team
- Support and help with family conflicts
- Financial questions
- Behavioral health resources

Development and behavioral health

Single ventricle heart disease patients have an increased risk for developmental and behavioral disabilities.

These may include social problems, speech delays, inattention, impulsive behavior, and problems in school performance. Anxiety, attention issues, and depression are also common.

Because of this, patients often need school and behavioral health support services. Parents and siblings may also have posttraumatic stress disorder, depression, or anxiety because of the stress of the disease.

The clinic can help patients meet with the neurodevelopmental clinic, refer your child to the Heart Center Neurodevelopmental Program (HCNP), and find behavioral health resources.

Research opportunities

One of the single ventricle survivorship clinic's primary goals is fostering research. The clinic is actively working on many research projects with national and international collaborators to improve care and outcomes for single ventricle patients.

Our single ventricle survivorship medical team are world renowned experts in this field and are leaders in medical research in this area. Patients may be asked to participate in new or ongoing research studies.

Intermountain Healthcare's Primary Children's Hospital and University of Utah pediatric cardiology program is focused on translating scientific discoveries from the lab into leading-edge treatments and therapies for patients with single ventricle heart defects.

Improving care of single ventricle heart defects is an area of intense investigation and research. As is the case for other research studies, participation is completely optional and will not affect the care and evaluation you or your child receives through this clinic and the Heart Center.

Developing a healthy lifestyle

Single ventricle heart disease patients need life-long medical care and support to overcome physical and psychological challenges and to gain control over their disease to live normal lives. As they transition to adult care, however, they must be aware of lifestyle decisions that can impact their health. This section includes information on body art and piercings, drug and alcohol use, and pregnancy precautions.

> *"I live a healthy lifestyle, but I always know someone is looking out for me and has my best interests in mind. I never push myself too hard, even if someone tells me I can do more, because I know my strength and my heart."*
>
> *– Patient, Single ventricle survivorship clinic*

Piercings and tattoos

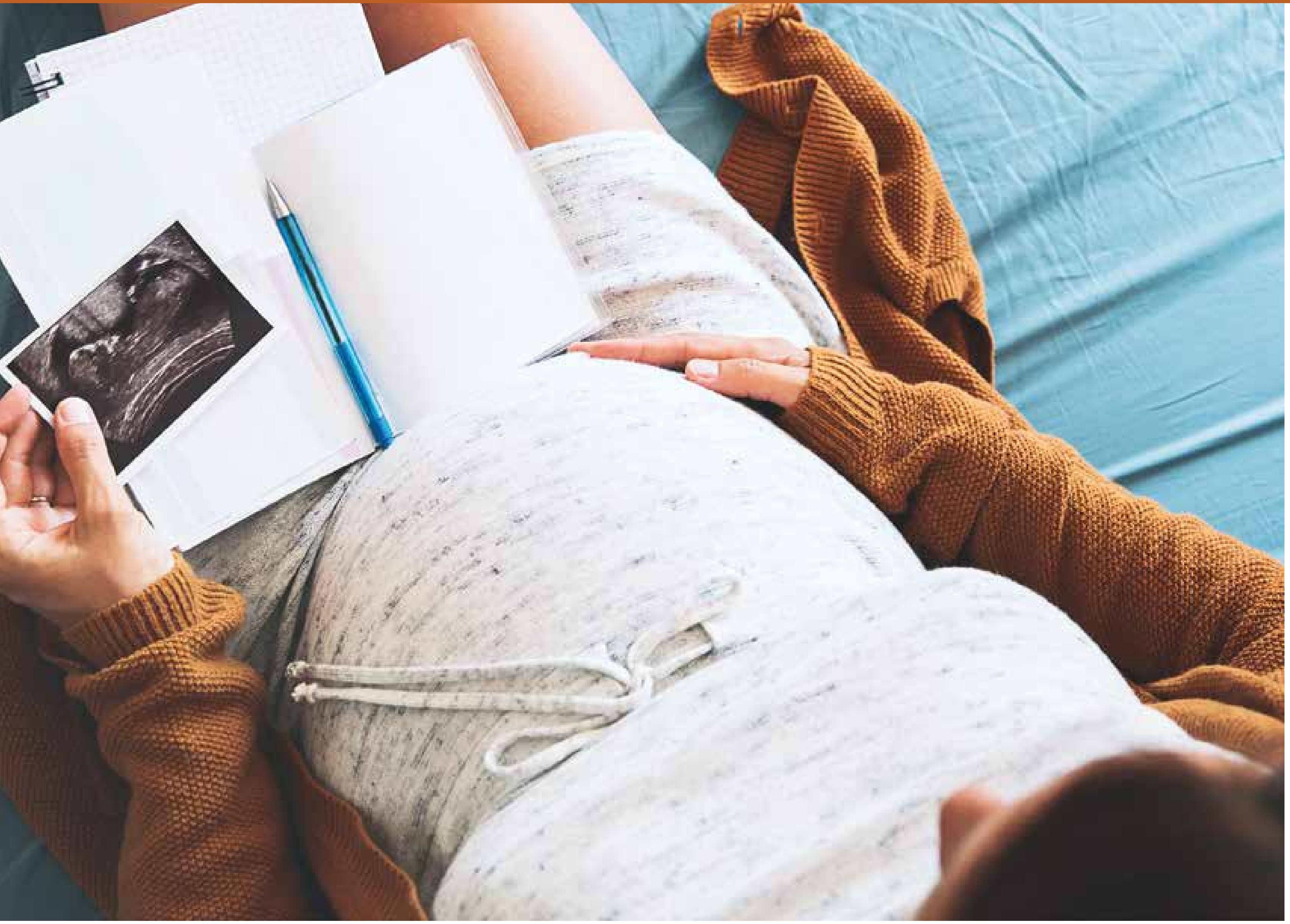
Like dental procedures, tattoos and piercings can potentially introduce bacteria or viruses into the bloodstream. If needles aren't sterilized, patients can also acquire skin and muscle infections and blood infections, like HIV and hepatitis. The American Heart Association recommends single ventricle patients take an antibiotic before getting any piercings or tattoos. The antibiotic only protects against bacterial infections, not viral infections like HIV or hepatitis.

