Heart Center
High Risk Program

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


University Hospital
Eccles Primary Children's Outpatient Services

Dear families,

Having a new baby can be an overwhelming experience on its own, but having a new baby with single ventricle heart disease can be anxiety-provoking and frightening as well. The Primary Children's Heart Center started the High Risk Program in 2012 to closely monitor babies born with single ventricle heart disease.

The High Risk Program team is here to provide care and guidance throughout the early months of your baby's life. This begins at your child's birth, continues as you transition from the hospital to your home, and extends until after your baby's second surgery, which usually happens between birth and 4 to 6 months of age.

The High Risk Program has a dedicated team of advanced practitioners (nurse practitioners and physician assistants); doctors; nutritionists; speech and physical therapists; and social workers. We compiled this booklet to help you prepare for what you and your baby may experience while in the hospital and at home. This booklet is intended to help educate you and to help you feel more comfortable with the care your baby receives in the hospital and the care that will be needed at home. This booklet can be looked at and reviewed at your leisure, but please feel free to ask the High Risk Program team any questions or concerns you may have.

Our healthcare providers recognize the burden of healthcare issues your baby faces as well as the demands you as a parent are facing with time, family, and financial commitments. We are committed to working with you, your primary cardiologist, and your primary care provider to improve the health, growth, and the quality of life of your baby with single ventricle heart disease.

Sincerely,

The High Risk Program team

Heart Center High Risk Program

at Primary Children's Hospital

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NURSE PRACTITIONER
intermountain healthcare
ANNIE

The Heart Center High Risk Program

The Heart Center High Risk Program at Primary Children's Hospital was developed by a team of specialized healthcare providers to closely monitor babies born with single ventricle heart disease during the "interstage" period. This is the time between birth and 4 to 6 months when a baby is likely to have their second surgery. Babies in the High Risk Program typically have:

- Hypoplastic [hy-poh-PLAS-tik] left heart syndrome
- Unbalanced atrioventricular [ay-tree-oh-ven-TRIK-yoo-lahr] septal defects
- Tricuspid [tri-KUS-pihd] atresia [ah-TREE-zhia]
- Pulmonary [PUL-mon-air-ee] atresia with intact ventricular septum
- Double inlet left ventricle
- Some types of double outlet right ventricle

The following information will help you learn more about what to expect when your baby is born, how the High Risk Program works, and what resources can provide support.

Prenatal Diagnosis

If your baby was diagnosed with single ventricle heart disease before birth, a pediatric cardiologist in Primary Children's Fetal Heart Program will follow your baby's progress and work closely with your obstetrician. The cardiologist will recommend you deliver your baby at University of Utah Hospital and then transfer to Primary Children's Hospital after birth. (The 2 hospitals are side-by-side and connected by a hallway).

Your baby will have an echocardiogram (heart ultrasound) shortly after delivery to help the cardiologist plan the next steps of your baby's care. A lot of parents get stressed when planning for their baby's arrival. It can help to talk with other parents of children with single ventricle heart disease. If this sounds like something you'd like to do, ask the fetal coordinator to help you get connected.

What to expect after your baby is born

After delivery, your baby will be admitted to the **cardiac intensive care unit** (CICU) or **neonatal intensive care unit** (NICU). The unit is determined by your baby's heart defect and whether they were born prematurely.

Your baby may need a medication called **prostaglandin** [pros-teh-GLAND-in] (PGE), which helps keep the **patent** [PAT-ehnt] **ductus** [DUCK-tuss] **arteriosus** [are-tear-ee-OH-suss] (PDA) blood vessel open so blood flows to the lungs or body.

Healthcare providers will closely monitor your baby. Many **echocardiograms** [ek-oh-CAR-dee-oh-gramz] may be needed to diagnose your baby's specific heart defect and to aid in surgical planning.

Your baby may not need surgery as a newborn if their blood flow is balanced. This means your baby is getting just the right amount of blood flow to the lungs and body. Deciding whether or not to do surgery when your baby is a newborn may take several days. If surgery isn't needed, your baby will be transferred to the **Cardiac Care Unit (CCU)** for further monitoring before going home.

Caring for your baby after surgery

If your baby needs heart surgery as a newborn, they'll recover in the CICU. There, your baby will get a breathing tube and IV medicines to control pain and improve the heart's function. Your baby will also be given **diuretics** [dy-yur-RETT-iks] (medicines that help reduce fluid) to help swelling go down.

