

Learning About Lupus

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Lupus Society of Illinois

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Systemic lupus erythematosus (SLE)

Systemic lupus erythematosus (SLE), commonly called lupus, is an immune-system disorder that may affect many organs of the body, including the skin, joints, heart, lungs, kidneys, brain, and/or blood vessels.

The immune system is the body's natural defense against disease. It produces small antibodies (a type of protein) that protect the body against viruses and foreign substances. However, in lupus patients, the antibodies appear to harm the individual's own healthy cells and tissues leading to inflammation and damage in the patient's organs.

The antibodies in lupus patients attach to proteins known as antigens, and together the antibody and antigen complex circulates in the patient's blood, deposits itself on the surface of an internal organ and initiates a process of inflammation.

An example of inflammation is when someone gets a splinter in their finger. The finger gets swollen, tender and red. These are the symptoms of inflammation. When the inflammation occurs internally around the joints or such organs as the heart, lungs and kidneys, the same process and symptoms occur. The common symptoms of lupus are fatigue, painful and swollen joints, morning stiffness, fever over 100°F and skin rashes, particularly on sun-exposed areas of the body such as the face

(see “Symptoms used to diagnose lupus” on page 3). Other symptoms may include chest pain with deep breathing (due to inflammation around the heart or lungs), unusual amounts of hair loss, and white or blue fingertips with exposure to cold (a condition known as Raynaud’s phenomenon).

When lupus attacks the kidneys, it is a painless process and the individual often has no symptoms, although sometimes the patient’s legs may swell. However, a test of the individual’s urine, called a urinalysis, may show abnormal results.

Lupus occurs predominantly in young women by a ratio of 9 to 1 over men. Animal studies have shown that there is a hormonal connection between the symptoms of lupus and the body’s immune system.

In addition, lupus is much more common in women of childbearing age than in premenstrual or post-menopausal women.

Symptoms used to diagnose lupus

The American College of Rheumatology has developed a list of symptoms used by physicians to diagnose lupus. Self-diagnosis is discouraged because many people suspect they have the disease without having the necessary medical evidence. Other conditions, such as rheumatoid arthritis, fibromyalgia and chronic fatigue syndrome, may share similar symptoms. Laboratory tests and tissue biopsies are required to make a specific diagnosis.

The symptoms of lupus are

- ◆ Fever higher than 100°F for an extended period of time
- ◆ Inexplicable fatigue
- ◆ Skin rashes, such as a butterfly-shaped rash over the face, with the wings of the butterfly appearing over the cheeks and the body over the nose
- ◆ Paleness from anemia
- ◆ Swelling of the feet
- ◆ Pain in the chest on deep breathing
- ◆ Sun sensitivity leading to a rash
- ◆ Extensive hair loss
- ◆ Raynaud's phenomenon (fingers turn white or blue when exposed to cold)
- ◆ Neurological problems, including confusion and seizures
- ◆ Ulcers in the mouth or nose for more than a few days at a time

Diagnosing lupus

To make a diagnosis of lupus, physicians need to ask about the individual's symptoms and medical history, conduct a physical examination, and order lab tests.

Both primary care physicians and rheumatologists can diagnose lupus. Specific major organ problems may be treated by other specialists such as nephrologists for kidney involvement, dermatologists for skin involvement and hematologists for blood abnormalities.

The laboratory tests used to diagnose lupus include blood tests for lupus antibodies and other blood elements (see "Antibody tests for lupus patients" on page 6). Lupus patients commonly have low red blood cell counts indicating anemia. They also sometimes have low white blood cell counts, low counts of platelets (small blood particles important for blood clotting), high sedimentation rates and high levels of C-reactive protein (indicators of inflammation).

A urinalysis is helpful in detecting lupus of the kidneys, especially measuring the urine protein level. Measurements of a certain group of proteins in the blood known as complement may also be helpful in the diagnosis, because lupus antibodies consume complement therefore complement levels tend to be low when lupus is active.

Diagnosing lupus

Although studies have shown that lupus tends to run in families and has a genetic basis, the offspring of lupus patients are affected only about 5% of the time. However, family members of lupus patients often test positive for lupus antibodies, but they may not develop any symptoms of the disease.

African-American, Hispanic and Asian women have higher rates of lupus than Caucasian women, presumably due to a genetic susceptibility to the disease. While some of the genes associated with a susceptibility to lupus have been identified, scientists do not know the exact combinations of genes that predispose individuals to the disease.

Lupus tends to occur in flare-periods when the disease is more active. In between the flare-ups, patients may be completely without symptoms. No one knows for sure what triggers the flare-periods. Occasionally, sunlight, medications, stress or pregnancy are the precipitating factors, but more often than not, the specific trigger is not known.

Diagnosing lupus

Antibody tests for lupus patients

Specific antibodies found in lupus patients include antinuclear antibodies (ANAs), anti-DNA antibodies, anti-Sjögren's syndrome A and syndrome B antibodies and anti-Sm (Smith) antibodies.