Single ventricle patients are often taking blood-thinner medicines. They have a higher risk for bleeding and problems during piercing and tattooing. Patients should talk to their cardiologist if they're considering body piercings or tattoos.

Alcohol, tobacco, and drugs

Patients with a congenital heart defect have a higher risk of harmful effects from tobacco and illegal drugs. Nicotine increases the risk of cardiovascular disease and causes cancer, and many illegal drugs cause heart problems ranging from abnormal heart rate to heart attacks. These drugs can be even more dangerous if the heart is already stressed. Alcohol can be dangerous for patients taking blood-thining or other medication, and it can make liver problems worse.

> *"Chicks dig scars. I love my scar and have never been afraid to show it off. I don't focus on it, mostly because I'm focusing on my ab muscles."*
>
> *– Patient, Single ventricle survivorship clinic*



Pregnancy and reproductive health

Pregnancy can be risky for a single ventricle patient. It puts stress on a woman's heart when blood volume and pressure increase.

Pregnancy also increases the risk of arrhythmia, blood clots, and heart failure. Healthcare providers may advise some single ventricle patients not to become pregnant. However, pregnancy may be possible for healthy patients, although there are concerns for premature birth and miscarriage.

Patients planning to become pregnant should talk to their cardiologist and obstetrician-gynecologist about the risks.

Before using any birth control, patients must talk to their cardiologist. Single ventricle and Fontan patients have an increased risk of blood clots and should be careful using birth control with estrogen. Progestin-only birth control is safe for all heart conditions, but patients must take the pill at the same time every day.

Other birth control options include an implanted contraceptive in the upper arm or hormone injections every 3 months.

If a patient decides they never want to have a baby, tubal ligation (having tubes tied) may be an option.