If the chest was left open after surgery, the surgeon will likely close it 2 to 3 days later, depending on the level of swelling and how your baby is doing. After surgery, your baby will get nutrition through an **IV** [eye-VEE]. Once your baby is stable, a healthcare provider will pass a feeding tube through their nose and into the stomach or small intestine to feed your baby.

As your baby heals and gains strength, the breathing tube will be removed, but your baby may still need oxygen. Healthcare providers will increase the amount of formula or breastmilk your baby receives based on feeding plans developed especially for single ventricle babies. Your baby may need a swallow study to see whether they can safely eat by mouth. It is not unusual for babies to go home with a feeding tube. A healthcare provider won't remove the feeding tube until your baby is able to take by mouth all the calories they need.

Your baby will be transferred to the CCU once the medical team decides they are doing well enough to be moved. The High Risk team will teach you more about caring for your baby before you leave the hospital.

Going Home

Before you can take your baby home, you must:

- Complete CPR training
- Learn how to mix feeds
- Learn to give your baby medicines
- Learn to use home medical equipment
- Spend at least 1 night at the hospital
- Review the High Risk Program binder with the High Risk Advanced Practice Provider
- Complete a discharge checklist

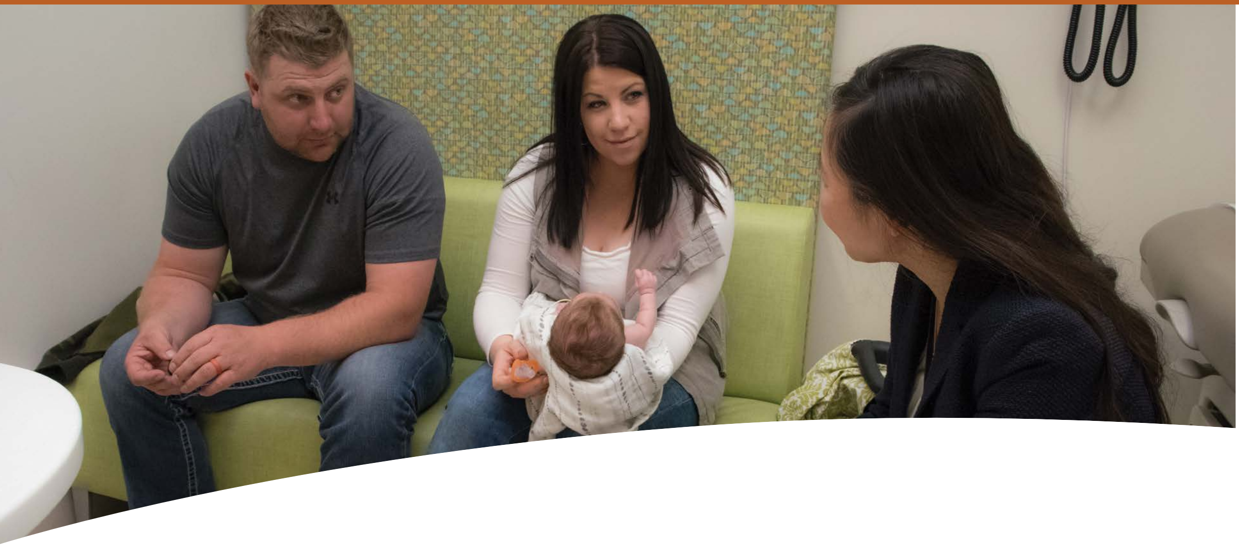
You will get the chance to practice all of these skills while your baby is in the CICU and CCU. The nursing staff will be there to help and answer questions. We want you to feel ready to go home and feel confident caring for your baby.

Ongoing care after you take your baby home

When your baby goes home, you will remain in close contact with the High Risk team. The **Advanced Practice Provider** will call weekly to see how you and your baby are doing. They will ask about oxygen saturations, feeding, and weight gain, and answer any questions you might have. Your baby will also have:

