EMPOWER
MY CONGENITAL HEART

A Guide To *Confidently* Understanding and Navigating Your CHD Tests

Which Test is Right For Me?

Choosing the right cardiac (heart) test depends on many factors your cardiologist carefully considers. This guide educates you on what each test excels at, why it might be ordered, and some risks. This will prepare you to feel more confident asking informed questions and sharing any concerns or risks your cardiac team may not be aware of.

These cardiac tests are important for providing your cardiology team with information to:

- Evaluate your heart rhythm, structure, and function.
- Determine if any of your symptoms are related to your heart.
- Make informed decisions about adjusting or changing medications.
- Plan for procedures or surgeries effectively.

General Categories of Tests

Imaging-Based Tests

Create visual representations of the heart's structure and function, such as Echo, MRI, CT, and Stress Echo.

Non-Imaging Based Tests

Assess the heart's electrical activity and overall performance, such as EKG, Heart Monitor, and Stress Test.

Comparing Results Across Tests

Different heart tests, such as echocardiograms, cardiac MRIs, and CT scans, examine your heart in slightly differently. For example, your ejection fraction (EF)—a measure of how well your heart pumps blood—might not be the same when measured by an MRI versus an echocardiogram. It's important to understand that these differences don't necessarily mean one result is wrong. When tests are performed close together, differences in values may reflect the different methods used to measure your heart.

Where Should I Do My Imaging Tests?

Typically, all **cardiac imaging** should be done at a specialized CHD center.

Why? Specific expertise of the congenital heart care team and machines are required to obtain accurate images of your heart, which are often only available at CHD centers.

EMPOWERMENT

Mark Norris, MD, MS

ACHD Cardiologist

"CHD doctors often face problems when medical tests, even at well-known adult centers, are unclear, incomplete, or incorrectly read. This might require them to order the same test again, causing delays with insurance, extra radiation exposure, or delaying treatment."

EMPOWERMENT If you had imaging tests done at another clinic or hospital, try to get the images of the test on a CD to take to your ACHD visit.

What should I look for in Imaging Reports?

There are many nuances to how a report is generated. Below is a general guide.

1. **Heart Function Details:** Your Echo, MRI, or CT report will often contain a summary of your heart valves and heart function, including:
   - **Stenosis:** Describes a valve narrowing that can restrict blood flow. It is often assessed by the valve's pressure gradient across it. *(e.g., 20 mmHg)* ***Lower numbers are usually better.***
   - **Regurgitation/Insufficiency:** Describes the percentage of blood that leaks backward through a valve. *(e.g., 10%)* ***Lower numbers are usually better.***
   - **Ejection Fraction (EF):** Measures how much blood your heart pumps each beat. (e.g., 50%) ***Higher numbers are usually better.***
   - **Chamber Size:** Measures how big or small the atria or ventricles are. It is often assessed by measuring the volumes *(e.g., LVEDV, RVEDV, LAV)** ***Lower numbers are usually better.***

**Sometimes, these details are described subjectively such as normal, mildly, moderately or severely abnormal.*

What Should I Look for in Imaging Reports?

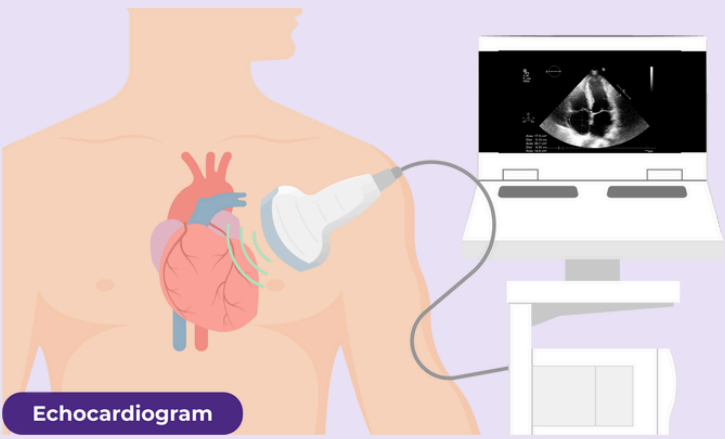
2 **Structural Details:** How the heart structures appear, such as narrow or enlarged blood vessels.

3 **Focus on the Change:** You have lived with your CHD all your life. So, how your heart function or structure appears on one report is less important than what has changed over time. Discuss any changes with your cardiologist, noting if new or worsening symptoms accompany them.

