

Scleroderma Education Program

Chapter 7

Heart, Lungs and Kidneys

Chapter Highlights

1. Heart Disease in Scleroderma
 - What the heart does
 - What can go wrong
2. Lung Disease in Scleroderma
 - What the lungs do
 - What can go wrong – symptoms of lung disease
3. Kidney/Renal Disease in Scleroderma
 - What the kidneys do
 - What can go wrong

This seventh chapter usually takes about 15 minutes.

Remember:

Many of the things discussed in this chapter are scary. No one with Scleroderma will have all or even most of the problems described in this manual. We want to include most of the problems that could develop in Scleroderma so that all patients will feel informed. It's important to discuss your concerns with your doctor.

Heart Disease in Scleroderma

Who Develops Heart Disease

Many people with Scleroderma do not develop heart disease. Some do.

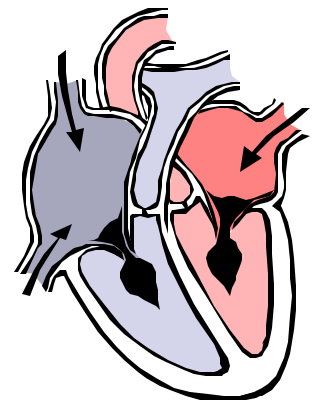
If there is a problem with the heart, the person with Scleroderma may be totally unaware of it at first. That's because there are usually no symptoms of heart disease in the early stages of Scleroderma. Doctors use tests to find out if the heart has been affected.

What the Heart Does

The heart pumps blood to the body and to the lungs

The circulatory system is made up of 2 parts:

1. Circulation to the body (Systemic)
This part sends blood to the body and oxygen to the organs
2. Circulation to the lungs - (Pulmonary)
This part sends blood to the lungs to get oxygen.



The heart has 4 chambers:

- 2 upper chambers (atria).
- 2 lower ones (ventricles).

Blood flows through chambers, valves, and arteries.

- First, blood enters the heart from the body at the right atrium.
- Then, it flows to the right ventricle.
- Then, the pulmonary arteries send the blood to the lungs to get oxygen.
- Then, the blood flows back to the left atrium.
- Then, the blood flows into the left ventricle.
- Finally, the aorta sends blood out to the body.

This flow of blood through the heart is controlled by 4 different valves.

Next we'll review different heart problems that are sometimes seen in Scleroderma patients.

Irregular Heartbeat - Arrhythmias

This is the most common symptom of heart disease in Scleroderma.

Any change from the normal rhythm of your heart is called an arrhythmia (a-RI TH-mee-a). The most common symptom of heart disease in Scleroderma is an irregular heartbeat. The only way to see an irregular heartbeat is on an EKG (electrocardiogram). But a person can feel some of the changes.

Symptoms of irregular heartbeat are:

- ? light-headedness
- ? palpitations
- ? irregular pulse
- ? fainting or dizziness.

Scar tissue can block the natural flow of electricity to the heart.

Heart rhythm is controlled by a natural electrical system. In some cases of Scleroderma, scar tissue gets in the way of the path of this natural electrical current. There can be a partial or complete block to the current, depending on how much scar tissue there is. Your doctor may suggest an EKG to test for an abnormal heart rhythm.

A heart monitor can help pick up an irregular heartbeat.

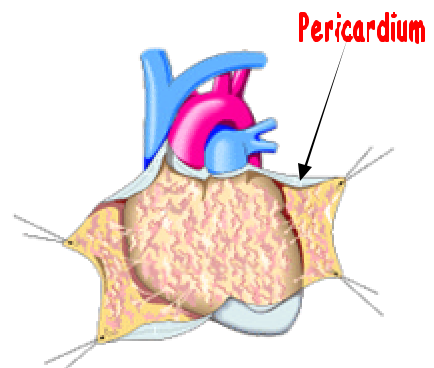
Sometimes the abnormal heart rhythm is noticed by the patient. But sometimes it is not. If you have symptoms and the EKG is normal you may be asked to wear a portable Holter monitor. A Holter monitor measures your heart rhythm for an entire day. It can pick up heart irregularities that happen only once in awhile and may be missed during the short period of time that the EKG is recording.

Treatment is usually not necessary, but medication can be used that helps keep the heartbeat regular. A pacemaker can be used to override the natural electrical system that controls the heartbeat.

Inflammation of the Heart - Pericarditis

You may have chest pain that involves the heart

Another way that scleroderma can affect the heart is that the outside membrane of the heart can get inflamed and swollen. This is called pericarditis.