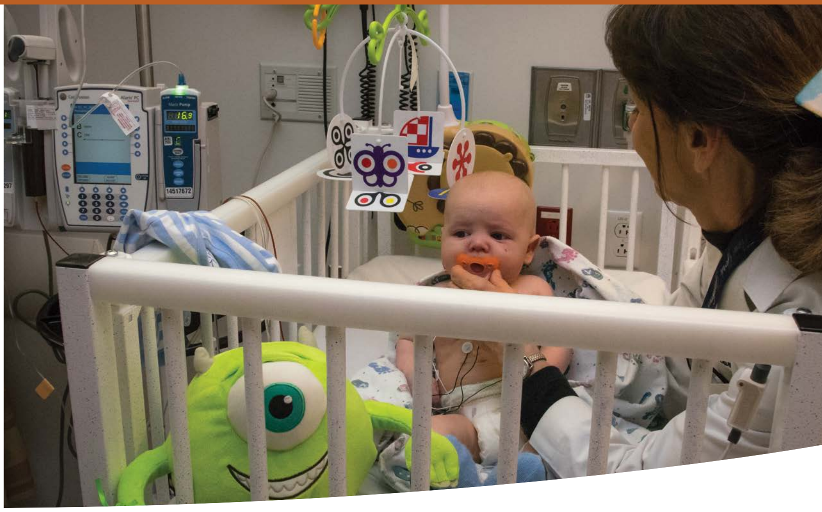
- Cardiology clinic appointments every 2 weeks, alternating between the High Risk Clinic and your baby's primary cardiologist. (A primary cardiologist is a heart doctor who oversees the overall health of people with heart disease.)
- Monthly echocardiograms
- Periodic electrocardiograms (ECGs) and chest X-rays
- Regular pediatrician visits

In addition, you can also call the High Risk Program anytime you have questions or concerns. Our phone number is on the back of this booklet.



Common Tests

- **Chest X-ray:** Special black-and-white images created with radiation to evaluate heart size and lung function.
- **Electrocardiogram** [ee-lek-troh-CAR-dee-oh-gram] **(ECG or EKG):** Test that records the heart's electrical activity using electrodes (sticky patches on your baby's chest).
- **Echocardiogram (echo):** Ultrasound test that allows cardiologists to see the heart's function and blood flow. If there are specific concerns, your baby may need echocardiograms more often.
- **Cardiac catheterization** [kath-eh-teh-reye-ZAY-shun]**:** Procedure in which a catheter (thin, flexible tube) is passed into the heart through a blood vessel in the groin, neck, or arm. The catheter is used to measure pressures within the heart and lungs. Cardiologists can also improve blood flow to the lungs or body during cardiac catheterization without the need for open-chest surgery. Babies often need cardiac catheterizations before the **Glenn surgery**, and some need them more often if not enough blood is flowing to the lungs or body.
- **Cardiac MRI:** Detailed motion and non-motion images of the heart and blood vessels created by radio waves, magnets, and a computer. A cardiac MRI is sometimes used instead of cardiac catheterization before the Glenn surgery.
- **CT scans:** Detailed still images of the heart and blood vessels created with radiation. CT scans are often faster than cardiac MRIs, so your baby may not need general anesthesia or sedation.
- **Holter monitor:** A small monitor your baby wears (along with electrodes) for 24 to 48 hours at home to record their heart rhythm over time. Your baby may need a Holter monitor if they've had abnormal heart rhythms in the past.



Common Issues during Interstage (Between Birth and Second Surgery)

Many babies with single ventricle heart disease, especially those who had newborn heart surgery, have trouble feeding by mouth, and some have feeding problems throughout the interstage. Your baby may need to have feeds completely, or partially, through an NG or NJ tube (different types of feeding tubes). The High Risk team works closely with nutritionists and speech therapists to help your baby eat and get enough nutrition. You may be able to participate in a specialized feeding clinic as well (see page 17 for more information on the feeding clinic, or ask a member of the High Risk team).

Blood in the stool

Bloody stool can be caused by anal fissures (small tears or cracks in the anus lining) or milk protein allergies. Your baby's milk protein allergies may improve by changing to a different formula or by not eating dairy if you breastfeed.

However, some babies born with heart disease can develop an intestinal infection called **necrotizing** [nek-roh-TIZE-ing] **enterocolitis** [en-ter-coh-LYE-tiss] (NEC). Your baby may need further testing if they have bloody stool, including an abdominal X-ray or a visit with a **pediatric gastroenterologist** [gas-troh-en-ter-AWL-uh-jist] or GI doctor.

Arrhythmias

There are many types of **arrhythmias** [ah-rith-mee-uhs] or abnormal heart rhythms. Sometimes an arrhythmia can cause poor blood flow from the heart. If this happens, your baby's heart may need to have electric shocks and possibly medicine to restore a normal heart rhythm. Your medical team will teach you arrhythmia signs to watch for and when to call the doctor.