EMPOWERMENT It is important to confirm whether any variations from test to test reflect actual differences in your condition. Variations can sometimes reflect changes in the technique used in completing the study or reporting the test results. So, schedule a follow-up appointment with your ACHD team to discuss your test results.

Your Echo, Your Heart

Echocardiography is the first-line imaging choice for assessing heart health. It effectively evaluates function of the heart muscle and valves, and blood flow patterns.



What It Is

An **Echocardiogram**, or Echo, is an ultrasound test that uses sound waves to examine the **structure** and **function** of the heart.

How It Works

During an Echo, a cardiac sonographer uses an ultrasound device (transducer), which takes live images of your heart valves and chambers.

Prep: ~10-15 minutes; Test: ~45-60 minutes

Your Echo, Your Heart

Sound + Images

The sounds you hear during an Echo are blood flowing through your heart. These sounds, combined with images, help the cardiac sonographer and cardiologist assess:

- The speed and direction of blood flow.
- Whether valves are narrowed (stenosis) or leaking (regurgitation), and their severity.
- Whether there is abnormal blood flow between heart chambers, such as a hole in the heart wall, and the extent of this abnormal flow.

PEER-EMPOWERMENT

Drezden Plotkin *- Pulmonary Stenosis and Ventricular Septal Defect Patient*

> You should not be concerned if the test is longer, shorter, or sounds different than other tests you've taken. The experience can vary considerably based on the sonographer, their equipment, and the part(s) of the heart they are observing.

Your Echo, Your Heart

Other Important Assessments

- How well the heart muscle is pumping.
- The size of your pulmonary artery and aorta—the blood vessels carrying blood to and from your heart.
- Sometimes, an IV might be needed for the sonographer to inject saline or other nontoxic substances to obtain better pictures of the heart.

EMPOWERMENT Echocardiography is so popular because it uses **no radiation**, is extremely **cost-effective**, and can give instantaneous results.

Your Echo, Your Heart

Two Types of Echos

The sounds you hear during an Echo are blood flowing through your heart. These sounds, combined with images, help the cardiac sonographer and cardiologist assess: - The speed and direction of blood flow. - Whether valves are narrowed (stenosis) or leaking (regurgitation), and their severity. - Whether there is abnormal blood flow between heart chambers, such as a hole in the heart wall, and the extent of this abnormal flow.

Transthoracic Echocardiogram (TTE)

The most common type of “heart echo” performed by placing the ultrasound transducer outside your chest (thoracic area).

Transesophageal Echocardiogram (TEE)

Performed by inserting a thin, flexible probe with an ultrasound transducer down your esophagus (swallowing tube). It shows your heart and valves in greater detail than a transthoracic echo because your bones and tissues aren’t between the transducer and your heart. TEE is only performed when your cardiologist needs to better visualize any of your heart structures.

Your Echo, Your Heart

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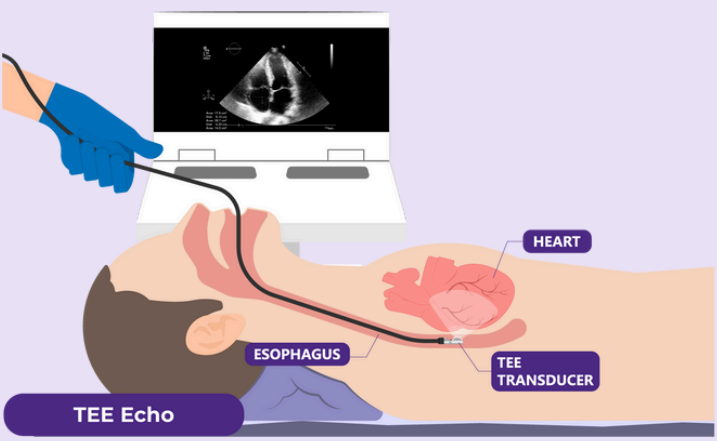
During a TEE, the flexible probe is lubricated to help it easily slide down your throat and esophagus. While this may feel uncomfortable, it usually won't harm you. You may need to swallow to move the transducer into the right spot. Also, you are usually put to light sleep during the TEE, so arrange transportation as you cannot drive yourself.



Heart Function Unlocked, Embrace the Tube

Cardiac MRI provides higher-resolution and more accurate measurements of many heart features assessed by an echocardiogram, including heart function, valve performance, and tissue health. It does this by excelling at high-resolution soft tissue imaging.

What It Is:

A Cardiac MRI (Magnetic Resonance Imaging) test uses powerful magnets and radio waves to create detailed pictures of your heart's structure and function.