The pericardium is a sac that surrounds the heart. Too much fluid builds up when the sac gets swollen.

Symptoms of pericarditis are:

- ? chest pain when you breathe deeply
- ? shortness of breath
- ? high fever.

The pain often is worse when you lie down.

You may have chest pain that does NOT involve the heart.

Often chest pain in Scleroderma comes from heartburn or reflux. This type of chest pain usually occurs after eating (See Chapter 3).

A physical exam and some tests will find the problem.

The way to diagnose pericarditis is by a physical examination, EKG and echocardiogram.

Treatment depends on how severe it is.

Your doctor may not give you any treatment if the amount of fluid is very small and you have no symptoms.

Severe Pericarditis is rare in Scleroderma.

Treatment for mild swelling

NO Treatment is given:

If there is a small amount of fluid, and it isn't pushing on the heart, and it doesn't get in the way of heart function, it is often left alone. It is NOT treated with medication, and it causes NO HARM.

Treatment for severe swelling

Two (2) kinds of medications are used:

The type of Drug

- o Steroids
- o Nonsteroidal anti-inflammatory drugs (NSAIDs)

The drug name

Prednisone.
Arthritis medications (e.g. Alleve, Motrin)

Surgery can help

A surgical procedure called a **pericardial window** may need to be done if the amount of fluid is large and is hurting how well the heart works.

This lets the fluid drain out into the lung cavity where there is more space. The extra fluid will slowly get absorbed by your body.

Pericarditis is temporary.

It may happen again, but it will get better with treatment.

Heart Failure

An uncommon heart problem that could occur in the later stages of Scleroderma is heart failure. When scar tissue grows throughout the heart muscle it makes the heart a less effective pump. The heart can't pump out as much blood as it gets. The heart can fail on the right side. Usually failure on this side is due to pulmonary hypertension (see section on lung disease).

Symptoms of Right sided heart failure:

- ? People find it hard to breathe
- ? The legs may become swollen and people may gain weight from retaining more fluids.

Symptoms of Left sided heart failure:

- If the left side of the heart fails there are similar symptoms.
- ? People find it hard to breathe when they are active.
 - ? People may not be able to lie flat in bed and wake up in the middle of the night short of breath.
- These symptoms are due to the body retaining more fluids.

Certain tests can find the problem

Tests such as an echocardiogram or an electrocardiogram (EKG), a stress test and a cardiac catheterization may need to be done to get a good picture of what part of the heart isn't pumping the way it should.

Treatments include medications, such as ACE inhibitors.

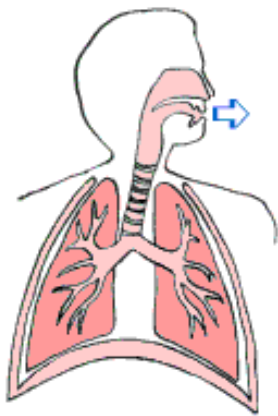
Summary of Heart Problems in Scleroderma

1. **Arrhythmias** are problems with heart rhythm that can be treated with medication or a pacemaker.
2. **Pericarditis** is an inflammation of the sac that surrounds the heart. Treatment isn't necessary if it isn't causing pumping problems of the heart. When it is more severe, it is treated with medication or with a surgical procedure called a pericardial window.
3. **Heart failure** is the inability of the heart chamber to pump out as much blood as it receives. This is caused by problems with the heart muscle. It is usually treated with medication.

Summary of Cardiac Tests

1. **EKG:** An electrocardiogram. This is done in a doctor's office and measures heart rate and rhythm.
2. **Holter monitor:** A portable EKG monitor usually worn for 24 hours to pick up heart rhythm irregularities that may be present only sometimes and were not found in a usual EKG.
3. **Echocardiogram:** A test of the heart that measures how well the chambers of the heart are pumping, how well the heart valves are working, estimates the pulmonary artery pressures, if pulmonary hypertension (discussed in the next section) is present and if there is any excess fluid in the sac around the heart.
4. **Cardiac catheterization:** A test where a plastic tube is inserted in a large blood vessel, usually in the groin or in the arm and threaded to the heart. IV dye is injected into the heart or into the arteries that supply blood to the heart to see if any vessels are blocked. Pressure in the heart and in the pulmonary artery can also be measured to see if you have pulmonary hypertension.
5. **Cardiac stress test:** A test that sees if the heart is getting an adequate blood supply during exercise.