Increased oxygen need

Your baby may need more oxygen because of a lung infection, narrowing shunt, or a shunt that's becoming too small. Signs that this is happening to your baby include a deeper blue tone in their lips and skin and lower oxygen saturation levels. If your baby suddenly needs a lot more oxygen, they may need a cardiac catheterization to see if the shunt is narrowing. Your baby may also need to be hospitalized.

Neurodevelopment

Studies have shown that congenital heart disease can increase the risk of neurodevelopmental and learning or behavioral disorders. Your baby may be at risk if they:

- Had surgery within a month after birth
- Have genetic syndromes
- Have cyanotic [sie-an-OTT-ik] heart disease (which includes single ventricle heart disease)

To help your baby early, you'll get a developmental plan. Your baby will also be checked by a physical therapist before going home. The High Risk Program will also refer your baby to your local Early Intervention program. You can also participate in the **Heart Center Neurodevelopmental Program** (see page 17).

Routine healthcare for your baby

In addition to the specialized care your baby receives for their heart, they will need routine vaccinations and regular visits to their primary healthcare provider.

Immunizing your family

Close family members should be up-to-date on the pertussis vaccine and get a flu shot to help protect your baby from illness. Your baby is too young to get the flu vaccine, and flu and pertussis can be life-threatening to your baby.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Caring For Your Baby After The Next Surgery

Your baby will likely have surgery when they are 4 to 6 months old (usually the Glenn surgery). After this surgery, babies' blood flow tends to be more stable. At this time, your baby will be out of the interstage period and will no longer need the High Risk Program.

However, you'll need to continue to take your baby to their primary cardiologist for checkups. Clinic appointments are often less frequent, and you'll no longer need home monitoring (including daily weight and saturation checks and weekly phone calls).

To ease the transition, a High Risk Program provider is available by phone even after the Glenn surgery. Remember, you are not alone. You can still contact the cardiology department at any time if you have questions.

Other Heart Center Specialty Clinics

Primary Children's Hospital has several other specialty clinics for children with congenital heart disease, including the:

- **Feeding clinic.** Speech therapists, physical therapists, and nutritionists help your baby learn to eat by mouth and make sure they are continuing to steadily gain weight.
- **Heart Center Neurodevelopmental Program (HCNP).** Neurodevelopmental pediatricians (doctors who are experts in children's growing brains and their ability to learn, focus, and remember) and pediatric cardiologists evaluate your baby to identify possible developmental challenges early in their life so they can get the help they need. You may be referred to this clinic at some point.
- **Single Ventricle Survivorship Clinic.** Children 5 years and older who have had Fontan surgery (connecting the inferior vena cava to the lungs) meet with pediatric cardiologists, gastroenterologists, and endocrinologists for help managing their health.

