



## SYSTEMIC LUPUS ERYTHEMATOSUS (LUPUS)

Systemic Lupus Erythematosus, usually referred to as SLE or lupus, is sometimes labeled the “great imitator.” Why? Because of its wide variety of symptoms, it can often be confused with other disorders. Lupus, which affects the joints, kidneys, and skin, can be fatal. However, there is much reason for hope. Improvements in therapy have significantly increased these patients’ quality of life and their life expectancy.

### Fast facts

- Lupus affects 10 times as many women as men.
- Treatment depends on the symptoms and their severity.
- Because it is a complex disease, lupus requires treatment by a rheumatologist and the patient’s active participation in working towards good health.

### What is lupus?

Lupus or SLE is a chronic inflammatory disease that can affect the skin, joints, kidneys, lungs, nervous system, and other organs of the body. Usually, patients have skin rashes and arthritis, as well as fatigue and fever. Lupus attacks can vary from mild to severe, and usually alternate between periods of activity and periods when the disease is mostly quiet.

### What causes lupus?

The immune system normally protects the body by producing antibodies that attack foreign germs and cancers. With lupus, the immune system misfires. Instead of producing protective antibodies, the auto-immune disorder begins manufacturing “auto-antibodies” which attack the patient’s own tissues. (Doctors refer to this as a “loss of self-tolerance.”)

As the attack continues, other immune system cells join the fight. This leads to inflammation and blood vessel abnormalities ([vasculitis](#)). These antibodies then end up in the immune system of cells in organs where they cause tissue damage.

Why this inflammatory reaction begins is not known. It is probably the result of a combination of inherited tendencies and environmental factors (such as viruses, the ultraviolet rays in sunlight, Silica dust, and allergies to medications). People affected by lupus may also have an impaired process for clearing old and damaged cells from the body, which then causes an abnormal reaction in the immune system.

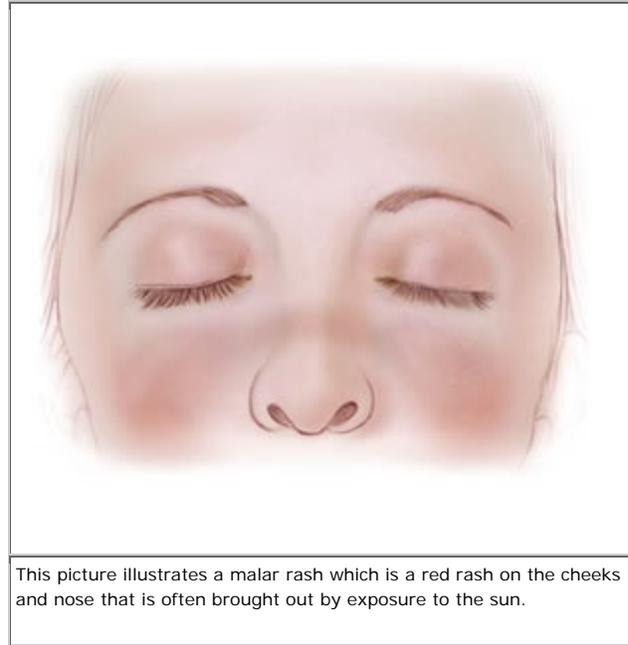
## Who gets lupus?

Lupus typically develops in people in their 20s and 30s, and 10 times more often in women than in men. It is more common in certain ethnic groups, particularly blacks and Asians, who also tend to be more severely affected.

## How is lupus diagnosed?

Diagnosis of lupus is difficult. Suspected cases must be confirmed by a series of blood tests. The most significant test measures [ANA](#), the antinuclear antibody which is present in virtually all lupus patients. Additional, more specific tests, such as the anti-double strand DNA (dsDNA) and anti-smith antibodies (Sm), confirm the diagnosis of lupus. Levels of certain complement proteins (a part of the immune system) in the blood are also measured to help diagnose and track the disease.

The presence of other types of antibodies (anti-phospholipid antibodies) can help doctors diagnosis lupus. These antibodies signal an increased risk of specific complications such as miscarriage and/or blood clots that may lead to stroke or lung injury.