LUNG DISEASE IN SCLERODERMA



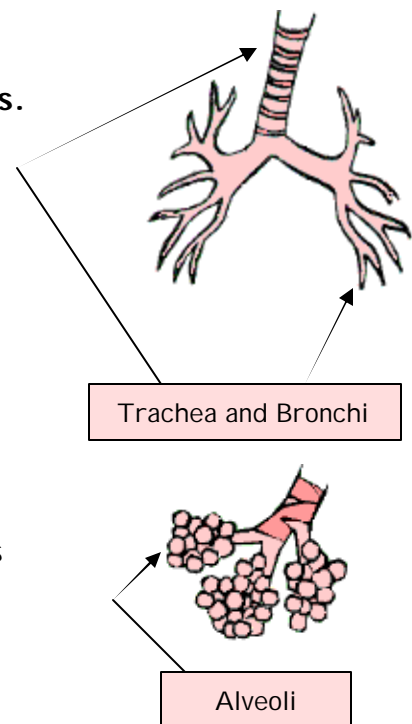
What the Lungs Do

The lungs take oxygen in, and let **carbon dioxide** out.

The lungs are two spongy organs surrounded by a thin, moist membrane called the pleura. The lungs get oxygen from the air we breathe and get rid of carbon dioxide. Carbon dioxide is a waste product from our bodies changing food to energy.

A breath of air travels through pipes, tubes, and sacs.

- When a person breathes in, the air is carried
- ? first, through your **windpipe** (the trachea)
- ? then, through your two flexible airways called **bronchial tubes** (bronchi)
- ? then, into each one of your two **lungs**
- ? then through millions of smaller branching airways called **bronchioles**
- ? then, into millions of even smaller clusters of microscopic sacs called **alveoli**
- ? then, through the thin membranes of these alveoli sacs
- ? then, into the tiny capillaries of the bloodstream
- ? and finally, throughout the entire body



Your breath travels in and out using the same route

Oxygen and carbon dioxide pass to and from capillaries. through the thin membrane of the alveoli sacs.

What Can Damage the Lungs

Because the membrane of the alveoli sacs is made up of collagen, Scleroderma patients sometimes have problems with their lungs. Scleroderma causes too much collagen to be put down in this membrane. This makes it thicker than normal. The thickness makes it harder and harder for the oxygen to get through to the capillaries.

Next we'll review different lung problems that are sometimes seen in Scleroderma patients.

Alveolitis

This is the name used to describe the disease process in the lungs of Scleroderma patients. In alveolitis there is inflammation in the small air sacs. This leads to hardening or fibrosis.

Sometimes this happens slowly, without any symptoms until after a lot of lung damage has happened. In the early stages a chest X-ray may be normal. It will only show up on a CAT scan of the lungs. Pulmonary function tests can pick up changes in lung function, but can't tell the difference between new fibrosis and old fibrosis.

Pulmonary Fibrosis

This is the name for scar tissue buildup in the lungs. It is also sometimes called interstitial lung disease and restrictive lung disease. It can be caused by things other than Scleroderma, but these other causes are rare in patients with Scleroderma. If pulmonary fibrosis shows up on a chest X-ray or pulmonary function tests it is probably due to Scleroderma.

Bronchitis

An inflammation of the bronchi, the main air passages to the lungs. Bronchitis causes a cough that produces mucus, shortness of breath, wheezing and fatigue.

Pleurisy

Although uncommon, another lung problem is called pleurisy. You feel a sharp pain when you are breathing deeply with pleurisy. The space between the walls in the chest becomes inflamed. It is more common in other rheumatic diseases but can occur in Scleroderma. You may also have a fever or a cough.

Pulmonary Hypertension

Serious pulmonary hypertension (high blood pressure in the lungs) is a problem that happens in about 10-15% of Scleroderma patients. It can happen with normal or even low blood pressure as measured in the arm.

What causes pulmonary hypertension?

The pressure in the pulmonary artery needs to be just high enough to push the blood through the lungs. Less pressure is needed than the pressure to push the blood through the rest of the body. The pulmonary pressure can get too high if the size of the lung decreases due to severe pulmonary fibrosis or if the lung's blood vessels are diseased.

How do you know if you have pulmonary hypertension?

Pulmonary hypertension is diagnosed using an echocardiogram with special attention given to the right side of the heart. Mild to moderate elevations in pulmonary pressure are common and may be followed without medications. With higher pressures, a cardiac catheterization may be done to carefully measure the pressure.

What can be done?

