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# Lupus

## WHAT IS LUPUS?

Lupus refers to several forms of a disease of the immune system that affects joints, skin, kidneys and other parts of the body. The immune system is your body's natural defense against infections, such as bacteria and viruses. In lupus, the immune system produces antibodies that react with the body's own tissues. Because of this, lupus is referred to as an autoimmune (aw-toe-im-MYOON) disease. In most cases the term "lupus" refers to the form known as systemic lupus erythematosus (sis-TEM-ick LOO-pus e-RIH-them-ah-TOE-sis), or SLE for short.

Lupus is an inflammatory condition that may be chronic. Inflammation refers to a reaction that results in pain, heat, redness and swelling. Chronic (cron-NIK) means the condition is long-lasting, which could mean it lasts for the rest of your life. You may not experience symptoms on a constant basis, however.

Many people with lupus have changes in signs and symptoms known as flares and remissions. A flare is a period when the disease becomes more active with increased symptoms. During a remission, few or no signs and symptoms of lupus

are present. Sometimes a person may have a complete or long-lasting remission, but this does not necessarily mean the disease has gone away.

## Systemic Lupus

About 70 percent of the people who have lupus have the systemic form, or SLE. A systemic disease is one in which several different body systems may be affected. In systemic lupus, the skin, joints, kidneys, nervous system, lungs, heart and/or blood-forming organs can be affected.

About half of the people with systemic lupus have a form that affects major internal organs, particularly the kidneys. In the other half of people with systemic lupus, mainly the skin and joints are affected. This second type is less likely to cause serious problems.

## Discoid Lupus

About 15 percent of people with lupus have a form known as discoid (DIS-coyd) lupus or cutaneous (cue-TANE-ee-us) lupus. This form of lupus results in a chronic skin rash that can sometimes cause scars. Discoid lupus may affect the skin without affecting other organs.

## Drug-Induced Lupus

The least common form of lupus develops as a result of drugs taken for other medical problems. This form is called drug-induced lupus. Signs and symptoms are similar to SLE, although people with this form of lupus rarely develop serious organ damage. Many different drugs can cause drug-induced lupus. Signs and symptoms usually improve and disappear once the drug is stopped.

## WHAT CAUSES LUPUS?

The cause of lupus, with the exception of drug-induced lupus, is unknown. Doctors and scientists refer to lupus as an autoimmune disease. The immune system fights off bacteria and viruses in several ways. One way is by creating special types of blood proteins called antibodies that attack and destroy invading substances.

In lupus, the immune system does not function properly, and produces antibodies (called autoantibodies) that react with and damage the body's cells, tissues and organs. This process is known as an autoimmune response (auto means self).

In lupus, the many different types of autoantibodies are formed, although the most common autoantibody is produced against the nucleus of cells, and therefore called antinuclear antibodies, or ANA for short. There are several different types of ANA. Types of ANA such as anti-double stranded DNA (anti ds-DNA) or anti-Smith (Sm) antibodies, are unique to SLE.

Antinuclear antibodies are found in almost all cases of lupus. ANA also may be found in people with other autoimmune diseases, such as rheumatoid arthritis (ROO-ma-toyd ar-THRY-tis), or RA, and Sjögren's (SHOW-grens) syndrome, and even can be found in some healthy people.

## WHO GETS LUPUS?

Studies suggest that some people may inherit the tendency to get lupus, and that genes play an important role in the disease. Researchers have found that new cases of lupus are more common in families in which someone already has the disease or related autoimmune disease, such as RA or diabetes (di-uh-BEE-tees). Most scientists believe that an environmental factor, such as a virus, serves to trigger symptoms in people who have a genetic tendency to develop lupus.

About 90 percent of people with lupus are women. In most cases, symptoms first appear in women of childbearing age (18 to 45). But lupus also occurs in children and in older people. African Americans tend to get lupus more often than Caucasians. Some studies suggest the disease also may occur more often in Asian and Hispanic populations than in Caucasians.

