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Systemic lupus erythematosus

Systemic lupus erythematosus (SLE) is an autoimmune disease. In this disease, the immune system of the body mistakenly attacks healthy tissue. It can affect the skin, joints, kidneys, brain, and other organs.

Causes

The cause of SLE is not clearly known. It may be linked to the following factors:

- Genetic
- Environmental
- Hormonal
- Certain medicines

SLE is more common in women than men. It may occur at any age. However, it appears most often in people between the ages of 15 and 44. The disease affects African Americans and Asians more than people from other races.

Symptoms

Symptoms vary from person to person, and may come and go. Everyone with SLE has joint pain and swelling at some time. Some develop arthritis. SLE often affects the joints of the fingers, hands, wrists, and knees.

Other common symptoms include:

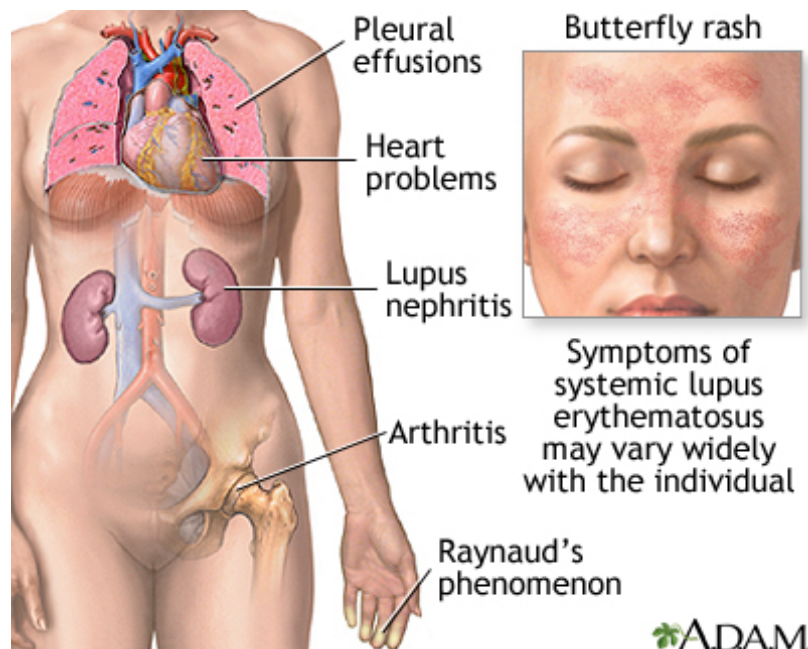
- Chest pain when taking a deep breath.
- Fatigue.
- Fever with no other cause.
- General discomfort, uneasiness, or ill feeling (malaise).

- Hair loss.
- Weight loss.
- Mouth sores.
- Sensitivity to sunlight.
- Skin rash: A "butterfly" rash develops in about half the people with SLE. The rash is mostly seen over the cheeks and bridge of the nose. It can be widespread. It gets worse in sunlight.
- Swollen lymph nodes.

Other symptoms depend on which part of the body is affected:

- Brain and nervous system: Headaches, numbness, tingling, seizures, vision problems, and personality changes
- Digestive tract: Abdominal pain, nausea, and vomiting
- Heart: Valve problems, inflammation of heart muscle
- Lung: Buildup of fluid in the pleural space, difficulty breathing
- Skin: Patchy skin color and fingers that change color when cold (Raynaud phenomenon)
- Kidney: Swelling in the legs

Some people have only skin symptoms. This is called discoid lupus.



Exams and Tests

To be diagnosed with lupus, you must have 4 out of 11 common signs of the disease. Nearly all people with lupus have a positive test for antinuclear antibody (ANA). However, having a positive ANA alone does not mean you have lupus.

The health care provider will do a complete physical exam. You may have a rash, arthritis, or edema in the ankles. There may be an abnormal sound called a heart friction rub or pleural friction rub. Your provider will also do a nervous system exam.