Virtually all lupus patients test positive for ANAs. However, it is also possible for people who do not have lupus to test positive for ANAs. In addition, most (but not all) lupus patients have test results that show elevated anti-DNA antibodies or low complement proteins.

The ANA test is reported as a quantity called a titer. The test is performed in a double fashion, often starting at a 1:40 titer, meaning one drop of the patient's blood for every 39 drops of saline (saltwater). The test is then performed at 1:80, 1:160, 1:320 titers and so on. The titer quantity reported to a physician is the highest titer that is still positive for ANAs.

Generally speaking, titers of 1:160 and higher are considered more significant than lower titers. Some people may have a significantly positive ANA test without having symptoms of lupus. Most of these people do not develop the disease.

Lupus treatments

Once a diagnosis had been firmly established, patients are assessed for damage to major organs, including the central nervous system, kidneys, heart or lungs. Treatment is tailored to the activity and severity of the disease. Many lupus patients have benign disease without major organ involvement and have a favorable course of treatment.

Treatment given early in the disease process may reduce the patient's chances of suffering permanent organ damage. Patients are often treated with: the antimalarial drug hydroxychloroquine (Plaquenil); nonsteroidal anti-inflammatory drugs (NSAIDs), such as naproxen (Aleve, Naprosyn) and ibuprofen (Advil, Motrin); or the NSAIDs known as COX-2 inhibitors (Celebrex). If a lupus patient is running a fever or has major organ involvement, the treating physician generally prescribes corticosteroids, such as prednisone, sometimes at high doses initially that are then tapered to a small dosage or discontinued. The dosage depends on the type of organ involvement, the patient's symptoms, blood-test results and any side effects caused by the steroid.

Drugs that suppress the immune system may be helpful in controlling the overactive but misdirected immune system in lupus patients. These drugs include azathioprine (Imuran), methotrexate, leflunomide (Arava), cyclophosphamide (Cytoxan), cyclosporine (Neoral), mycophenolate mofetil (CellCept), tacrolimus (Prograf) and rituximab (Rituxan). Belimumab (Benlysta) a B lymphocyte white blood cell inhibitor is given intravenously once a month. Other biologic response medications like Benlysta are currently under investigation for lupus treatment.

Some patients try dietary supplements and herbal medications on their own to help treat the disease, but these therapies have not been studied thoroughly in systemic lupus patients and no one knows what effect, if any, supplements may have.

Physicians, usually specialists such as rheumatologists, monitor the illness continually with laboratory tests that include complete blood counts, urinalysis, blood chemistry measurements, sedimentation rate measurements, antibody tests and blood complement levels. It is important that physicians monitor the patient's symptoms regularly to determine the success of the treatment.

Lupus is not contagious and is not a form of cancer.

One form of lupus is drug-induced. Stopping the incriminating medication may lead to remission. However, most cases of lupus are not related to drug treatment.

Patients may require an examination by such specialists as neurologists and nephrologists to establish treatment options. Patients with advanced cases of lupus may require kidney transplants or total hip replacements, but most patients rarely require hospitalization and many can look forward to leading a relatively normal life.

Lupus and pregnancy

Most women with lupus can become pregnant. However, pregnancy should be avoided during times of active disease or when the disease has affected major organs. Patients with active disease have miscarriages and abnormally small babies more frequently than other women. The miscarriages are sometimes related to the presence of antibodies in the blood called antiphospholipid, anticardiolipin or lupus anticoagulant antibodies. If these antibodies are detected by blood tests, patients are usually treated with low doses of aspirin (81 mg), and sometimes with more potent blood thinners.

The safest, although not always the most effective, method of birth control for women with lupus is a diaphragm used with a contraceptive jelly as an intrauterine device. Many women with lupus take birth control pills, but taking hormones is considered somewhat controversial for lupus patients. Most women with mild forms of the disease do not experience flares when taking oral contraceptives. Some birth control pills and shots do not contain estrogen and may be safer.

More information about lupus is available on the Lupus Society of Illinois website (www.lupusil.org), or if you have any questions, please call the Lupus Society of Illinois at (312) 542-0002.

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The Lupus Society of Illinois (LSI), formed in 1973, is the Midwest's leading non-profit health organization dedicated to finding the causes and cure for lupus.

The LSI's mission is to promote lupus awareness and complement the work of health care professionals by providing personalized resources for the lupus community while supporting research.

The LSI is the only organization in Illinois committed to assisting lupus patients and their families cope with this often devastating disease. Programs and services include:

- Patient education programs and materials
- Referrals to physicians, community services, resources, and assistance
- Health educators /navigators available to answer questions and provide medically sound information about lupus
- Information and updates from local and international lupus medical experts
- Local events and support groups
- Newsletters and alerts
- Living with Lupus grant provides financial assistance for a lupus-related expense to qualifying applicants

LSI's programs and services are accessible via phone, online, and in-person. The Lupus Society of Illinois is a non-profit, 501(c)3 organization. Our programs and services are funded through the generosity of individuals, corporations, foundations and special events.



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