

Lupus Overview

Lupus -- also known as systemic lupus erythematosus -- is a disease of the immune system. Normally, the immune system protects the body from infection. In lupus, however, the immune system inappropriately attacks tissues in various parts of the body. This abnormal activity leads to tissue damage and illness.

Who Gets Lupus?

According to the Lupus Foundation of America, approximately 1.5 million people in the U.S. have lupus. People of African, Asian, and Native American descent are more likely to develop lupus than are Caucasians. Although it can occur in both men and women, 90% of people diagnosed with the disease are women. Women of childbearing age (14 to 45 years old) are most often affected and as many as 1 in 250 people may develop lupus.

What Are the Symptoms of Lupus?

The symptoms of lupus differ from one person to another. Some people have just a few symptoms, while others have many. In addition, there are many different symptoms of lupus because the disease can affect any part of the body. Some of the more common symptoms include:

- Achy joints (arthralgia)
- Unexplained fever (more than 100 F)
- Swollen joints (arthritis)
- Prolonged or extreme fatigue
- Skin rash
- Ankle swelling and fluid accumulation
- Pain in the chest when breathing deeply (pleurisy)
- A butterfly-shaped rash across the cheeks and nose

- Hair loss
- Sensitivity to the sun and/or other light
- Seizures
- Mouth or nose sores
- Pale or purple fingers or toes from cold or stress (Raynaud's phenomenon)

What Problems Can People With Lupus Have?

Many people with active lupus feel ill in general and complain of fever, weight loss, and fatigue. People with lupus also develop specific problems when the immune system attacks a particular organ or area in the body. The following areas of the body can be affected by lupus:

- **Skin** . Skin problems are a common feature of lupus. Some people with lupus have a red rash over their cheeks and the bridge of their nose -- called a "butterfly" or malar rash. Hair loss and mouth sores are also common. One particular type of lupus that generally affects only the skin is called "discoid lupus." With this type of lupus, the skin problems consist of large red, circular rashes that may scar. Skin rashes are usually aggravated by sunlight. A common lupus rash called subacute cutaneous lupus erythematosus is often worse after exposure to the sun. This type of rash can affect the arms, legs, and torso. An uncommon but serious form of lupus rash results in the development of large blisters and is called a "bullous" lupus rash.
- **Joints**. Arthritis is very common in people with lupus. There may be pain, with or without swelling. Stiffness and pain may be particularly evident in the morning. Arthritis may be a problem for only a few days or weeks, or may be a permanent feature of the disease. Fortunately, the arthritis usually is not crippling.
- **Kidneys**. Kidney involvement in people with lupus can be life threatening and may occur in up to half of those with lupus. Kidney problems are more common when someone also has other lupus symptoms, such as fatigue, arthritis, rash, fever, and weight loss. Less often, kidney disease may occur when there are no other symptoms of lupus.
- **Blood**. Blood involvement can occur with or without other symptoms. People with lupus may have dangerous reductions in the number of red

blood cells, white blood cells, or platelets (particles that help clot the blood).

Sometimes, changes in blood counts (low red cell count, or anemia), may cause fatigue, serious infections (low white cell count), or easy bruising or bleeding (low platelet count). Many patients do not have symptoms from low blood counts, however, so it is important for people with lupus to have periodic blood tests in order to detect any problems.

Blood clots are more common in people with lupus. Clots often occur in the legs (called deep venous thrombosis or DVT) and lungs (called pulmonary embolus or PE) and occasionally in the brain (stroke). Blood clots that develop in lupus patients may be associated with the production of antiphospholipid (APL) antibodies. These antibodies are abnormal proteins that may increase the tendency of the blood to clot. Blood can be tested for these antibodies.

- **Brain and Spinal Cord.** Brain involvement is, fortunately, a rare problem in people with lupus. When present, it may cause confusion, depression, seizures, and, rarely, strokes. Involvement of the spinal cord (transverse myelitis) can cause numbness and weakness.
- **Heart and Lungs.** Heart and lung involvement often is caused by inflammation of the covering of the heart (pericardium) and lungs (pleura). When these structures become inflamed, patients may develop chest pain, irregular heartbeat, and accumulation of fluid around the lungs (pleuritis or pleurisy) and heart (pericarditis). The heart valves and the lung itself can also be affected by lupus, resulting in shortness of breath.