How It Works:

During the test, you lie on a table that slides into a large, tube-shaped MRI scanner. The scanner uses powerful magnets, which rotate around you, and radio waves to create highly detailed images of your heart, focusing on soft tissues like the heart muscle and valves. The radiology tech **may administer contrast** (Gadolinium) through an IV to enhance image clarity.

Prep: ~30 minutes; Test: 1-2 hours

Heart Function Unlocked, Embrace the Tube

Cardiac MRI focuses on detailed imaging of soft tissues, **does not involve radiation**, and is ideal for evaluating heart muscle, inflammation, and scarring.

EMPOWERMENT MRIs could be very **loud and long**. Wear headphones during the test. The MRI technologists usually offer one or you may be able to use your own during the test.

Cardiac MRI Strengths in Heart Imaging

- Measures how much blood the ventricles pump into your body or lungs (ejection fraction [EF]).
- Measures the size of your heart chambers and identifies any enlargement (dilation).
- Assesses valve insufficiency (regurgitation) or narrowing (stenosis) and their severity.
- Visualizes blood flow through your heart to examine the blood vessels to and from the heart.
- Detects inflammation or scarring in the heart muscle if present.

Heart Function Unlocked, Embrace the Tube

Setting Yourself Up for a Successful MRI

Some people (especially those who fear **closed spaces,** also known as ‘claustrophobia’), often find it difficult to tolerate a cardiac MRI. Your doctor may be able to give you some medication to relax you.

$ MRI is **significantly more expensive** than an Echo, so check with your insurance or doctor to find out what you’ll owe after insurance. Being proactive can help you **avoid unexpected bills**.

MRI uses magnets, so metal in your body can affect the images. Avoid wearing any jewelry during the MRI. If you have a pacemaker, defibrillator, or other implanted device, **bring your device card** and inform your doctor or MRI technician about any metal in your body, like a spinal rod.

Your Anatomy, Your Answers

While echocardiography and cardiac MRI are excellent for evaluating heart function and tissue health, Cardiac CT is particularly valuable for visualizing **complex cardiac anatomy** and **blood vessels**. It does this by excelling at high-resolution, three-dimensional spatial imaging.

What It Is:

A Cardiac CT uses X-rays to create 3D images of your heart structure and blood vessels.

How It Works:

During the test, you lie on a table that slides through a large, doughnut-shaped CT scanner. The scanner uses X-rays to take detailed, high-resolution images of your heart and blood vessels. The radiology tech **usually administers contrast** (Iodine) through an IV to enhance image clarity.

Prep: ~30 minutes; Test: 30-60 minutes

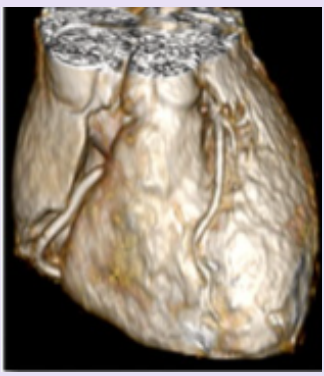
Your Anatomy, Your Answers

Cardiac CT is faster than Cardiac MRI, uses X-rays, and has some radiation exposure. It excels in capturing detailed images of blood vessels, bones, and large structures like the aorta and pulmonary artery. Cardiac CT is usually better tolerated than MRI if you have claustrophobia. Like MRI CT is **expensive**.

Cardiac CT Strengths in Heart Imaging

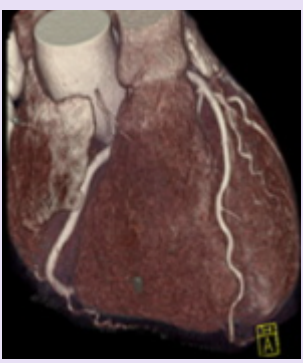
- Provides detailed visualizations of calcifications and anatomical differences that can impact valve function
- Evaluates the aorta and pulmonary arteries for conditions like enlargement (aneurysms) or narrowing (e.g., coarctation).
- Detects blockages, narrowing, and calcification in the coronary arteries, which can affect blood flow to the heart muscle.
- Visualizes abnormal blood flow patterns (such as fistula, collaterals, etc.)
- Help in planning for procedures.

MRI and CT Key Takeaways



MRI Key Takeaways

- No Radiation
- May Use Contrast
- Excels at Measuring Function



CT Key Takeaways

- Radiation
- Usually Uses Contrast
- Excels at Visualizing Structure

Non-Imaging Tests

Non-imaging tests often measure the electrical signals from your heart. By studying these signals, doctors can see how well your heart's natural electrical system works, find problems with blood flow, and check the size of its chambers.