## SYMPTOMS OF LUPUS

You may develop several of the symptoms mentioned in this section or just a few. No two people with lupus have the same symptoms.

The American College of Rheumatology (ACR) has developed guidelines to help doctors diagnose lupus. If you have four or more of the following 11 symptoms listed, it is likely that you have lupus or a similar condition.

### Main Symptoms and Signs

If you have four or more of the signs or symptoms listed below, talk to a doctor who can determine whether you have lupus or one of many other conditions that can cause similar symptoms. The 11 main symptoms and signs that may indicate lupus include:

- A rash across the cheeks and the bridge of the nose (called a "butterfly rash")

- Scaly, disk-shaped rash on the face, neck, ears, scalp and/or chest
- Sensitivity to sunlight, such as severe rashes or fever from minimal sun exposure
- Painless sores on the tongue, inside the mouth and/or in the nose
- Arthritis (pain, stiffness and swelling in the joints)
- Pain in your chest and side when you breathe, indicating inflammation of the lining of the heart (pericarditis) or lungs (pleurisy)
- Kidney problems
- Neurologic (brain) problems, including seizures and mental problems
- Low white or red blood cell count
- Presence of specific autoantibodies measured in the blood
- The presence of antinuclear antibodies, the most commonly seen autoantibody in SLE

### *Other Symptoms and Signs of Lupus*

People with lupus can develop other symptoms and signs, including serious organ involvement, not included in the 11 criteria described above. These symptoms and signs include:

- Blood clots
- Strokes
- Heart attacks
- Eye inflammation
- Fever
- Weakness, fatigue
- Weight loss
- Raynaud's (ray-NODES) phenomenon, a condition in which the fingers, toes, nose and/or ears may become unusually sensitive to cold and may turn white or blue when exposed to the cold.
- Muscle aches
- Swollen lymph nodes
- Loss of appetite

- Hair loss
- Sjögren's syndrome, a chronic condition that causes dryness of the eyes and mouth. Women may experience vaginal dryness.
- Depression or difficulty concentrating

### **DIAGNOSIS**

Lupus can be hard to diagnose. It is important to see a rheumatologist (ROO-ma-tall-o-jist), a doctor who specializes in arthritis and related diseases, like lupus. The rheumatologist will begin by asking you questions and conducting a physical exam. You'll also have laboratory tests, including ones to see if you have too few red blood cells, white blood cells or platelets (blood cells that help to control bleeding and clotting). Blood chemistry tests and urine studies can help determine whether your organs, such as the kidneys and liver, are functioning normally.

If your rheumatologist thinks you may have lupus, he or she will order a blood test called an ANA, which detects a group of autoantibodies found in the blood of people with lupus. These autoantibodies attack the body's own cells. However, this test does not diagnose lupus.

Other tests to find certain antibodies, such as anti-DNA or anti-Smith, may be helpful in diagnosing lupus or related diseases. Antibodies to phospholipids, including cardiolipin, also are common in lupus. They are associated with an increased risk for blood clotting, strokes and recurrent miscarriages. Tests to measure the level of complement proteins in your blood may be obtained. Levels of complement are often low or reduced in people with lupus.

Other blood tests may be conducted for diagnosis and as an aid in following disease activity. Because kidney problems often occur, you'll need a urinalysis, which is an examina-

tion of your urine. If protein is found, you may be asked to collect all the urine you pass in a 24-hour period for analysis. If your doctor suspects kidney problems, you also may have a kidney biopsy. This is when a small piece of tissue from one of your kidneys is removed and examined. This procedure requires an overnight hospital stay.

Your doctor may request a chest X-ray or heart studies such as an electrocardiogram (EKG) or an echocardiogram to determine if the disease is affecting your lungs or heart.

## TREATMENTS

The treatment plan for lupus includes taking medications to reduce the inflammation and reduce the activity of the immune system, balancing rest with exercise and eating a proper diet.