When the pulmonary hypertension is severe, many patients do well on oral medications including water pills, blood thinners and vasodilators. Getting oxygen at night helps since oxygen helps to relax the smooth muscle of the pulmonary artery. Most blood pressure medications that work for systemic high blood pressure don't seem very helpful for pulmonary hypertension. Anti-coagulation drugs (medicines that reduce the blood from clotting) may be helpful.

One class of medicines called prostacyclin analogues are helpful. Unfortunately, this medication must be given by a continuous IV (intravenously) because it doesn't work in the pill form. The IV therapy must be given for the rest of the person's life or if severe, until lung transplantation. New pills for Scleroderma pulmonary hypertension are being made and researched.

Symptoms of Lung Disease

Shortness of Breath

In Scleroderma, people can have trouble with their lungs without feeling sick. But the most common symptom of Scleroderma lung disease is shortness of breath. People are not able to breathe easily when they are being active. Walking a distance on flat surface or up a flight of stairs can cause shortness of breath.

The shortness of breath usually takes time to develop and you may not notice it at first. It occurs gradually over time. Sudden shortness of breath is uncommon in Scleroderma and may mean something else, like a lung has collapsed or an infection.

Coughing

Coughing can be a symptom of lung disease in Scleroderma. A cough can occur for a variety of reasons.

- ? One reason is that the membranes in the mouth and the upper airways become dry.
- ? Another reason for a cough can be lung disease. This happens later in Scleroderma when the lung is damaged from scarring. The lung is no longer shaped normally. In this case people may cough to clear mucous. Discolored mucous can be a sign of bronchitis or even an infection.
- ? The third cause of a cough can be because of changes in the bronchial tubes. The breathing tubes may become altered and then they become more sensitive to cold air or dust.
- ? Gastrointestinal reflux disease may also cause you to cough. This occurs when the esophagus is not working properly. The esophagus is the tube that pushes food from the mouth to the stomach. (See chapter 4) When it does not work correctly you may feel like you have to cough. People usually feel like this for short periods at a time.
- ? In rare cases there can be bleeding from the lung, and blood is coughed up. This is very uncommon. It can happen in a small group of people who have both pulmonary hypertension and lung disease. Coughing of

blood may be secondary to infection or lung tumor. Your doctor should be notified immediately.

Chest Pain

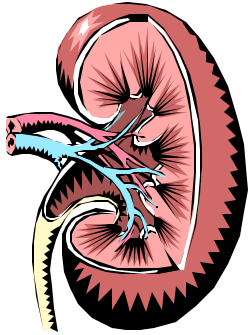
There is usually no chest pain when you are feeling breathless with Scleroderma lung disease. If you do experience some chest pain, it is probably caused by something else. Chest pain in people with Scleroderma is usually felt as an ache. It is usually caused by muscle and joint problems or reflux instead of lung problems.

Summary of Treatment for Alveolitis/Pulmonary Fibrosis

1. **Stop smoking.** Smoking will only make the lung problems of Scleroderma worse and will also make Raynaud's symptoms worse.
2. **Improve reflux.** Use suggestions in Chapter 4 on how to improve Reflux symptoms.
3. **Take medications, as needed.** Medications that suppress your immune system with or without steroids may help if you have alveolitis or pulmonary fibrosis.
4. **Get tested regularly.** If you have non-progressive pulmonary fibrosis you should get regular pulmonary function tests.
5. **Arrange for home oxygen therapy, if needed.** If you have severe fibrosis you may need home oxygen therapy.

KIDNEY/RENAL DISEASE IN SCLERODERMA

Who Develops Kidney Disease



Scleroderma affects the kidneys in about 1 out of 10 people. The good news is that 9 out of 10 people with Scleroderma will never get this complication. The other good news is that there is treatment when it does occur.

The bad news is that it is difficult to know which 10% will have kidney involvement. Kidney problems occur most often in the first five years after the diagnosis. Kidney problems tend to happen in people with diffuse skin involvement (skin thickness that reaches the upper arms and trunk).

What the Kidneys Do

The kidneys are fancy trash collectors.

