

The Heart Center
Primary Children's Outpatient Services
81 North Mario Capecchi Drive
Salt Lake City, UT 84113

For more information or to refer a patient:

801.213.3599

primarychildrens.org/heartcenter



SINGLE VENTRICLE SURVIVORSHIP CLINIC AT THE HEART CENTER



SINGLE VENTRICLE HEART DEFECT

Children with a single ventricle heart defect are born with one effective pumping chamber instead of two. This defect is a serious condition that requires 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 children born with this defect live to adulthood. To survive the first few years of life, these patients often undergo multiple open-heart surgeries and typically have a Fontan operation to allow them to survive with only one pumping ventricle.

The challenges don't end there. Patients living with the Fontan circulation will usually face a reduced life expectancy and are at risk for life-

long medical complications that may include liver dysfunction, reduced exercise capacity, abnormal heart rhythm, decreased heart function, reduced bone mineralization, delayed sexual maturation, short stature, abnormal clotting, protein-losing enteropathy, and plastic bronchitis.

WHY A SINGLE VENTRICLE SURVIVORSHIP CLINIC?

Now that survival rates have improved, the Single Ventricle Survivorship Clinic has been established with a new focus—to help single ventricle patients achieve the good health necessary to enjoy life, and to help manage any complications that arise related to single ventricle physiology. Since the health issues that Fontan patients experience can vary and often involve multiple organ systems, the Single Ventricle Survivorship Clinic uses a team approach and involves specialists from different medical fields to provide patient-centered, collaborative evaluation and treatments.



WORKING TOGETHER TO IMPROVE RESULTS

The team includes:

- Cardiologist
- Liver and gastrointestinal disease specialist
- Endocrinologist
- Lung disease specialist
- Nurse coordinator
- Nurse practitioner

Based on the individual needs of each patient, the team may also include:

- Hematologist
- Radiologist
- Cardiothoracic surgeon
- Holistic care provider

Based on each patient's clinical condition, tests or studies may be performed. The Single Ventricle Survivorship Clinic will work with health insurance providers to try and obtain approval for all tests ordered. If a patient's primary cardiologist or other health care provider has already performed any of these tests, the clinic may not repeat them.



THE PROCESS

Patients will be invited to visit the Single Ventricle Survivorship Clinic every three to five years, starting at five years of age. This clinic visit will be in addition to their regular visits. The clinic will work closely with each patient's regular cardiologist and primary care physician to provide updates on the findings and recommendations of the clinic's different specialists.

Cardiologists may refer a child to the clinic earlier than the age of five, particularly if the child develops Fontan complications like protein-losing enteropathy, plastic bronchitis, liver disease, or Fontan failure.

COMMON TESTS

Based on a patient's clinical condition and age, a variety of tests will be performed to evaluate the health of their heart, liver, muscle, bones, blood, and lungs.

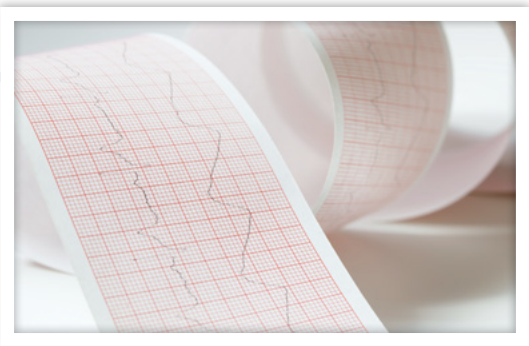


HEART TESTS

ELECTRICAL ACTIVITY HEART TESTS

Multiple open-heart surgeries, in combination with 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 prompt the heart to beat at a normal rate. The clinic will evaluate the heart's rhythm using these screening tests:

- An electrocardiogram (EKG or ECG)
An ECG translates the heart's electrical activity into line tracings on paper giving information on heart rhythm and condition.
- 24-hour Holter monitor
Electrodes (small electricity-conducting patches) are stuck onto a patient's chest and attached to a small recording monitor. The battery-operated monitor is carried in a pocket or small pouch worn around the patient's neck or waist.
 - The monitor records the heart's electrical activity.
 - Patients keep a diary of activities performed while wearing the monitor, noting how they feel.
 - After 24-48 hours, patients will return the monitor to the clinic.
 - A physician at the clinic will look at the recordings for any abnormal heart rhythms.
- Pacemaker testing



HEART FAILURE TEST

B-Type Natriuretic Peptide (BNP) is a substance secreted from the lower chambers of the heart (ventricles) that is caused by increased pressure from developing or worsening heart failure. We will check the BNP with a blood test.

