Summary Article: **Lupus (Systemic Lupus Erythematosus)**
From *Harvard Medical School Health Topics A-Z*

**What Is It?**
Lupus is thought to develop when the immune system mistakenly attacks the body's own tissues. The immune system attacks many different parts of the body, including the joints, skin, kidney, nervous system (brain, spinal cord and nerves), blood, heart, lungs, digestive system and eyes, causing inflammation and tissue damage.

Abnormal antibodies develop (called autoantibodies) that can attach themselves to proteins in the body forming abnormal molecules called immune complexes which can trigger additional inflammation and injury when they are deposited in various organs and tissues.

The exact cause of lupus remains a mystery, although scientists are investigating many different possibilities and believe several factors may play a role in the development of the disease. Since 90% of lupus patients are women, usually of childbearing age, researchers think hormones may be involved.

Lupus tends to run in families, so genetic factors may play a role. There is evidence that the illness may be more common in people of African, Native American, West Indian and Chinese descent.

Some researchers think lupus may be triggered by a virus or another type of infection in people who are genetically susceptible to the disease.

Lupus is relatively rare, affecting less than one in 2,000 people. The scientific name of the disease is systemic lupus erythematosus, or SLE.

**Symptoms**
In some people, lupus causes only mild illness, but in others it leads to potentially deadly complications. Symptoms tend to come and go. Periods of intensified symptoms are called flares and periods when symptoms disappear are called remissions. Flares can be triggered by many different factors, including sun exposure, infection, medication and possibly pregnancy, but often they occur for no apparent reason.

Lupus has the potential to affect many different parts of the body, so it can cause a wide range of symptoms including:

- Malaise (a general sick feeling) and fatigue
- Fever
- Loss of appetite
- Weight loss
- Muscle and joint pain, with pain and swelling of the joints
- A butterfly-shaped rash on the cheeks and bridge of the nose, called a malar rash
- Skin photosensitivity (a more widespread rash and flu-like symptoms after exposure to sunlight)

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Hair loss
A "discoid" rash, which appears as firm, round red plaques with raised borders
Painful ulcers in the mouth, nose and genital areas

Other possible symptoms of lupus include:

- Neurological symptoms (headaches, seizures, trouble thinking or stroke)
- Psychiatric symptoms, including psychosis, in which hallucinations may occur
- Heart problems (abnormal heart rhythms, heart failure, inflammation of the heart muscle or lining)
- Lung symptoms, especially pleurisy, which causes painful breathing
- Eye redness or loss of vision
- Pain or swelling in an extremity due to thrombosis (abnormal blood clots)

Some people develop a form of lupus that involves only the skin, called cutaneous lupus or discoid lupus erythematosus. Another form of lupus follows exposure to certain drugs (drug-induced lupus) including procainamide and hydralazine. While drug-induced lupus may cause rash, arthritis and fever that appear similar to the systemic form of lupus, it tends to be milder.

Women with lupus can have normal pregnancies and deliver healthy babies. However, pregnant women with lupus are at risk for certain complications such as high blood pressure or worsening kidney function. If the lupus is "quiet" prior to becoming pregnant, these risks are reduced.

Women with lupus tend to have an increased risk of having small birthweight babies. Babies born to women with lupus may have a rash, low blood counts and heart problems, a condition called "neonatal lupus."

Women with lupus who are planning pregnancy should see an obstetrician experienced in such pregnancies and a rheumatologist. This is particularly important so that medications that may be risky for the fetus can be adjusted or stopped and the lupus can be closely monitored. If the lupus is active, a woman may be advised to delay pregnancy.

**Diagnosis**

Your doctor will begin by reviewing your symptoms, your medical history and your exposure to factors that can trigger lupus flares. Next, he or she will examine you, looking for skin rashes on your face or on sun-exposed skin, tenderness or swelling of the joints and ulcers inside your mouth or nose. Your doctor will listen to your heart and lungs with a stethoscope, checking for signs of inflammation of the membrane covering the heart (pericarditis) or inflammation of the membranes covering the lungs (pleuritis).