Where Should I Do My Non-Imaging Tests?

Typically, you can do these tests at any center that offers them and is most convenient for you.

Non-Imaging Test Strengths

- **Heart Rhythm:** Checks how the heart's electrical system works and shows if it beats at a normal rate and pattern.
- **Arrhythmia**: Looks for irregular heartbeats (arrhythmia) and extra electrical pathways that can lead to abnormal heart rhythms.
- **Heart Block**: Finds blocks in the heart's electrical signals.
- **Chamber Size:** Shows if the heart's chambers (atria and ventricles) are larger than normal.
- **Blood Flow Issues:** Helps detect signs of a blocked artery causing reduced blood flow to the heart muscle.

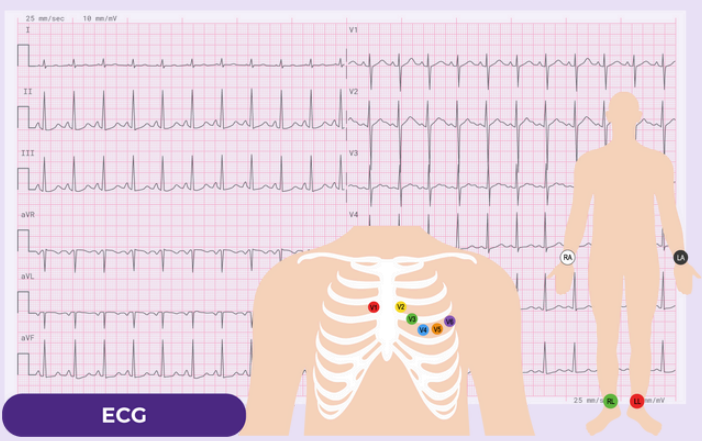
My Beat, My Rhythm

An **Electrocardiogram** (ECG) is the first-line **non-imaging** choice for assessing heart health. It provides valuable information about the heart's electrical system and can also give clues about the heart muscle and chamber sizes.

Also known as an EKG!

What It Is

An **ECG** is a non-imaging test that records the electrical activity of your heart.



EMPOWERMENT: Discuss any new or changed symptoms with your cardiologist, such as palpitations or chest pain at rest or exertion.

My Beat, My Rhythm

How it Works

An **ECG** is a simple test where small stickers (called electrodes) are placed on your chest, arms, and legs. These electrodes are connected to an ECG machine. The electrodes pick up electrical activity from the heart and create a picture of your heart rhythm.

Prep: ~5 minutes; Test: 5-10 minutes

An **ECG** helps your doctor to evaluate:

- If there are heart rhythm disturbances such as arrhythmia
- If there are blocks in the electrical conduction system
- If you have a pacemaker, ECG helps determine how well it is working
- If there are signs of blockages in arteries that supply the heart muscle
- If there are indirect signs of enlargement in heart chamber size

Hide and Seek, Find the Beat

A **Heart Monitor** is a non-imaging tool that tracks your heart's electrical activity over time. It provides valuable information about your heart rhythm, helps detect irregular heartbeats (arrhythmias), and heart blocks.

Holter Monitor

Event Monitor

Prep: ~15 - 20 minutes; Test: 1 – 30 days

What It Is

A **Heart Monitor** is a portable, long-term ECG device worn for 24-48 hours (Holter Monitor) or 7-30 days (Cardiac Event Monitor), which provides information about the heart's electrical system.

Your doctor may recommend a **Holter Monitor** or **Cardiac Event Monitor** based on availability, insurance coverage, and the nature of your symptoms.

Hide and Seek, Find the Beat

How It Works

A **Heart Monitor** is done by attaching small stickers (electrodes) to your chest. These electrodes are connected to a small device that **continuously records your heart's activity**.

You can wear a Heart Monitor for 1 to 30 days, depending on the type. It keeps track of your heart rhythm during that time. After you return the monitor, your doctor will check the recordings for heart rhythm problems.

Benefits of a Heart Monitor

Patients with congenital heart disease are at risk for abnormal heart rhythms and blocks in the electrical system of the heart. A heart monitor checks your heart's rhythm over a **longer period** than a regular ECG. Helping your doctor to evaluate:

- **Slow heart rate** (bradycardia), **fast heart rate** (tachycardia), or **arrhythmias** such as atrial fibrillation, atrial flutter, or ventricular tachycardia.
- If there are **blocks** in the heart's electrical signals.
- How well a pacemaker is working if you have one.

