

# What You Should Know About Lupus

**Y**ou've been diagnosed with lupus, and you may feel both relieved at knowing the cause of your symptoms and overwhelmed by fears and questions. You're probably angry, depressed, worried about work and family, and afraid that you might die. What you need to know is that help is available, and that you have more control over the situation than you may think.

## What is lupus?

Lupus, more correctly called *systemic lupus erythematosus* or *SLE*, is a chronic condition that can cause inflammation in many parts of the body, including the skin, joints, blood, and kidneys. It's an *autoimmune* condition in which the body attacks its own cells. The body can't identify "invaders" (*antigens*) such as viruses and bacteria, and begins to manufacture disease-fighting proteins (*antibodies*) that target healthy cells. These *autoantibodies* cause *immune complexes* to accumulate in the tissues, leading to inflammation, injury, and pain. Lupus is usually mild to moderate, but in some cases it can have severe, life-threatening complications.

## What causes lupus?

While the exact cause of lupus is unknown, it's probably due to a combination of environmental and genetic factors. Patients with a genetic predisposition may develop lupus when exposed to environmental triggers such as infections, antibiotics and other medications (especially sulfa and penicillin drugs), ultraviolet light, extreme stress, or hormones.

*This Patient Handout was prepared by Patricia L. Van Horn using material from IntelliHealth/Aetna (<http://www.intelihealth.com/IH/ihIHLH>), the Lupus Foundation of America (<http://www.lupus.org/>), and WebMD (<http://my.webmd.com/index>).*

However, only 10% of lupus patients have a close relative with the disease, and only about 5% of children born to women with lupus develop the disease.

## Who gets lupus?

The total number of lupus patients in America may be as high as 1.5 million. Lupus appears to favor women in their 20s and 30s; in fact, 90% of patients are women. It tends to occur more often, and with greater severity, in blacks, Native Americans, and Asian Americans. The female hormone estrogen may account for the predominance in women, and may also explain why symptoms tend to increase before menstrual periods and during pregnancy.

## Are there different forms of lupus?

Yes. The three main types of lupus are: *Discoid (cutaneous) lupus*, which only affects the skin. Its main feature is a rash on the face, neck, and scalp. Discoid lupus progresses to affect the organs in about 10% of patients. *Systemic lupus*, which can attack almost any organ or system in the body. Most lupus patients have the systemic form. Some patients only have skin and joint symptoms, while others have symptoms in the joints, lungs, kidneys, blood, and other organs. No two patients with systemic lupus have the same symptoms, although most have healthy periods (*remissions*) alternating with periods of symptom activity (*flares*).

*Drug-induced lupus*, which can occur after the use of certain prescription drugs such as hydralazine or procainamide. The symptoms generally disappear when the drug is discontinued.

## How is lupus diagnosed?

Lupus symptoms tend to mimic those of other illnesses, making diagnosis difficult. You may need to visit several doctors, including a *rheumatologist* or *immunologist*. No single test can detect lupus, so diagnosis generally involves a complete medical work-up, plus routine and specialized laboratory tests. A patient is considered to have lupus if she has four or more of the symptoms (not necessarily all at the same time). The symptoms include most of those listed above, plus:

- Inflammation of organ linings (*serositis*) in the lungs (*pleuritis*) or heart (*pericarditis*)

## Symptoms of Lupus

- Joint aches (*arthralgia*)
- Fever
- Swollen joints (*arthritis*)
- Extreme fatigue
- Red, circular rashes on the body (*discoid rash*)
- "Butterfly" rash on the cheeks and nose (*malar rash*)
- Low red blood cell count (*anemia*)
- Kidney disorders
- Chest pain (*pleurisy*)
- Sun sensitivity (*photosensitivity*)
- Hair loss (*alopecia*)
- Abnormal blood clotting
- Fingers turning white or blue in the cold (*Raynaud's phenomenon*)
- Seizures or confusion
- Ulcers in the mouth, nose, and/or vagina.

- Blood disorders such as anemia, low white blood cell count (*leukopenia*, *lymphopenia*) or low platelet (clotting cell) count (*thrombocytopenia*)
- Presence of *antinuclear antibodies* (ANA), a particular type of autoantibody
- Immune disorders associated with certain other autoantibodies.

It may also be helpful to measure blood levels of complement, a protein that combines with antibodies to attack bacteria. Low C3 or C4 complement values with a positive ANA result is highly suggestive of lupus.