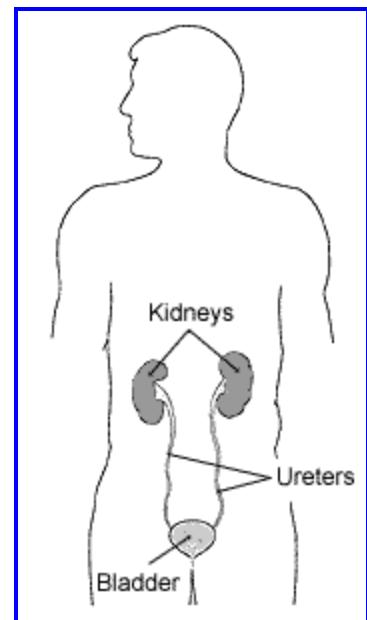
It's helpful to first understand what the kidneys do. Your kidneys are bean-shaped organs that are about the size of your fist. They rest just below your rib cage and near the middle of your back.

Kidneys remove wastes from your blood.

Every day, your kidneys filter about 200 quarts of blood and filter out about 2 quarts of waste products and extra water. The waste and extra water become urine. The urine flows to your bladder and it is stored until you go to the bathroom.

Without kidneys, waste can build up and cause damage.

The wastes in your blood come from the normal breakdown of muscle and from the food you eat. Your body uses the food for energy and self-repair. After your body has taken what it needs from the food, waste is sent to the blood. Without your kidneys removing these wastes, the wastes would build up in the blood and damage your body.



What Can Damage the Kidneys

Sudden and severe high blood pressure can damage the kidneys.

Kidneys can be damaged in Scleroderma due to a sudden and severe increase in blood pressure. This is called **malignant phase hypertension**. This is very different from the usual type of high blood pressure. The usual type of high blood pressure is also known as hypertension. The usual kind of hypertension starts slowly and only causes organ damage after many years. In Scleroderma, blood pressure can go from normal levels to dangerously high levels in a matter of days. This is called a **Scleroderma Renal Crisis**.

Symptoms of Kidney Damage

Kidney damage can occur in a matter of hours or days. When **malignant phase hypertension** continues without treatment the kidneys can be permanently damaged.

Symptoms of Malignant Phase Hypertension: As with the usual type of high blood pressure, there are no symptoms in the early stages. After the first few days, malignant phase hypertension can cause headaches, nausea, and vomiting.

Symptoms of renal failure are a sense of weakness, nausea, vomiting, dizziness, and generally feeling sick.

How to Prevent Kidney Damage

Detect and treat high blood pressure quickly

The way to prevent kidney damage is to detect and treat the Scleroderma-related high blood pressure as soon as possible. Your doctor can tell you whether you are at higher risk for this kind of high blood pressure.

Use a home blood pressure monitor

Patients who may be at high risk should buy and use a home blood pressure monitor. If your doctor suggests you check your blood pressure at home you should measure your blood pressure twice a week. If it starts to go up, your blood pressure should be taken every day. Your blood pressure should not go over 150 (high number) and 90 (the low number).

Call your doctor if your blood pressure is high

If your blood pressure goes over 150 for the high number or 90 for the low number, you should contact your doctor IMMEDIATELY. Contact your doctor if your blood pressure rises above its usual values, even if it does not go beyond these numbers, which are the upper limits for normal blood pressure.

Get your blood pressure down right away

If your blood pressure starts to go up, it is very important to get it down to normal as fast as you can. This means that you need to get frequent blood tests to check how well your kidneys are working. Often the medication dose will have to be changed. Many patients have to be hospitalized if a Scleroderma renal crisis is suspected. Rapid control of the blood pressure is essential.

How to Treat Kidney Disease

If malignant phase hypertension is caught early, the high blood pressure can be brought under control with medication. ACE inhibitors (angiotensin-converting enzyme inhibitors) are most frequently prescribed. Other types of blood pressure medications do not seem to work as well as ACE inhibitors and will not prevent this problem.

TIPS ON BLOOD PRESSURE

Not every headache or every episode of nausea or vomiting is a sign of uncontrolled high blood pressure.

This is why it is important to be able to take your blood pressure at home. People with limited Scleroderma (skin tightness limited to areas below the elbows and below the knees) do not need to take frequent blood pressures.

- ✍ It is important to take your blood pressure at the same time every day. Blood pressure changes quite a bit during the day and on a daily basis.
- ✍ The range of normal pressure is 100-140 for systolic (the upper number) and 60-90 for diastolic (the lower number).

A single reading above this may not be cause for alarm but you should take your blood pressure every day for the next few days.

Summary

Scleroderma can affect your internal organs such as your heart, lungs and kidneys. It is frightening to learn about all the things that can go wrong. But, knowing what symptoms to look for, learning about tests, medications and treatment options can help you play an important role in your health care. Remember, it is very unlikely that you will face all of these complications of Scleroderma. If you have specific questions or concerns, talk with your doctor.