Lupus is an unpredictable disease. Signs of the disease appear and disappear, sometimes for no apparent reason. Because lupus has so many different forms and can change, finding the right treatment for you may take time. Your treatment will depend on the symptoms you experience and the organs affected. Once an effective treatment program has been started, continue to follow it. If your symptoms change, let your doctor know so that you can work together to adjust your program.

## Medications

Medications are a necessary part of treatment for most people with lupus. The particular medication prescribed by your doctor will depend on the extent of disease and how active it is. The type and amount of medications prescribed may change over time, depending on the signs and symptoms of lupus that are present.

Many medications can take several weeks or months to start working. You should not change

the amount or frequency of medication you take without talking to your doctor. Changing your schedule on your own can make it more difficult for your doctor to evaluate what is happening in your disease and how the medication is working. Taking a drug without medical advice could cause harm, either by interacting with other medications you take or by causing side effects.

## NONSTEROIDAL ANTI-INFLAMMATORY DRUGS

**What they do:** Nonsteroidal anti-inflammatory drugs (NSAIDs) can help control the arthritis and inflammation associated with lupus. Some examples of NSAIDs include aspirin, ibuprofen and naproxen. NSAIDs are available in prescription and over-the-counter forms.

**Side effects:** NSAIDs can cause stomach irritation, bleeding from the stomach or intestines, ulcers or diarrhea. Your doctor may prescribe anti-ulcer medications to help protect you from these side effects. Newer NSAIDs called COX-2 specific inhibitors may further minimize stomach side effects.

**Tips:** Some people may feel stomach upset when they take large doses of NSAIDs. You may be able to ease this side effect by taking your medication with meals or with a large glass of water or milk. You may want to try safety-coated tablets, which help protect the stomach. Taking an antacid about 30 minutes after meals and at bedtime also can help protect your stomach.

NSAIDs can affect the platelets in the blood and can extend bleeding time. If you are scheduled for surgery, you may need to temporarily stop taking these drugs. Care should be taken when using NSAIDs with blood thinners, such as warfarin. COX-2 specific inhibitors do not increase bleeding time. Most NSAIDs can rarely damage the liver and decrease kidney function.

The damage usually can be reversed when you stop taking the medication or take it in smaller doses.

#### DISEASE-MODIFYING ANTIRHEUMATIC DRUGS

**What they do:** Disease-modifying anti-rheumatic drugs (DMARDs) may be used to treat symptoms of lupus, such as arthritis, skin rashes and pleurisy, which is inflammation of the lining of the lungs. These drugs can help treat and prevent lupus rashes and systemic problems such as fever and fatigue.

The most common DMARD used for treating lupus is hydroxychloroquine. Prescribing hydroxychloroquine for mild disease may decrease the risk of lupus spreading. These medications are usually prescribed long term, because stopping them may result in a lupus flare.

**Side effects:** The most common side effects of hydroxychloroquine are mild nausea, vomiting and diarrhea.

#### Medication Tip

In rare cases that usually follow prolonged use, hydroxychloroquine may injure the retina, the lining of the eye that enables you to see. Early detection of this unusual occurrence can minimize damage. See an eye specialist who is familiar with this toxicity regularly – usually every six months – to help reduce the slight risk from this drug. If you notice any changes in your vision while taking hydroxychloroquine, call your doctor right away.

#### GLUCOCORTICOIDS

**What they do:** Glucocorticoids (such as prednisone) are drugs that restrain the immune system and help to reduce inflammation. They are

related to cortisol, a hormone that occurs naturally in the body and controls many necessary body functions.

During times when your lupus is very active, your doctor may prescribe large doses of glucocorticoids. Lower doses may be used to treat lupus when it doesn't threaten internal organs. As symptoms and laboratory test results improve, your doctor may reduce the dose over time.

**Side effects:** Common side effects of high doses of glucocorticoids include weight gain, rounding of the face, easy bruising, slow healing and mood swings. Other possible side effects include fluid retention with swelling of the legs and high blood pressure; triggering or worsening of diabetes; increased risk of infection; the death of bony tissue, called osteonecrosis (ah-stee-oh-ne-CRO-sis); acne; and, rarely, stomach bleeding.

