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Cardiac Issues in Thalassemia

by Craig Butler



Many people with thalassemia develop cardiac issues (heart-related issues) that can have a significant impact on their health. Because cardiac-related complications are the leading cause of death among people with thalassemia, it is very important for patients who do not have cardiac issues to take steps to prevent these complications from developing, and for patients who already do have cardiac issues to work to reverse them or prevent them from getting worse.

Why are cardiac issues common in thalassemia?

Both the anemia caused by thalassemia and the iron overload that is related to the disorder and its treatment can contribute to cardiac issues.

Untreated anemia, in which hemoglobin levels are consistently too low, often causes tachycardia, (a heartbeat that is too fast), as the heart tries to compensate for the lack of oxygen being carried through the body. Over time, the heart also becomes enlarged.

Living with Thalassemia

Issues in Thalassemia Care

Iron overload, which is a consequence of transfusion therapy but which also occurs in patients who are not regularly transfused, results in excess iron being deposited in organs throughout the body, including the heart. This cardiac iron may cause heart problems such as arrhythmia (an abnormal heartbeat, such as tachycardia) and congestive heart failure (a decreased ability of the heart to pump blood).

What symptoms are related to cardiac issues and how are these issues diagnosed?

Symptoms may include shortness of breath or difficulty breathing (sometimes even when at rest); palpitations; fainting; chest pain; feeling very tired very easily; and unexplained swelling around the ankles.

However, symptoms of cardiac complications often don't show up until the complication is already advanced. Because cardiac issues are common in thalassemia, it is therefore necessary for patients to undergo regular tests that can pick up warning signs in the early stages. These tests may include:

- Chest x-ray
- Electrocardiogram
- 24-hour Holter monitor
- Cardiac Stress Test
- Echocardiogram
- MUGA Scan
- T2*
- Functional Cardiac MRI



shortness of breath; and Beta blockers, which slow heart rate and are beneficial in fighting arrhythmias.

It's important to remember that there is a great deal that can be done both to prevent and to treat cardiac complications in thalassemia. Making sure that you undergo a complete annual comprehensive care evaluation is essential to maintaining a healthy heart.

For more information about thalassemia and cardiac issues, or about other issues involved with thalassemia care, talk with your health care professional, or contact the Cooley's Anemia Foundation (800-522-7222 or info@cooleysanemia.org) or one of the Thalassemia Centers of Excellence.

The Thalassemia Centers of Excellence have the most highly trained thalassemia experts in the country. They are located at:

- Children's Hospital Boston
- Children's Hospital Los Angeles
- Children's Hospital Oakland
- Children's Hospital of Philadelphia
- Children's Memorial Hospital (Chicago)
- Weill Medical College of Cornell University (New York)

Many other hospitals are satellite centers affiliated with these Centers. Please contact the Cooley's Anemia Foundation for a list of these satellite centers.



heart's health. Smoking can have negative effects on your heart, as can too much alcohol. You also need to watch what you eat; as with the general population, eating too much of some foods can damage the heart. And people with thalassemia also need to be careful about eating foods that are too rich in iron or that increase iron absorption.

Stress and a positive outlook on life can also have an effect on cardiac health. Finding ways to relieve stress - whether through meditation, peaceful walks, or simply allowing yourself time to do things you enjoy - can be helpful.

How are cardiac issues treated?

Fortunately, progression of heart failure and other cardiac issues can be halted and in many cases reversed. The key to this is aggressive chelation therapy. This may take the form of 24-hour Desferal treatments, administered intravenously, or therapy involving both Desferal and the oral chelator deferiprone.

In addition, dietary changes may be required, as well as making sure the patient receives sufficient rest. There are some drug-based therapies that may also be utilized, such as ACE (Angiotensin Converting Enzyme) inhibitors, which help reduce blood pressure and improve heart function; Digitalis, which slows fast heartbeats; Diuretics, which reduce high blood pressure and help with

After a certain age - usually around 10 - 12 years - it is recommended that patients undergo most of these tests on an annual basis - and more frequently if indications of possible cardiac issues are present.

What is involved with these tests?

A **chest x-ray** is simply a picture of the inside of the chest, including the heart. It can be useful in getting a look at heart size and detecting any unusual swelling.



An **electrocardiogram** (often called "ECG" or "EKG") records electrical activity in the heart and helps detect arrhythmias and assess heart function. Several electrodes are positioned around the chest and attached to a machine which gathers the information over a short period of time (usually just a few minutes).

A **24-hour Holter monitor** is a small ECG machine that the patient wears for 24 hours to get a sustained picture of electrical activity in the heart. As with a regular electrocardiogram, electrodes are attached to the patient's chest, but they connect to the smaller Holter monitor, which can usually be attached to the patient's belt.

A **Cardiac Stress Test** also involves an ECG. Usually, electrodes are attached to the patient and to the ECG machine, after which the patient is asked to undergo some form of physical exercise, such as running on a treadmill. In addition, a blood

pressure cuff is used to monitor blood pressure during the test. The length of the test may vary from a few minutes to perhaps half an hour, depending upon your physical condition and the kind of information your doctor requires. The Cardiac Stress Test helps to determine if your heart is getting the oxygen that it needs.

An **Echocardiogram** is an ultrasound for the heart. Unlike an ECG, in which electrodes are attached to specific areas of the chest, in an echocardiogram a “probe” is placed on the chest and then moved to different positions to get a picture of the heart created through soundwaves.



A **MUGA Scan** is a Multiple Gated Acquisition Scan. In this test, a small amount of a radioactive fluid is injected into the patient’s blood. This fluid is then observed by a special camera, which monitors it as it enters and leaves the heart, allowing the doctor to get a good idea of how well the heart pumps. The MUGA may be conducted under either a state of relaxation or of exercise, depending on what information the doctor is seeking.

A **T2*** reading is obtained from an MRI (Magnetic Resonance Imaging) machine using special software. As with a normal MRI, a patient usually lies down on a flat surface, which is pushed inside of a scanner. The patient may be asked to hold his/her breath during the scans. The T2* is especially

helpful in giving an indirect measurement of how much iron is in the heart and of how that iron is affecting heart function.

A **Functional Cardiac MRI** is an MRI of the heart that tells much of the same information as an echocardiogram. It is increasingly being used because it is more accurate and sensitive to cardiac dysfunction. The T2* and functional MRI are often done together.

What can I do to help prevent cardiac issues?

Aside from making sure you undergo regular testing (as above), there are some lifestyle factors that can play a big role in preventing cardiac issues. The most important of these is to do everything possible to remain well-chelated. Maintaining compliance with an effective chelation therapy is the best method for keeping your heart strong and healthy.

As with people who don’t have thalassemia, making sure that you get an appropriate amount of exercise - whether it is as simple as walking or as involved as going to the gym - is also helpful in maintaining your heart’s health. Discuss with your doctor an appropriate exercise schedule for you - what kind of exercise, how often, for how long, etc.



What you put inside your body can also affect your