Tests used to diagnose SLE may include:

- Antinuclear antibody (ANA)
- CBC with differential
- Chest x-ray
- Serum creatinine
- Urinalysis

You may also have other tests to learn more about your condition. Some of these are:

- Antinuclear antibody (ANA) panel
- Complement components (C3 and C4)
- Antibodies to double-stranded DNA
- Coombs test -- direct
- Cryoglobulins
- ESR and CRP
- Kidney function blood tests
- Liver function blood tests
- Rheumatoid factor
- Antiphospholipid antibodies and lupus anticoagulant test
- Kidney biopsy
- Imaging tests of the heart, brain, lungs, joints, muscles or intestines

Treatment

There is no cure for SLE. The goal of treatment is to control symptoms. Severe symptoms that involve the heart, lungs, kidneys, and other organs often need treatment from specialists. Each person with SLE needs evaluation regarding:

- How active the disease is
- What part of the body is affected
- What form of treatment is needed

Mild forms of the disease may be treated with:

- NSAIDs for joint symptoms and pleurisy. Talk to your provider before taking these medicines.
- Low doses of corticosteroids, such as prednisone, for skin and arthritis symptoms.
- Corticosteroid creams for skin rashes.
- Hydroxychloroquine, a medicine also used to treat malaria.
- Belimumab, a biologic medicine, may be helpful in some people.

Treatments for more severe SLE may include:

- High-dose corticosteroids.
- Immunosuppressive medicines (these medicines suppress the immune system). These medicines are used if you do not get better with corticosteroids, or if your symptoms get worse when you stop taking them.
- Most commonly used medicines include mycophenolate, azathioprine and cyclophosphamide. Because of its toxicity, cyclophosphamide is limited to a short course of 3 to 6 months. Rituximab (Rituxan) is used in some cases as well.
- Blood thinners, such as Coumadin, for clotting disorders.

If you have SLE, it is also important to:

- Wear protective clothing, sunglasses, and sunscreen when in the sun.
- Get preventive heart care.
- Stay up-to-date with immunizations.
- Have tests to screen for thinning of the bones (osteoporosis).
- Avoid tobacco and drink minimal amounts of alcohol.

Support Groups

Counseling and support groups may help with the emotional issues involved with the disease.

Outlook (Prognosis)

The outcome for people with SLE has improved in recent years. Many people with SLE have mild symptoms. How well you do depends on how severe the disease is. Most people with SLE will require medicines for a long time. Nearly all will require hydroxychloroquine indefinitely.

The disease tends to be more active:

- During the first years after diagnosis

- In people under age 40

Many women with SLE can get pregnant and deliver a healthy baby. A good outcome is more likely for women who receive proper treatment and do not have serious heart or kidney problems. However, the presence of certain SLE antibodies or antiphospholipid antibodies raises the risk of miscarriage.

Possible Complications

LUPUS NEPHRITIS

Some people with SLE have abnormal immune deposits in the kidney cells. This leads to a condition called lupus nephritis. People with this problem may develop kidney failure. They may need dialysis or a kidney transplant.

A kidney biopsy is done to detect the extent of damage to the kidney and to help guide treatment. If active nephritis is present, treatment with immunosuppressive medicines including high doses of corticosteroids along with either cyclophosphamide or mycophenolate are needed.

OTHER PARTS OF THE BODY

SLE can cause damage in many different parts of the body, including:

- Blood clots in arteries or veins of the legs, lungs, brain, or intestines
- Destruction of red blood cells or anemia of long-term (chronic) disease
- Fluid around the heart (pericarditis), or inflammation of the heart (myocarditis or endocarditis)
- Fluid around the lungs and damage to lung tissue
- Pregnancy problems, including miscarriage
- Stroke
- Bowel damage with abdominal pain and obstruction
- Inflammation in the intestines
- Severely low blood platelet count (platelets are needed to stop any bleeding)
- Inflammation of the blood vessels

SLE AND PREGNANCY

Both SLE and some of the medicines used for SLE can harm an unborn child. Talk to your provider before you become pregnant. If you become pregnant, find a provider who is experienced with lupus and pregnancy.

When to Contact a Medical Professional

Call your provider if you have symptoms of SLE. Also call if you have this disease and your symptoms get worse or a new symptom occurs.

Alternative Names

Disseminated lupus erythematosus; SLE; Lupus; Lupus erythematosus; Butterfly rash – SLE; Discoid lupus

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Updated by: Gordon A. Starkebaum, MD, Professor of Medicine, Division of Rheumatology, University of Washington School of Medicine, Seattle, WA. Internal review and update on 03/28/19 by David Zieve, MD, MHA, Medical Director, Brenda Conaway, Editorial Director, and the A.D.A.M. Editorial team.

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