Research Opportunities

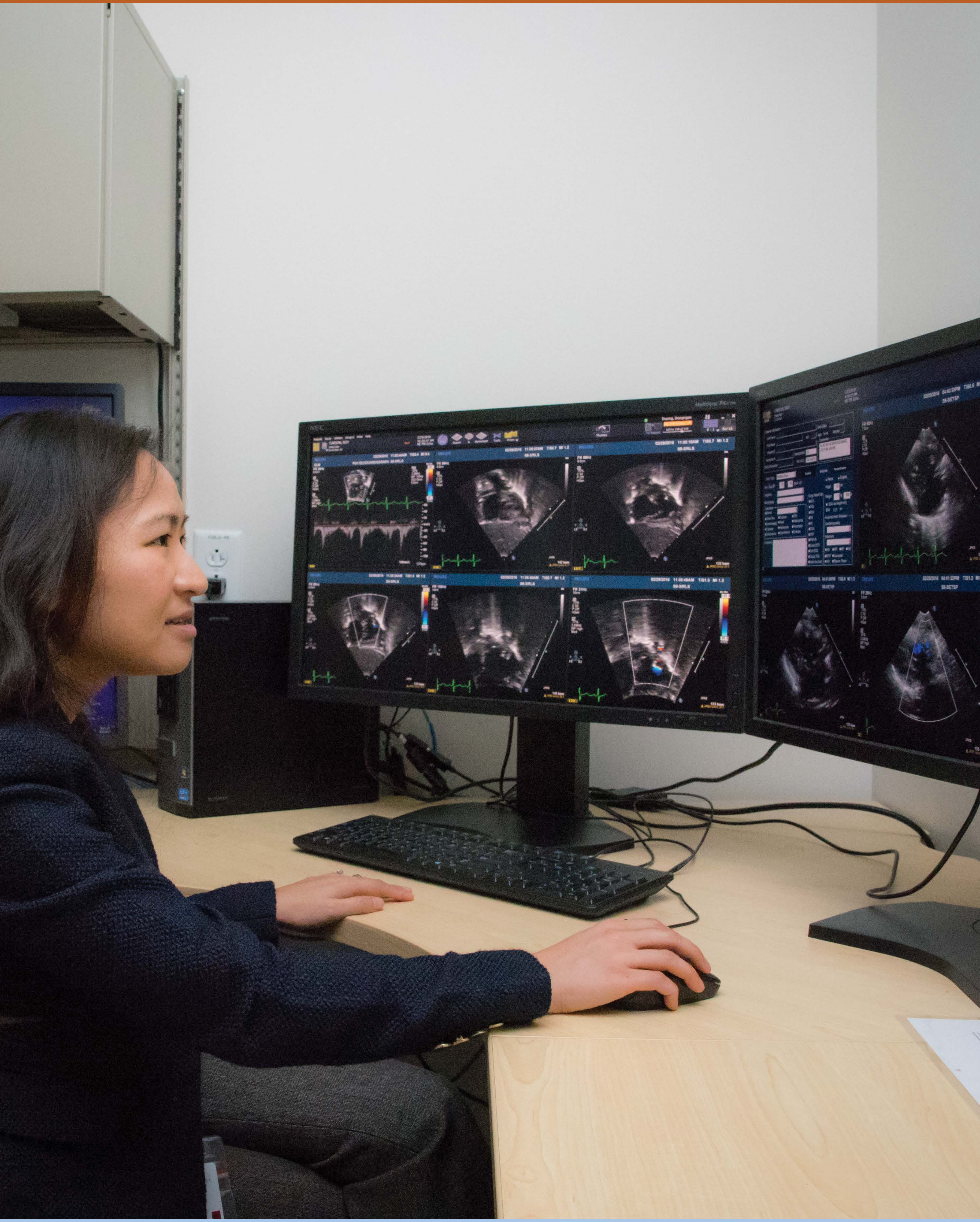
You may be asked about enrolling your child in congenital heart disease research studies. Participating is completely voluntary and does not affect your child's care. If you are interested in congenital heart disease research opportunities, please ask the medical team or visit the following organizations for more information:

- American Heart Association (**heart.org**)
- The Bench to Bassinet (**benchtobassinet.com**)
- The Congenital Heart Surgeons' Society (**chss.org**)
- The National Heart, Lung, and Blood Institute (**nhlbi.nih.gov/health-topics/about-clinical-trials**)
- The National Pediatric Cardiology Quality Improvement Collaborative (**npcqic.org**)
- The Pediatric Cardiac Critical Care Consortium (**pc4quality.org**)
- The Pediatric Heart Network (**pediatricheartnetwork.org**)

Resources for Families

Several local and national resources are available to help support families facing congenital heart disease, including:

- **Family to Family:** Primary Children's Hospital Heart Center has family volunteers who have children born with heart disease, including single ventricle heart disease. They can speak with you either in the hospital or by phone. Ask a social worker to help arrange this if you're interested.
- **Intermountain Healing Hearts:** Families throughout the Intermountain West who have children with congenital heart disease can join this support group (**Intermountainhealinghearts.org**).
- **It's My Heart:** This book, developed by the Children's Heart Foundation, describes various heart defects and explains common congenital heart disease tests, surgical procedures, and treatments. Ask the High Risk Program nurse practitioner for a free copy.
- **Mend A Heart Foundation**: This organization was founded by parents of a child with hypoplastic left heart syndrome. The Mend a Heart Foundation supports initiatives that extend and enrich the lives of heart kids by supporting medical practitioners and researchers who advance congenital heart defect research.
- **National Pediatric Cardiology Quality Improvement Collaborative:** This group of pediatric cardiology centers across the country work to improve patients' survival rates and quality of life after having a Norwood procedure (**npcqic.org**).
- **Sisters by Heart:** This national support group is available for families who have children with single ventricle heart disease (**sistersbyheart.org**).



The Heart Center

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For more information:

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primarychildrens.org/heartcenter

Find our support group on facebook at
https://intermountainhealthcare.org/primary-childrens/programs-specialties/heart

Special thanks to Clayhaus Photography
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