

Systemic lupus erythematosus

Some call it the “great imitator,” because its symptoms vary so widely that it is often mistaken for other disorders. Systemic lupus erythematosus, often referred to simply as lupus, is a serious disease affecting the joints, kidneys, and skin. Although lupus can be a fatal disease, there is much reason for hope. Improvements in therapy have significantly increased life expectancy and quality of life.

Fast Facts

- Lupus affects 10 times as many women as men.
- Treatment depends on the type and severity of symptoms you experience.
- Given its complex nature, lupus requires treatment by a rheumatologist and your own active participation in maintaining your health.

What is lupus?

Systemic lupus erythematosus (also called SLE or lupus) is a chronic inflammatory disease that can affect the skin, joints, kidneys, lungs, nervous system, and/or other organs of the body. The most common symptoms include skin rashes and arthritis, often accompanied by fatigue and fever. The clinical course of SLE varies from mild to severe, and typically involves alternating periods of remission and relapse.

What causes lupus?

SLE is an autoimmune disorder which develops when the body's own immune system, which normally protects against cancers and invading infections, begins to attack the patient's own tissues (known medically as a “loss of self-tolerance”). This occurs first through the production of “auto-antibodies” (antibodies are immune system cells that attack foreign microbes; auto-antibodies attack a person's own cells). As the attack continues, other immune system cells join the fight. This leads to inflammation, blood vessel abnormalities (vasculitis) and deposition of immune system cells in organs which causes tissue damage.

It is not known why this inflammatory reaction begins, but it probably occurs because of some combination of inborn or hereditary predispositions and environmental factors (such as viruses, the ultraviolet rays in sunlight, Silica dust, and allergies to medications). Recent research suggests that people affected by lupus may have a defect in the normal biological process of clearing old and damaged cells from the body, which then causes an abnormal stimulation of the immune system.

Who gets lupus?

SLE occurs 10 times more often in women than in men. The disorder typically develops in people in their twenties and thirties. SLE is more common in certain ethnic groups, particularly in blacks and Asians, who also tend to be more severely affected.

How lupus is diagnosed?

Diagnosis of SLE may be suspected on the basis of symptoms, but is confirmed by a series of blood tests. Of particular interest is the antinuclear antibody (ANA), which is present in virtually all the patients with lupus. Other tests such as the anti-double strand DNA (dsDNA) and anti-smith antibodies (Sm) are more specific and are used to confirm the diagnosis of lupus. The

levels of certain complement proteins (a part of the immune system) are also used to help diagnose and monitor the disease.

If anti-phospholipid antibodies are present, this not only helps to establish a diagnosis of lupus, but also indicate there is an increased risk of specific complications. These include an increased risk of miscarriage and an increased risk for developing blood clots that may lead to stroke or lung injury. Typical clinical features include:

- Fever, fatigue, and weight loss
- Arthritis involving multiple joints for several weeks
- Butterfly-shaped rash over the cheeks or other rashes
- Skin rash appearing in areas exposed to the sun
- Sores in the mouth or nose for more than a month
- Loss of hair, sometimes in spots or around the hairline
- Seizures, strokes and mental disorders
- Blood clots in different locations
- Miscarriages in some patients
- Blood or protein in the urine or tests that suggest poor kidney function
- Low blood counts (anemia, low white blood cells or low platelets)
- Other symptoms include chest pain when the patient breathes deeply, heartburn, abdominal pain and poor circulation to the fingers and toes.

All of these symptoms can develop gradually, making lupus hard to diagnose. (See section below on the role of a rheumatologist.)

How lupus is treated?

Management of SLE can be a challenge. Treatment depends on symptoms and their severity. Careful and frequent medical evaluation is therefore important for monitoring symptoms and adjusting treatment as necessary.

Conservative treatment is appropriate for patients with muscle or joint pain, fatigue, skin manifestations (such as rashes), and other features that are not life-threatening. Conservative options include nonsteroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen (*Motrin*, *Advil*) and naproxen (*Naprosyn*) and anti-malarial medications such as hydroxychloroquine (*Plaquenil*).

More aggressive therapy is required for life-threatening and more serious manifestations such as kidney inflammation, lung or heart involvement, and central nervous system symptoms. Treatment in these circumstances might involve high dose corticosteroids such as prednisone (*Deltasone*) and other immunosuppressive drugs such as azathioprine (*Imuran*), cyclophosphamide (*Cytoxan*), and cyclosporine (*Neoral*, *Sandimmune*). Recently mycophenolate mofetil (*CellCept*) has been used to treat severe lupus kidney disease. Sometimes several medications must be combined 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.

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.

Broader health impact of lupus

Even when it is not active, SLE may cause complications later on. 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. This makes it crucial to try to prevent such complications by reducing other risk factors for heart disease such as smoking, high blood pressure, and high cholesterol. SLE may also cause kidney disease, which can progress to renal failure and require dialysis. This can be prevented by the early and aggressive treatment of the first signs of kidney disease.