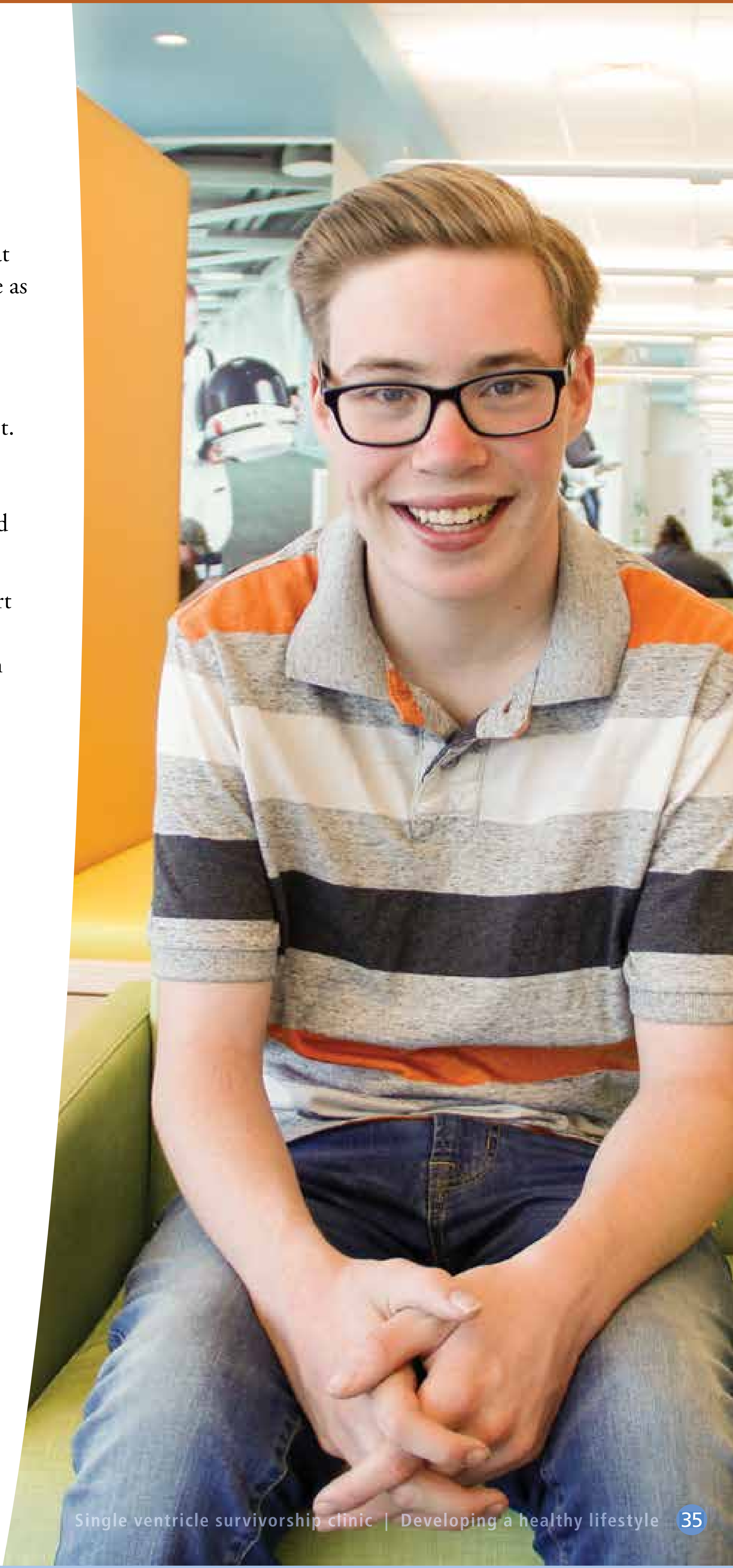
Transition to adult care

There is no cure for patients born with single ventricle heart disease. All surgeries and procedures are palliative, meaning that they improve survival and help patients be as healthy as possible.

Patients born with complex heart disease should receive lifelong care from an adult congenital heart disease (ACHD) specialist. These specialized cardiologists help adults born with congenital heart defects with lifestyle and family planning questions and provide ongoing cardiac care.

As children born with single ventricle heart defects enter adulthood, they can talk to their cardiologist about transitioning from pediatric to adult care.

> *"I see myself as a normal teen, someone who goes to school, stays after school to help set up a musical, or hangs out with his girlfriend or friends. I am a normal person, mostly thanks to my parents. I know I am unique. I just feel normal in a different way."*
>
> *– Patient, Single ventricle survivorship clinic*



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Find our support group on facebook at
Single Ventricle Survivorship

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