If your doctor suspects you have lupus, he or she will order a blood test to look for a type of antibody, called the antinuclear antibody (ANA), that almost all people with lupus have in their blood. However, since the ANA test can be positive in people who do not have lupus, your doctor may order follow-up blood tests to look for other types of antibodies. Lupus cannot be diagnosed only on the basis of the ANA test.
Your doctor may evaluate your condition using the criteria established by the American College of Rheumatology. Your doctor may diagnose lupus even if you don't meet all of these criteria, which were developed for research studies. If you have had 4 of the 17 lupus criteria at some time during your illness, even if fewer than four are active at the time of diagnosis, the diagnosis is more certain and you may be eligible for entry into a research study of lupus.

At least one of the positive criteria must be "clinical" (causing symptoms or affecting a particular part of the body) and one must be a laboratory abnormality (such as an abnormal blood test). The diagnosis can be established without meeting 4 criteria if a kidney biopsy shows evidence of lupus kidney disease along with certain antibodies (including antinuclear antibodies or anti-ds-DNA) present in the blood.

The lupus criteria include the following:

- Certain types of rashes (called acute or chronic cutaneous lupus)
- Discoid rash
- Hair loss
- Ulcers in the mouth or nose
- Arthritis
- Pericarditis, confirmed by physical exam or electrocardiogram (EKG), or pleuritis, confirmed by physical findings or chest X-ray
- Kidney disorder, confirmed by finding high levels of protein in the urine or other specific urine abnormalities, especially red cells suggesting inflammation in the kidney
- Neurological disorder, including seizures or psychosis (a serious psychiatric illness)
- Blood disorder, including evidence of red blood cell destruction (hemolytic anemia), low white blood cells (leukopenia) or low platelets (thrombocytopenia)
- Immune disorder — This is established by the finding of certain antibodies in the blood, which may include a positive ANA, anti-ds-DNA test, or anti-Smith antibody test, a positive test for syphilis even though you don't have syphilis or a positive antiphospholipid antibody test (an antibody associated with miscarriage or blood clots).
- Low complement levels (proteins involved in inflammation)
- Antibodies linked with red blood cell destruction, called a positive Coombs' test

Other tests that may be performed to help diagnose lupus include:

- Erythrocyte sedimentation rate (ESR), a blood test that indicates the presence of inflammation
- A skin or kidney biopsy (taking a small tissue sample for laboratory examination)
- Additional blood tests for autoantibodies

**Expected Duration**

Lupus is a long-lasting (chronic) condition, although its activity often varies over time with periods of active disease (flares) and periods in which the illness is relatively inactive or even completely quiet.
Prevention
Since doctors haven't determined the cause of lupus, there's no way to prevent it. You may be able to prevent flare-ups of the illness by taking medications as prescribed, avoiding exposure to the sun as much as possible and using sunscreen when you are in the sun.

Treatment
Lupus may be treated with several different types of medication, including:

- Nonsteroidal anti-inflammatory drugs (NSAIDS), such as ibuprofen (Advil, Motrin and other brand names) or naproxen (Aleve, Naprosyn and others)
- Antimalarials, such as hydroxychloroquine (Plaquenil), chloroquine (Aralen), or quinacrine. Recent studies suggest that lupus patients treated with antimalarial medications have less active disease and less organ damage over time. Therefore, many experts now recommend antimalarial treatment for all patients with systemic lupus unless they cannot tolerate the medication.
- Corticosteroids, such as prednisone (Deltasone and others), hydrocortisone, methylprednisolone (Medrol and others), or dexamethasone (Decadron and others)
- Immunosuppressives, such as azathioprine (Imuran), cyclophosphamide (Cytoxan, Neosar), methotrexate (Rheumatrex, Folex, Methotrexate LPF), or mycophenolate mofetil (CellCept), or belimumab (Benlysta)

When To Call a Professional
Call your doctor if you have any of the symptoms of lupus, especially if you develop skin symptoms (malar or discoid rash, photosensitivity, ulcers in your mouth or nose), together with fatigue, fever, joint pain, poor appetite and weight loss.

Prognosis
Most people with lupus have a normal life span. However, life expectancy and quality of life vary widely depending on severity of illness. Cardiovascular disease, including heart attack, is more common among people with lupus. The presence of cardiovascular disease worsens prognosis. The outlook is also worse if the disease has seriously affected the kidneys or brain.

Additional Info
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1 AMS Circle
Bethesda, MD 20892-3675
Phone: 301-495-4484
Toll-Free: 1-877-226-4267

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