What Causes Lupus?

The cause of lupus is unknown. However, there appears to be something that triggers the immune system to attack various areas of the body. That's why suppressing the immune system is one of the main forms of treatment. Finding the cause is the object of major research efforts.

Factors that may contribute to the development of lupus include viruses, environmental chemicals and a person's genetic makeup.

Female hormones are believed to play a role in the development of lupus because women are affected by lupus much more often than men. This is especially true of women during their reproductive years, a time when hormone levels are highest.

The observation that lupus may affect more than one member of the same family has raised the possibility that the tendency to develop lupus may be inherited. Having such a tendency, however, does not predict that a relative will develop lupus. Only about 10% of people with lupus have a close relative with the disease.

Drug-induced lupus can occur after the use of some prescription medications (such as hydralazine and procainamide). These symptoms generally improve after the drug is discontinued.

How Is Lupus Diagnosed?

Lupus is diagnosed when a person has several features of the disease (including symptoms, findings on examination, and blood test abnormalities). The American College of Rheumatology has devised criteria to assist doctors in making the correct diagnosis of lupus. A person should have at least four of the following 11 criteria, either at the same time or one after the other, to be classified as having lupus. These criteria include:

1. **Malar rash**, a "butterfly" rash that appears on the cheeks.
2. **Discoid rash**, red, scaly patches on the skin that cause scarring.
3. **Photosensitivity**, a skin reaction or sensitivity to sunlight.
4. **Oral ulcers** (open mouth sores).
5. **Arthritis**, pain, inflammation, or swelling of the joints.
6. **Kidney disorder**, either excess protein in the urine (proteinuria) or red blood cells in the urine.
7. **Neurological disorder**, seizures, or psychosis.
8. **Inflammation** of the lining around the lungs (pleuritis) or of the lining around the heart (pericarditis)
9. **Blood disorder**, either low red blood cell count (anemia), low white blood cell count (leukopenia), decrease in lymphocytes (lymphopenia), or decrease in blood platelets (thrombocytopenia).
10. **Immunologic disorder**, including the presence of certain cells or autoantibodies, or a false-positive test result for syphilis.
11. **Abnormal blood work**, a positive antinuclear antibody (ANA) test result from blood work.

What Is an Antinuclear Antibody Test

An antinuclear antibody (ANA) test is a sensitive screening tool used to detect autoimmune diseases, including lupus. Antinuclear antibodies (ANAs) are antibodies that are directed against certain structures within a cell's nucleus (thus, antinuclear antibody). ANAs are found in particular patterns in people with autoimmune diseases (those in which a person's immune system works against his or her own body).

An ANA test is done on a sample of a person's blood. The test determines the strength of the antibodies by measuring how many times the person's blood must be diluted to get a sample that is free of antibodies.

Does a Positive ANA Test Mean That I Have Lupus?

Not necessarily. The antinuclear antibody (ANA) test is positive in most people who have lupus, but it also may be positive in many people who are healthy or have another autoimmune disease. Therefore, a positive ANA test alone is not adequate for the diagnosis of lupus. There must be at least three additional clinical features from the list of 11 features for the diagnosis to be made.

How Is Lupus Treated?

The type of lupus treatment prescribed will depend on several factors, including the person's age, type of drugs he or she is taking, overall health, medical history, and location and severity of disease.

Because lupus is a condition that can change over time and is not always predictable, a critical part of good care includes periodic visits with a knowledgeable, accessible doctor, such as a rheumatologist.

Some people with mild features of the disease do not require treatment, while people with serious involvement (such as kidney complications) may require powerful medications. Drugs used to treat lupus include:

- **Steroids** . Steroid creams can be applied directly to rashes. The use of creams is usually safe and effective, especially for mild rashes. The use of steroid creams or tablets in low doses can be effective for mild or moderate features of lupus. Steroids also can be used in higher doses when internal organs are threatened. Unfortunately, high doses also are most likely to produce side effects.