Using glucocorticoids over a long period of time may lead to cataracts, glaucoma, muscle weakness and/or thinning of the bones, called osteoporosis (ah-stee-oh-po-RO-sis). Your doctor may recommend that you take calcium and vitamin D supplements or other medications to help prevent bone loss.

**Tips:** If you are taking glucocorticoids, tell any doctor or dentist before you have surgery or undergo any other type of invasive procedure. Your doctor may need to increase your dosage at such times to enable your body to handle the stress. You also should wear a medical identification bracelet to alert others that you take glucocorticoids in case you cannot tell them during a medical emergency.

#### OTHER IMMUNOSUPPRESSIVE DRUGS

**What they do:** Most immunosuppressive drugs reduce the activity of the immune system. These drugs often are prescribed with gluco-

corticoids when lupus affects major organs. This combination may allow a reduction in your dosage of glucocorticoids.

The immunosuppressive drugs most often used for lupus are azathioprine, methotrexate and cyclophosphamide. Cyclosporine and mycophenolate mofetil also are used.

### *DHEA Supplements*

DHEA is short for dehydroepinandrosterone, a weak male hormone found naturally in the body. Nutritional supplements of this chemical are made from extracts of the wild Mexican yam. This supplement has been touted as a remedy for a variety of diseases, including lupus. Some studies have found evidence that DHEA may help to ease certain lupus symptoms and reduce the need for medication in patients with lupus that affects only the skin and joints.

How DHEA works is not completely understood and the full risks and benefits are not yet known. People with lupus should not take this supplement without first seeking a doctor's advice. Because nutritional supplements are regulated differently from drugs, the quality and quantity of ingredients in DHEA products may vary widely. If you are considering taking DHEA, consult your doctor about the potential risks and benefits.

**Side effects:** These drugs can have serious side effects. If you are taking an immunosuppressive drug, you should have regular blood tests because the drug can interfere with the proper formation of blood cells. You also may get infections more easily because the drugs

reduce the activity of the immune system. Tell your doctor if you develop a fever or any other new symptoms while taking these medications.

### **Physical Activity**

Exercising every day is very important. Working out may be easier when your lupus is less active and you feel better. However, even during a flare, doing gentle range-of-motion (ROM) exercises can keep you from feeling stiff. Exercise also can help prevent weak muscles so that your muscles are better able to support sore joints.

A physical therapist can work with you to set up an exercise program that fits your schedule, physical condition and other needs. This may be as simple as designing a walking program you can adjust to fit how you feel each day. In fact, simple ROM exercises such as stretching can be as effective in improving your mood, sense of well-being and fatigue level as aerobic activity.

### **Diet**

Eating a balanced diet is an important part of your treatment plan. At times when your lupus is active and your appetite is poor, taking a daily multivitamin may help your body get the nutrients it may be missing. However, high doses of vitamins can have side effects. Some herbs and dietary supplements can make your lupus worse. Be sure to tell your doctor about any vitamins, herbs or dietary supplements you take.

Your doctor may suggest a special diet to help problems caused by lupus symptoms, such as kidney disease. A low-salt diet will help prevent fluid retention that can lead to swelling. If you have kidney disease, you also may need to limit how much protein you eat so that you don't overwork your kidneys.

Simply taking a daily dose of folic acid can help lower levels of a chemical called homocysteine (ho-mo-SIS-teen). High levels of homocysteine, which have been linked with heart problems, have been found in people with lupus. Ask your doctor about the recommended dose of folic acid to help reduce homocysteine. High cholesterol also may be a problem; discuss this with your doctor.

Drinking alcohol has the potential to harm your liver. You should avoid drinking alcohol if you take certain drugs such as methotrexate, which increases the risk of liver damage.