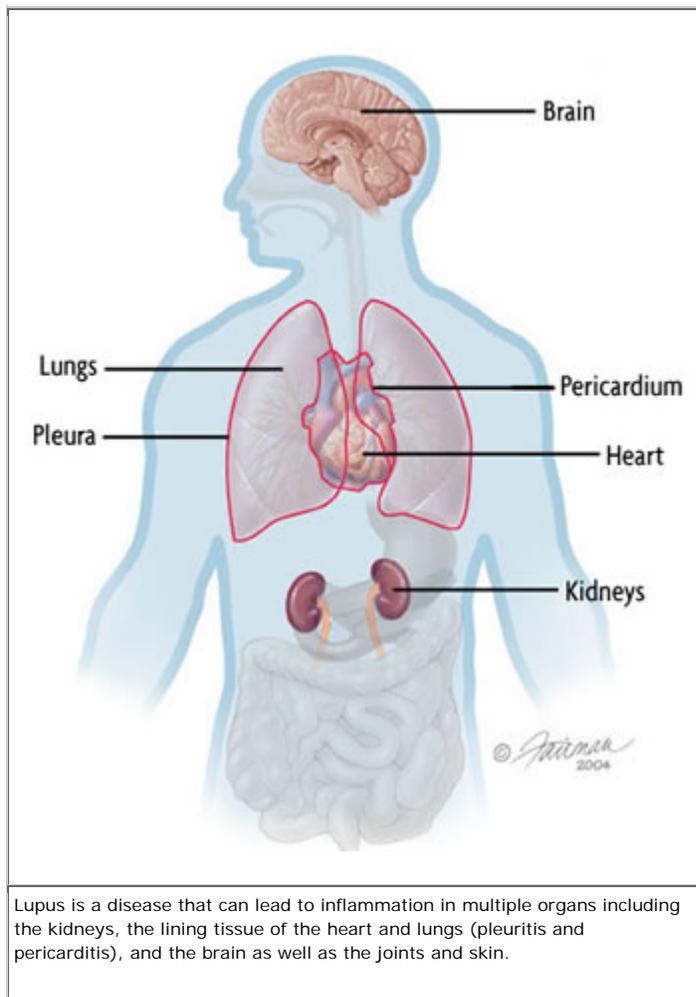
Physicians frequently use the [1997 update of the 1982 Revised Criteria for Classification of Systemic Lupus Erythematosus](#) to aid them in considering whether a patient with symptoms may have lupus. Although this table was originally designed for [clinical research studies](#) it can be helpful when considering an individual patient as well. Please note that most of the findings listed in the table must be documented by a physician. There are many findings in lupus that are not listed in this table. Therefore, a [rheumatologist](#) will use this table only as a starting point.

Patients with lupus typically experience:

- Fever, fatigue, and weight loss
- Arthritis lasting for several weeks in multiple joints
- Butterfly-shaped rash over the cheeks or other rashes
- Skin rash appearing in areas exposed to the sun
- Sores in the mouth or nose lasting for more than a month
- Loss of hair, sometimes in spots or around the hairline
- Seizures, strokes, and mental disorders (depression)
- Blood clots
- Miscarriages
- Blood or protein in the urine or tests that suggest poor kidney function
- Low blood counts (anemia, low white blood cells, or low platelets)

Patients may also have chest pain when breathing deeply, heartburn, abdominal pain, and poor circulation to the fingers and toes.

All of these symptoms can develop gradually, making lupus hard to diagnose. [Rheumatologists](#) specialize in the diagnosis and treatment of autoimmune disorders such as lupus, and will therefore be able to provide patients with the best advice about treatment options.



## How is lupus treated?

There is no cure for lupus and its management can be a challenge. However, in the past several years, treatment has improved considerably and we currently have very medications that are proving effective. Treatment decisions are based on symptoms and the severity of those symptoms.

Patients with muscle or joint pain, fatigue, skin abnormalities (such as rashes), and other responses that are not life-threatening can be treated conservatively. These options include [nonsteroidal anti-inflammatory drugs](#) (NSAIDs) such as ibuprofen (*Motrin, Advil*) and naproxen (*Naprosyn*) and anti-malarial medications such as [hydroxychloroquine](#) (*Plaquenil*). *Remember, some of these NSAIDs can cause serious side effects like stomach bleeding or kidney damage. If you have lupus, always check with your doctor before taking any of the over the counter medications.*

Those with serious and life-threatening problems such as kidney inflammation, lung or heart involvement, and central nervous system symptoms need more aggressive therapy. This may include high dose corticosteroids such as

prednisone (*Deltasone*) and other drugs that suppress the immune system such as [azathioprine](#) (*Imuran*), [cyclophosphamide](#) (*Cytoxan*), and [cyclosporine](#) (*Neoral, Sandimmune*). Recently [mycophenolate](#) (*CellCept*) has been used to treat severe lupus kidney disease. Health care providers may combine several medications to control the disease and prevent tissue damage.