Hide and Seek, Find the Beat

Preparing for a Smooth Heart Monitor Experience

- You should resume day-to-day activities, including sports and exercise.
- Some people experience discomfort or skin irritation from the electrodes.
- Some devices, like cell phones and magnets, can interfere with the heart monitor's signal. To avoid this, keep them at least 6 inches away from the monitor.
- If wearable event monitors fall off, use tape to put them back on.
- Water can damage some Heart Monitors. The technician will tell you whether you can swim or shower while wearing it.

EMPOWERMENT Keep a diary to note your activities and any symptoms while wearing your monitor. If you notice any symptoms, such as palpitations, chest pain, fast heartbeat, or dizziness, push the button on the monitor to mark the event. This will help your doctor assess whether these symptoms are related to an abnormal heart rhythm. Don't worry if you forget to press the button. The monitor will be continuously recording.

Your Exercise Test, Pushing Your Limits

Stress Tests evaluate your heart's response to exercise. There are three common types for CHD patients: an Exercise EKG Test, a Cardiopulmonary Exercise Test (CPET), and a Stress Echo.

PEER-QUESTION

Mary Jane Klein - *Aortic Valve and Subaortic Stenosis Patient*

"Are stress tests necessary when you have a pacemaker?"

PROVIDER-ANSWER

Leigh Reardon, MD *- Pediatric and Adult Congenital Cardiologist*

"Having a pacemaker does not limit the ability to do a stress test or affect the accuracy of the results. Stress tests are ordered for various reasons—from assessing one's exercise capacity, blood pressure and heart rate response to exercise, or signs of compromised blood supply to the heart. Ask your physician to explain why they ordered the test."

Your Exercise Test, Pushing Your Limits

There are Three Common Types of Exercise Test for CHD Patients

- Exercise EKG Test
- Cardiopulmonary Exercise Test
- Stress Echo

Exercise EKG Test

- You exercise on a bike or treadmill, which gets harder over time.
- Records your heart rate, blood pressure, and EKG during exercise.
- Helps your cardiologist evaluate your cardiac health and clear you for sports or physical activities.
- If you have any symptoms, an exercise test helps your cardiologist determine if your **symptoms are related to your heart**.

Exercise Time: 8-30 minutes; Total Time (Prep, Exercise, Cool Down): 1-2 hours

All other stress tests build off the foundation of the exercise EKG test.

EMPOWERMENT Let your cardiologist know if you have been told by another doctor that you have to limit your exercise, for example due to your knees or back.

Your Exercise Test, Pushing Your Limits

Cardiopulmonary Exercise Test (CPET)

- You breathe into a mask or mouthpiece, and a breath analysis (ventilatory gas exchange) is recorded.
- This analysis measures how much oxygen you inhale and how much carbon dioxide you exhale.
- It helps the cardiologist evaluate how your heart and lungs respond to exercise and can help determine if symptoms are related to **heart issues, lung function, or both.**

Exercise Time: 8-30 minutes; Total Time (Prep, Exercise, Cool Down): 2-3 hours

EMPOWERMENT Wear comfortable shoes and clothes for exercise. Let your cardiologist know if you have been told by another doctor that you have to limit your exercise. Avoid exercising on the day of the CPET as this can affect the results.

Your Exercise Test, Pushing Your Limits

Stress ECHO

- A standard Echo is performed both **before** exercise and during **peak** exercise.
- The results of the two Echos are compared for any differences in heart function when your heart is exercising.
- A stress echo helps detect issues like **ischemia** (reduced blood flow to heart muscle) or **valve dysfunction**, which may only become apparent when the heart is under physical stress.

Exercise Time: 8-30 minutes; Total Time (Prep, Exercise, Cool Down): 3-4 hours

Scheduling Tips

Need to schedule multiple tests?

If convenient, you can schedule all or most of your tests on the same day! However, this might require you to call multiple departments, such as Radiology, to schedule your MRI and the cardiology department to schedule your echo or stress test. Often, ACHD programs have patient coordinators or navigators who can help schedule most of the tests for you on the same day—ask your ACHD team about it!

PEER-EMPOWERMENT

Joe Valente - *Tetralogy Patient*

> I will not schedule a Cardiac MRI and Cardiopulmonary Stress test on the same day unless necessary. For me, they are too exhausting both mentally and physically.

Scheduling Tips

Need to arrange transportation?