## SPECIAL CONCERNS

### Kidney Problems

Sometimes, even with the use of glucocorticoids and immunosuppressive drugs, kidney failure may occur. If this happens, dialysis (di-AL-ih-sis) can help cleanse waste products from the blood. One procedure, called hemodialysis, involves placing catheters in an artery and a vein to filter the blood through a cleansing machine. Blood is taken from the body, run through a machine that removes poisons and waste products, and then returned to the body.

Continuous ambulatory peritoneal dialysis (CAPD) is another cleansing procedure in which fluid is placed into and then drained from the abdominal cavity through a tube. This can be done at home, but careful supervision is necessary because people with lupus have an increased risk of infection.

When kidney failure is irreversible, a kidney transplant may be recommended. This procedure often can allow a person with lupus to return to a normal lifestyle, although many medications are required to prevent rejection of the new kidney.

### Heart Problems

People with lupus are at increased risk for heart attacks. This may be because lupus can damage blood vessels. Other heart disease risk factors, such as hypertension (high blood pressure), smoking, obesity and high cholesterol and homocysteine levels may further damage the blood vessels. Consult your doctor for ways to reduce or manage these risks.

### Infections

People with lupus are more likely to get infections. This is because the immune system may not be working properly and because of medication side effects, especially from glucocorticoids and immunosuppressive drugs. For some people with lupus, infections can trigger disease flares. If you have a fever, which could indicate infection, talk to your doctor. Yearly flu shots and other vaccines are important to protect people with lupus from infections.

### Sun Exposure

Exposure to the sun and certain fluorescent lighting is known to worsen the skin rashes and/ or other systemic problems linked with lupus in some people. About one-third of people with lupus are sun-sensitive. Sun exposure may cause lupus flares, resulting in fever, joint pain or even inflammation of the heart, lungs, kidneys or nervous system. Too much sun exposure for one person, however, may be fine for someone else. Degrees of sun-sensitivity vary from person to person.

### Sun Safety

A few simple precautions can help protect you from negative reactions to sun exposure. Fol-

low these steps to help prevent lupus-related rashes and sunburns from sun-sensitivity.

- Avoid outdoor activities during peak sunlight hours (usually 10 a.m. - 4 p.m.).
- Use sunscreen lotions or sun block regularly. Choose one with a sun protection factor (SPF) of 15 or higher. Like all makeup and lotions, sunscreens may cause allergic reactions in some people. If you have a reaction, stop using that product and try a different one or consult your doctor.
- Look for products that block both UVA and UVB rays – the label should note if the product blocks both types of ultraviolet rays. SPF ratings apply only to UVB light; ultraviolet light exposure is greater at higher altitudes.
- If you must be out in the sun, wear a hat to shade you from direct sun and clothing to cover your arms, legs and chest. Clothing made from special sun-blocking fabrics is available.

### Pregnancy and Birth Control

Pregnancy may present special problems for women with lupus. Lupus may worsen, or flare, during a pregnancy. Alternately, lupus may get better or stay the same.

Women with lupus have an increased chance of having a miscarriage either early or late in their pregnancy. Certain abnormal antibodies present in the blood of some women with lupus may increase the chance of miscarriage. Be sure to consult your doctor about any plans to become pregnant. The good news is that with careful planning, many women with lupus can have normal pregnancies and healthy babies.

In some cases, a woman with lupus can pass autoantibodies (anti-Ro and anti-La) through the placenta, which can cause a rash in the baby. The

rash eventually goes away, usually within six months after the baby's birth. A very rare heart problem can occur in the baby of a woman who has this specific autoantibody pattern. Talk to your doctor about this risk and options for treatment.

If your doctor advises against pregnancy, or if you do not wish to have children, it's important to use birth control because you could still become pregnant. An unplanned pregnancy during a period of active disease could cause health problems and expose the growing fetus to medications you take for lupus.