Treatment depends upon an individual assessment of risks and benefits. Most immunosuppressive medications, for instance, may cause significant side effects such as increased risk of infections, nausea, vomiting, hair loss, diarrhea, high blood pressure, and osteoporosis. Rheumatologists may also reduce or discontinue a medication after the disease goes into remission for a period of time. As a result, it is important to undergo careful and frequent medical evaluation to monitor symptoms and adjust treatment as necessary.

[Clinical trials](#) to evaluate new treatments are always being undertaken, with the hope that more promising drugs will be identified and made more widely available. Some of these new treatment options include new drugs already in use for other diseases called [biologics](#). This is an area of significant research.

## Broader health impact of lupus

Even when it is not active, lupus may cause problems later. One of these problems is accelerated atherosclerosis (clogging of the arteries), which increases the risk of heart attacks and other cardiovascular events like heart failure and strokes. Consequently, it is crucial that lupus patients reduce other risk factors such as smoking, high blood pressure, and high cholesterol.

Lupus may also cause kidney disease which can progress to renal failure and require dialysis. This can be prevented by early and aggressive treatment at the first signs of kidney disease.

## Living with lupus

While the treatment for lupus has improved and long-term survival has increased, it is still a chronic disease that can limit activities. Severe sickness usually results from the most serious forms of the disease. More often, quality of life is challenged by symptoms like fatigue and joint pains, which are not life threatening. Further, the unpredictability of lupus and the occasional lack of response to treatment can lead to depression, a loss of hope or the will to keep fighting, or other emotional distress such as irritability or anger.

The best way to control lupus is to:

- Maintain a good doctor-patient relationship and solid support from family and friends. These are key to learning to cope with this chronic and often unpredictable illness.
- Take all medications as prescribed, visit your physician regularly, and learn as much as you can about lupus, your medications, and your progress. Get involved in your care. Don't take a back seat.
- Stay active. This will usually help keep joints flexible and may prevent cardiovascular complications. This does not mean overdoing it; the best approach is to alternate light to moderate exercise with periods of rest or relaxation.
- Avoid excessive sun exposure. Ultraviolet rays in sunlight can cause a skin rash to flare, and may even trigger a more serious flare in the disease itself. Wear protective clothing (long sleeves, a big-brimmed hat) and use sunscreen liberally when outdoors on a sunny day to protect against such complications.

Young women with lupus who wish to have a baby should carefully plan their pregnancies. With physician guidance, pregnancies should be timed for a period when lupus activity is low. Pregnancies must be carefully monitored and certain medications avoided (such as [cyclophosphamide](#), [cyclosporine](#), and [mycophenolate](#)) that would harm the baby. Women whose disease is very active, or who are taking some of these harmful medications, should use birth control.

The possibility that the use of estrogen may cause or worsen lupus has always been a concern for rheumatologists. Recent research has shown that estrogen can trigger some mild or moderate flares of lupus, but does not cause very severe worsening of symptoms. However, because estrogen can increase the risk of blood clots, it should be avoided in patients with lupus whose blood tests have revealed the presence of antiphospholipid antibodies.

## Points to remember

Most people with lupus can live normal lives, but this disorder must be carefully monitored and treatment adjusted as necessary to prevent serious complications.

## The role of a rheumatologist in the treatment of lupus

Lupus is a complex disease. As specialists in the diagnosis and treatment of autoimmune disorders such as lupus, rheumatologists are able to provide patients with the best advice about treatment options.

### To find a rheumatologist

For more information about rheumatologists, [click here](#).

For a listing of rheumatologists in your area, [click here](#).

### For more information

The American College of Rheumatology has compiled this list to give you a starting point for your own additional research. The ACR does not endorse or maintain these Web sites, and is not responsible for any information or claims provided on them. It is always best to talk with your rheumatologist for more information and before making any decisions about your care.

The Arthritis Foundation  
[www.arthritis.org](http://www.arthritis.org)

The Lupus Foundation of America  
[www.lupus.org](http://www.lupus.org)

Lupus Research Institute  
[www.LupusResearchInstitute.org](http://www.LupusResearchInstitute.org)

National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse  
[http://www.niams.nih.gov/Health\\_Info/Lupus/default.asp](http://www.niams.nih.gov/Health_Info/Lupus/default.asp)

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Written by Ellen Ginzler, MD, and Jean Tayar, MD, and reviewed by the American College of Rheumatology Patient Education Task Force.

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