Medicaid Transportation

State **Medicaid** programs offer **transportation** to help **eligible individuals** access care, such as medical, dental, or mental health appointments, and to pick up prescriptions or supplies for **covered services**.

- **Non-Emergency Medical Transportation** (NEMT)**:** Specialized transportation
- **Non-Medical Transportation** (NMT)**:** Public or private transportation

Public Transit

Public transit agencies must provide paratransit (accessible) services for individuals with **disabilities** who cannot use standard bus or rail services. These services require an application and cost a fee.

Search online for paratransit services in your area to learn more.

EMPOWERMENT Talk to the social or case worker at your primary care or CHD clinic for help finding transportation resources in your area and assistance with applying.

Scheduling Tips

Prior Authorization

Does this test require prior authorization?

- Most insurance plans require **authorization** (approval) before tests. Your doctor's office or hospital is responsible for getting this approval. To avoid problems, it's a good idea to check that the test has been approved. You usually have to pay the full cost if a test is done without approval.
- Ask for a payment plan if you end up with a bill you cannot pay.
- An authorization does not necessarily mean that insurance will pay the full cost of the test. Depending on your insurance, you may still be responsible for your share of the cost.

PEER-EMPOWERMENT

Karla Deal *- Tetralogy Patient*

"I received a medical bill that was higher than expected; fortunately, I was able to set up a monthly payment plan. Which helped alleviate the financial stress of a large one-time payment."

EMPOWERMENT Before you have a test, know how much you will owe for the test.

Scheduling Tips

Managing Insurance

When do I need a letter of medical necessity?

A letter of medical necessity is a letter that an insurance company will request from your physician asking for the reason or medical necessity for the procedure. Sometimes, this is a letter, or it can be a peer-to-peer conversation between your provider and a provider employed by the insurance company.

If your test is denied

A **denial** means the insurance does not cover the cost of the test. Often, the insurance company needs more information to approve the test.

If your insurance company denies a test, procedure, or visit, contact your doctor's office immediately. Ask what they can do to help overturn the denial and start the appeal process. They can also guide you through the next steps, including contacting the hospital if necessary.

Scheduling Tips

If you receive a denial from your insurance company for a particular test (procedure or office visit), then please contact your doctor's office immediately. Make them aware of the denial and ask what they can do to overturn it. Your doctor's office will need to start an appeal process. If the denial is for a procedure at a hospital, you might need to call the hospital directly.

PEER-EMPOWERMENT

Karla Deal *- Tetralogy Patient*

"When my insurance company denied a test ordered by my cardiologist. I spoke with my cardiologist, and we discussed two possible solutions. The first option was to request a Peer-to-Peer Review. The second, suggested by my cardiologist, was to postpone the test to a later date when my insurance company would most likely approve it. I decided to postpone the test and have a virtual visit with my cardiologist instead."

Radiation

Some tests, like **CT** and **X-rays**, involve small amounts of radiation. The radiation risks are typically small compared to the study's benefits. However, these risks increase with age, pregnancy, and other conditions. If you are concerned, ask your provider for more information.

Long-Term Exposure: While the risk from a single imaging study is low, repeated radiation exposure over time can increase the risk of developing cancer. If you are undergoing repeated scans, particularly CT, you should be mindful of the cumulative dose. Discuss the necessity of each scan with your doctor and explore alternatives when possible.

Pregnancy: Radiation exposure during pregnancy carries a small risk to the developing fetus. Imaging studies involving radiation, such as CT scans or X-rays, are usually avoided, particularly in the first trimester, unless necessary.

Age: Children are more sensitive to radiation than adults. Clinicians prioritize echocardiograms or MRIs whenever possible.

Contrast

Contrast agents, used in imaging tests like **CT** or **MRI**, enhance the **clarity** of images by highlighting specific structures. While typically safe, they carry some risks, including:

Allergic Reactions: Include itching or a rash (rare) and difficulty breathing (extremely rare).

Side Effects: Include a metallic taste, nausea, or warmth during administration.

Impact on Kidney Function: In people with kidney conditions, contrast agents may temporarily or permanently affect kidney function.

Inform your healthcare team about any prior contrast reactions, allergies, or kidney issues so they can take appropriate precautions.

PEER-EMPOWERMENT

Karla Deal *- Tetralogy Patient*

"I was concerned when I had an allergic reaction to contrast because I did not know if the test would still be meaningful without the dye. I was happy to learn that these tests are still meaningful even if your provider has to take precautions."