EXERCISE STRESS TEST

An exercise stress test is an excellent way of monitoring heart performance and is a screening tool used to test the effects of exercise on the heart. To conduct the test, a patient walks on a treadmill or pedals an exercise bicycle. The patient will begin at a slow pace and will then be asked to walk or pedal faster and/or on an incline. During the test, the patient's breathing, exhaled gases, electrocardiogram, blood pressure, and oxygen saturation will be monitored by an exercise technician. An exercise stress test can be a valuable tool to monitor and record changes in the performance of a patient's heart over many years.



CARDIAC CATHETERIZATION

Cardiac catheterization is a procedure in which a catheter (a thin flexible tube) is passed into either side of the heart by way of a blood vessel in the groin, neck, or arm. Most patients have had catheterization procedures as part of their previous evaluations. Cardiac catheterization is the only way to precisely measure pressures within the Fontan circulation and lung arteries. High Fontan pressures may indicate problems with the lung or heart function and may require additional investigation and treatment. Cardiac catheterization may also allow transcatheter interventions that can improve blood flow or heart function without surgery.

CARDIAC MRI AND CT SCANS

Cardiac MRI uses radio waves, magnets, and a computer to create both still and moving pictures of the heart and blood vessels. Compared to echocardiograms, which are limited by the penetration of sound waves, cardiac MRI provides more detailed pictures of the beating heart and blood vessels in order to evaluate structure and function. Unlike CT scans, an MRI doesn't use ionizing radiation, so it doesn't increase the risk for cancer. Cardiac MRI is the only noninvasive imaging test that can differentiate healthy heart muscle tissue from dead or non-healthy tissue, while also providing valuable insight into heart function and determining if there is any narrowing in the arteries and veins. In patients with a pacemaker, ICD, or other metal implants, an MRI may not be recommended. These patients may undergo a CT scan instead.



LIVER TESTS



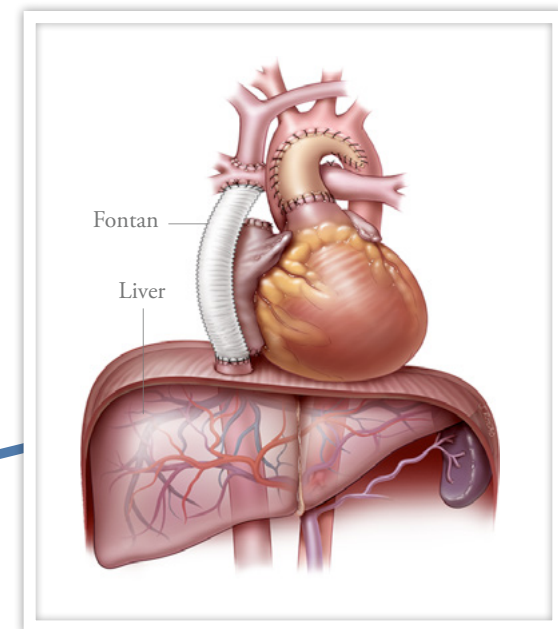
LIVER DISEASE BLOOD TEST

There has been growing awareness of the development of liver disease in Fontan patients. This is thought to be the result of a number of factors, including multiple surgeries, medications, overfilling of the veins with blood, and volume overload on the liver due to the lack of a ventricle to pump blood to the lung arteries. Blood tests and imaging studies are used for ongoing monitoring because they are low-to noninvasive. The clinic uses a combination of blood and imaging tests to identify patients who should undergo a liver biopsy, which is an invasive test.

- **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 assessment of liver fibrosis analyzes 10 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 Test**
Viral hepatitis can be a cause of liver dysfunction.
- **Alpha-Fetoprotein (AFP) Blood Test**
AFP is a protein normally produced by the liver and is used for screening of liver disease and protein-losing diseases of the intestines.

LIVER BIOPSY

Liver biopsy remains the gold standard for assessment of liver disease. A liver biopsy is a procedure using a needle to remove a small piece of the liver so that it can be examined under a microscope for signs of damage or disease. The test is conducted in a hospital as an outpatient procedure. Before the test is administered, patients are given medicines for sedation and pain. It is important for patients to remain as still as possible during the biopsy, which is commonly done through the abdominal wall with patients lying on their backs. After cleaning and numbing the area, a biopsy needle is inserted. Ultrasound is typically used to guide the needle or mark an optimal location. After the biopsy is obtained, the needle is removed quickly and a bandage is placed over the insertion site. The biopsy can also be done by inserting a needle into the jugular vein during cardiac catheterization. If the procedure is performed this way, X-rays will be used to guide the needle to the vein. A special needle and catheter is used to take the biopsy sample. Patients will need someone to drive them home.