- **Plaquenil** (hydroxychloroquine). Commonly used to help keep mild lupus-related problems, such as skin and joint disease, under control. This drug is also effective at preventing lupus flares.
- **Cytoxan** (cyclophosphamide). A chemotherapy drug that has very powerful effects on reducing the activity of the immune system. It is used to treat severe forms of lupus, such as those affecting the kidneys or brain.
- **Imuran** (azathioprine). A medication originally used to prevent rejection of transplanted organs. It is commonly used to treat the more serious features of lupus.
- **Rheumatrex** (methotrexate). Another chemotherapy drug used to suppress the immune system. Its use is becoming increasingly popular for skin disease, arthritis, and other non-life-threatening forms of disease that have not responded to medications such as hydroxychloroquine or low doses of prednisone.
- **Benlysta (belimumab)**. This drug weakens the immune system by targeting a protein that may reduce the abnormal B cells thought to contribute to lupus. People with active, autoantibody-positive lupus may benefit from Benlysta when given in addition to standard drug therapy.
- **CellCept** (mycophenolate mofetil). A drug that suppresses the immune system and is also used to prevent rejection of transplanted organs. It is being used increasingly to treat serious features of lupus, especially those previously treated by Cytoxan.
- **Rituxan** (rituximab). A biologic agent used to treat lymphoma and rheumatoid arthritis. It is used to treat the most serious features of lupus when other therapies are not effective.

What Is the Outlook for People With Lupus?

The outlook for lupus varies, depending on the organs involved and the severity of symptoms. The disease often includes periods of symptoms followed by periods of remission or lack of symptoms. Most people with lupus can expect to have a normal lifespan, especially if they follow their doctor's instructions and their treatment plans.

What Can Be Done to Improve Quality of Life With Lupus?

There is no cure for lupus, but there are steps you can take to improve your sense of well-being and your quality of life, including:

- **Exercise .** Low-impact exercises, such as walking, swimming, and biking can help prevent muscle wasting and lower your risk for developing osteoporosis (thinning of the bones). Exercise also can have a positive impact on mood.
- **Get enough rest.** Pace yourself, alternating periods of activity with periods of rest.
- **Eat well.** People with lupus should eat a nutritious, well-balanced diet.
- **Avoid alcohol.** Alcohol can interact with your medications to cause significant stomach or intestinal problems, including ulcers.
- **Don't smoke.** Smoking can impair circulation and worsen symptoms in people with lupus. Tobacco smoke also has negative effects on your heart, lungs, and stomach.
- **Play it safe in the sun.** People with lupus may develop rashes or disease flares when exposed to the sun. All lupus patients should protect themselves from the sun; limit time in the sun, especially between 10 a.m. and 2 p.m., wearing sunglasses, a hat, and sunscreen when you are out in the sun.
- **Treat fevers.** Take care of fevers and infections promptly. A fever may indicate an infection or a lupus flare-up.
- **Be a partner in your care.** Build an honest and open relationship with your doctor. Be patient. It often takes time to find the right medication and dosage that works best for you. Also, follow your doctor's treatment plan and don't be afraid to ask questions.
- **Get to know your disease.** Keep a record of your lupus symptoms, which parts of your body are affected and any situations or activities that seem to trigger your symptoms.
- **Ask for help.** Don't be afraid to recognize when you need help and to ask for it. Consider joining a support group. It often helps to talk to others who have been through similar experiences.

Living With Someone Who Has Lupus

If someone close to you has lupus, your life will likely be affected, as well. It is important to understand your loved one's illness and what he or she may be expecting from you. The following are some tips for living with someone who has lupus:

- Learn about lupus and its treatment. Understanding the illness can help you know what to expect, and provide better support and understanding.
- Don't push. Give your loved one enough space to deal with the illness and regain some control over his or her life.
- When possible, go with the person to the doctor. This is a good way to offer support and to listen to what the doctor says. Sometimes, a person feels overwhelmed and cannot take in everything the doctor says.
- Encourage the person to take care of himself or herself and to follow the doctor's treatment plan, but do it gently. Be patient and don't nag.
- Be open with the person. Talk about your own fears and concerns, and ask the person about his or her fears and needs.

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[Sources](#) ^

SOURCES:

The Lupus Foundation of America.

American College of Rheumatology.

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