### What causes lupus flares?

Flares can be linked to a number of factors. Triggers can include sun exposure, an infection such as a cold, use of a certain drug, or pregnancy. In many cases, though, there is no obvious trigger.

### Is there any way to tell how lupus will affect a patient?

Lupus is an unpredictable disease, and there is no sure way to know who will develop complications. Prompt treatment of flares is the key, because untreated attacks can cause lasting organ and joint damage, eye problems, atherosclerosis (“hardening of the arteries”), diabetes, and mental disturbances (memory loss, psychosis). With proper therapy, however, most patients can avoid serious damage.

### How is lupus treated?

There is no cure for lupus, so therapy is mainly directed at alleviating symptoms. Effective treatment can usually reduce inflammation and help organs to function normally. Measures to decrease flares include avoidance of sun exposure and regular application of sunscreens, an exercise regimen to prevent weakness and fatigue, immunization against specific infections, and stress relief through support groups, counseling, and help from family, friends, and doctors. “Don’ts” include smoking, alcohol consumption, misuse of prescription drugs, and postponing medical check-ups. The medications prescribed for lupus depend on the organ(s) involved and disease severity.

*Nonsteroidal anti-inflammatory drugs* (NSAIDs) reduce muscle and joint pain. Choices include aspirin, ibuprofen (Motrin, Advil), naproxen (Naprosyn, Aleve), indomethacin (Indocin), nabumetone (Relafen), tolmetin (Tolectin), sulindac (Clinoril), diclofenac (Voltaren), piroxicam (Feldene), ketoprofen (Orudis), diflunisal (Dolobid), etodolac (Lodine), and oxaprozin (Daypro). These drugs may cause stomach problems, and are best taken with meals, antacids, or prostaglandins such as misoprostol (Cytotec); newer drugs like diclofenac plus misoprostol (Arthrotec) combine medications. NSAIDs can also lead to kidney problems, so monitoring is essential.

*Corticosteroids* are hormones that decrease inflammation and suppress immune activity, and include prednisone (Deltasone), methylprednisolone (Medrol), or dexamethasone (Decadron). These drugs carry a high risk of side effects such as weight gain, rounding of the face, acne, bruising, bone thinning (*osteoporosis*), hypertension, cataracts, diabetes, greater susceptibility to infection, stomach ulcers, hyperactivity, and increased appetite. Therefore, it’s important to regulate the dosage for maximum benefit and minimum risk.

*Antimalarials* like chloroquine (Aralen) and hydroxychloroquine (Plaquenil) can

help skin and joint symptoms, but may take months to work. They have few risks, but it’s important to have your eyes checked regularly.

*Immunomodulating drugs* such as methotrexate (Rheumatrex, Folex, Methotrexate LPF), cyclosporine, azathioprine (Imuran), and cyclophosphamide (Cytoxan) are immunosuppressive agents. Possible side effects include anemia, leukopenia, and greater susceptibility to infection. They may also predispose some patients to developing cancer in the future. Finally, there are drugs that target specific immune cells and inhibit antibody production, and *anticoagulants* to prevent excessive blood clotting.

### Can women with lupus have a healthy pregnancy?

Women with systemic involvement including the central nervous system, kidneys, heart, or lungs should consult with their internist or a maternal/fetal specialist prior to conception. Otherwise, there is no compelling reason why a woman with lupus should avoid pregnancy. However, disease activity often increases during or immediately after pregnancy, and close monitoring is crucial. The main risks are high blood pressure (*hypertension*) and *antiphospholipid syndrome*, which can cause clotting problems that lead to miscarriage.

### Can patients with lupus ever lead a normal life?

The idea that lupus inevitably ends in an early death is outdated and untrue. Although a small number of patients develop fatal complications, those without multiorgan involvement can look forward to a normal lifespan if they follow their doctor’s orders, take drugs as prescribed, and seek help promptly for lupus symptoms or drug side effects. Given the great progress in treating lupus during the last decade, it only makes good sense to do everything you can to control your disease, because tomorrow is very likely to bring still better therapy or even a cure.

### Resources

For more information...

#### National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse

1 AMS Circle  
Bethesda, MD 20892-3675  
Phone: 877-226-4267  
E-mail: [niamsinfo@mail.nih.gov](mailto:niamsinfo@mail.nih.gov)  
Website: <http://www.nih.gov/niams/>

#### Lupus Foundation of America, Inc.

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