The safest method of contraception for women with lupus is a diaphragm used with contraceptive jelly. Take special care and consult your doctor about the benefits and risks of using intrauterine devices (IUDs) because they carry a risk of infection. Studies on the safety of birth-control pills for women with lupus are under way. Such information about their safety should be available in a few years. Note: Women with antiphospholipid (an-ty-fos-FO-li-pid) antibody should consult with their doctors prior to taking birth-control pills or estrogen replacement therapy (ERT) because of increased risk of thrombosis (blood clots).

### Dryness

Post-menopausal women with lupus or those who also have secondary Sjögren's syndrome may experience vaginal dryness. Lubricating products and estrogen creams can help restore moisture and ease discomfort. If you experience vaginal dryness, talk to your doctor about which products may be right for you.

### MANAGING LUPUS

Emotional problems often go along with having a chronic disease. Talking about your fears, anger and depression usually helps. It is



important to confide in your family, friends and doctor. Joining a support group and sharing your frustrations with others who have lupus may make you feel better emotionally and help you solve specific problems.

Reading about lupus can help you understand your symptoms and what to expect from the disease. You'll feel more in control of your condition and be able to communicate more effectively with your doctor about treatments.

There may be times, though, when attending support groups or reading about lupus may not be enough to overcome a sense of feeling overwhelmed. In that case, seeking counseling from a professional trained in helping people with chronic diseases may be helpful. Your doctor or local chapter of the Arthritis Foundation can give you suggestions about agencies and mental health professionals such as social workers, counselors and psychologists who may be able to offer their support.

## LUPUS RESEARCH

Researchers are gaining knowledge that will contribute to improvements in the quality of life of people with lupus. This information is leading to the development and testing of various techniques for helping people with lupus gain the knowledge, attitudes and coping skills they need to take control of their condition.

Ongoing Arthritis Foundation-funded studies are aimed at finding better ways to control lupus and improve the quality of life of those affected by it. Some examples include:

- Studies to increase understanding of how best to prevent premature cardiovascular disease in women with lupus;
- Studies of how estrogens foster the production of autoantibodies; and

- A study of people with SLE who have difficulty with memory and concentration to uncover associated signs, symptoms, medications, and behavioral and psychosocial impact.

## Alliance for Lupus Research

The Alliance for Lupus Research (ALR) is a new, nonprofit organization formed as a joint undertaking by the family of Robert Wood Johnson IV and the Arthritis Foundation to provide an aggressive and concerted effort to prevent, treat and cure systemic lupus erythematosus (SLE), or lupus. For more information about ALR, please call 800/867-1743 or visit [www.lupusresearch.org](http://www.lupusresearch.org)

## THE ARTHRITIS FOUNDATION

The mission of the Arthritis Foundation is to improve lives through leadership in the prevention, control and cure of arthritis and related diseases.

The Arthritis Foundation supports research with the greatest potential for advances and has invested more than \$320 million in these efforts since its inception in 1948. Additionally, the Arthritis Foundation supports key public policy and advocacy efforts at a local and national level in order to make a difference on behalf of 70 million people living with arthritis.

As your partner in taking greater control of arthritis, the Arthritis Foundation also offers a large number of programs and services nationwide to make life with arthritis easier and less painful and to help you become an active partner in your own health care.

Contact us at (800) 283-7800 or visit us on the Web at [www.arthritis.org](http://www.arthritis.org) to become an Arthritis Advocate or to find out how you can become involved.

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**For more information:** The Arthritis Foundation offers a wide variety of books, brochures and videos about different forms of arthritis, treatment and self-management techniques to help you take control of your arthritis. To order any of these products, become an Arthritis Foundation member or to subscribe to the Arthritis Foundation's award-winning consumer health magazine, *Arthritis Today*, call (800) 283-7800. Call or visit our Web site ([www.arthritis.org](http://www.arthritis.org)) to find out how you can take control of your arthritis and start living better today!

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This brochure has been reviewed by the  
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**MISSION STATEMENT:**

The mission of the Arthritis Foundation is to improve lives through leadership in the prevention, control and cure of arthritis and related diseases.



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