LIVER MRI

MRI scans provide detailed images of soft tissues in the body by using radio waves and strong magnets instead of X-rays. When MRI is used to look at the liver, several sets of images may be taken. After the first set is done, a contrast material called gadolinium is injected into a vein in order to help show details more clearly. New sets of images are taken over the next several minutes as the gadolinium moves through the liver and other parts of the body. This is known as dynamic contrast-enhanced MRI, which is beneficial for viewing liver abnormalities like fibrosis and cirrhosis, and can help physicians discern between benign and malignant tumors. The scans can also be used to look at blood vessels in and around the liver.

LIVER CT SCAN

For some patients, including those with metal implants such as a pacemaker or an implantable cardioverter-defibrillator (ICD), MRI is not feasible. These patients will have their livers evaluated with a CT scan. A CT scan is an X-ray test that produces detailed cross-sectional images of the body. Similar to MRI, CT scans are helpful in looking at liver abnormalities like fibrosis, cirrhosis, and both non-malignant and malignant tumors, as well as nearby blood vessels.



LUNG TESTS

In single ventricle patients, after the Fontan procedure, the blood has to flow passively through the lungs without a pump. Since normal functioning of a Fontan circuit depends on the lungs being in good health, we will perform pulmonary (lung) function tests.

SPIROMETRY

The health of the lungs will be assessed using spirometry which measures airflow by measuring how much air you inhale and how quickly the air is exhaled. During these tests you may be asked to perform a forced inhalation or exhalation after a deep breath, breathe normally and quietly or inhale a medicine to see how it changes your test results.

PLASTIC BRONCHITIS

Plastic bronchitis is a rare but serious lung complication that may develop following the Fontan operation. In plastic bronchitis, the lung is unable to clear the lymphatic secretions and large, rubbery, cheese like mucus cast forms within the airways which can obstruct the airflow within the airways. In this condition, patients can have severe respiratory symptoms including breathing difficulty, low oxygen saturation and coughing episodes during which the mucus casts are coughed out. The Single Ventricle Survivorship team includes a pulmonologist who will evaluate, manage and follow patients with plastic bronchitis. With the newer treatment options, some patients with plastic bronchitis may improve without the need for cardiac transplantation.





BONE TESTS

BONE HEALTH BLOOD TEST

Serum Vitamin D, calcium, phosphorus, parathyroid hormones (PTH), reduced exercise capacity, delayed onset of puberty, and liver abnormalities may cause Fontan patients to have weak bones and muscles and vitamin D deficiency. The clinic will check blood levels of vitamin D and recommend vitamin D supplementation, as necessary.



DXA SCAN TO MEASURE BONE HEALTH (Dual-Energy X-ray Absorptiometry, or DXA)

Patients with the Fontan circulation commonly have low bone density. DXA is the preferred technique for measuring bone mineral density because it is relatively easy to perform and the amount of radiation exposure is low. Patients with low bone density will be referred to a specialist for further evaluation and treatment.



BONE AGE & SEXUAL MATURITY TEST

Like other chronic diseases, chronic heart disease can delay the growth and maturation. A bone age study estimates the maturity of a child's skeletal system which can help doctors diagnose delayed growth and development. It's usually done by taking a low dose X-ray of the wrist, hand, and fingers. Sexual maturity will be assessed by the endocrinologist and your self-reporting of sexual maturity staging. If required, delayed growth and sexual maturation can be treated with medication.

OTHER TESTS





KIDNEY FUNCTION BLOOD TESTS

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

- BUN and creatinine levels
Blood tests of urea nitrogen (BUN) and creatinine levels are often done to assess how well the kidneys are working.
- Serum Vitamin D and PTH
Reduced exercise capacity, delayed onset of puberty, and liver abnormalities may cause Fontan patients to have weak bones and muscles and vitamin D deficiency. The clinic will check blood levels of vitamin D and recommend vitamin D supplementation, as necessary.

BLOOD CLOTTING

Patients with a single ventricle may have abnormalities in blood clotting that may predispose them to abnormal clot formation or bleeding. If indicated, tests for blood components and clotting will be performed.

LOSS OF PROTEIN

One of the most concerning complications of the Fontan operation is excessive loss of protein from the gut, known as Protein-Losing Enteropathy (PLE). A screening test for protein loss in the stool may be performed.



Visit date: _____

NOTES:

Visit one: _____

Visit two: _____

NOTES:

Visit three: _____

Visit four: _____

NOTES:

Visit five: _____

Visit six: _____

NOTES:

Visit seven: _____

Visit eight: _____
