

**SYSTEMIC LUPUS ERYTHEMATOSUS**

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ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that has protean manifestations and follows a relapsing and remitting course. It is characterized by an autoantibody response to nuclear and cytoplasmic antigens. SLE can affect any organ system, but mainly involves the skin, joints, kidneys, blood cells, and nervous system (see Clinica I). The diagnosis of SLE must be based on the proper constellation of clinical findings and laboratory evidence. American College of Rheumatology (ACR) criteria summarizes features necessary for diagnosis. (See Workup.) Management depends on disease severity and organ involvement. Periodic follow-up and laboratory testing are imperative to detect signs and symptoms of new organ-system involvement and to monitor the response or adverse reactions to therapies. Systemic lupus erythematosus (SLE) is a long-term autoimmune disorder that may affect the skin, joints, kidneys, brain, and other organs.

Key words: Systemic lupus erythematosus, nuclear and cytoplasmic antigens

INTRODUCTION

The term lupus (Latin for wolf) is attributed to the thirteenth century physician Rogerius who used it to describe erosive facial lesions that were reminiscent of a wolf's bite.¹⁻³ Classical descriptions of the various dermatologic features of lupus were made by Thomas Bateman, a student of the British dermatologist Robert William, in the early nineteenth century; Cazenave, a student of the French dermatologist Laurent Bielt, in the mid-nineteenth century; and Moriz Kaposi (born Moriz Kohn), student and son-in-law of the Austrian dermatologist Ferdinand von Hebra, in the late nineteenth century. The lesions now referred to as discoid lupus were described in 1833 by Cazenave under the term "erythema centrifugum," while the butterfly distribution of the facial rash was noted by von Hebra in 1846. The first published illustrations of lupus erythematosus were included

in von Hebra's text, Atlas of Skin Diseases, published in 1856.

Systemic lupus erythematosus (SLE) is a chronic, often life-long, autoimmune disease. It can be mild to severe, and affects mostly women. SLE may affect various parts of the body, but it most often manifests in the skin, joints, blood, and kidneys. SLE was first described in 1828. Its very name helps define the disease:

- Systemic is used because the disease can affect organs and tissue throughout the body.
- Lupus is Latin for wolf. It refers to the rash that extends across the bridge of the nose and upper cheekbones and was thought to resemble a wolf bite.
- Erythematosus is from the Greek word for red and refers to the color of the rash.

WHO classification of Lupus Nephritis:

Pattern	Immune fluorescence		Electron microscopy		
	Mesangial	Peripheral	Mesangial	Sub endothelial	Sub epithelial
I normal	0	0	0	0	0
II Mesangial deposit	+	0	+	0	0
III Mesangial hypercellularity	+	0	+	0	0
IV focal segmental GN	++	+	++	+	+
V diffuse GN	++	++	++	++	+
V membranous GN		++	+	+	++

Causes, incidence, and risk factors

Systemic lupus erythematosus (SLE) is an autoimmune disease, which means the body's immune system mistakenly attacks healthy tissue. This leads to long-term (chronic) inflammation. The underlying cause of autoimmune diseases is not fully known. SLE is much more common in women than men. It may occur at any age, but appears most often in people between the ages of 10 and 50. African Americans and Asians are affected more often than people from other races. SLE may also be caused by certain drugs. For information on this cause, see Drug-induced lupus erythematosus

Symptoms:

Symptoms vary from person to person, and may come and go. Almost everyone with SLE has joint pain and swelling.

Some develop arthritis. Frequently affected joints are 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, Mouth sores, Sensitivity to sunlight, Skin rash -- a "butterfly" rash over the cheeks and bridge of the nose affects about half of people with SLE. The rash gets worse in sunlight. The rash may also be widespread, Swollen lymph nodes

Other symptoms depend on what part of the body is affected: Brain and nervous system: headaches, numbness, tingling, seizures, vision problems, personality changes, Digestive tract: abdominal pain, nausea, and vomiting, Heart: abnormal heart rhythms (arrhythmias), Lung: coughing up blood and difficulty breathing, Skin: patchy skin color, fingers that

change color when cold (Raynaud's phenomenon). Some patients only have skin symptoms. This is called discoid lupus.

Signs and tests:

To be diagnosed with lupus, you must have 4 out of 11 typical signs of the disease. Your doctor will perform a physical exam and listen to your chest with a stethoscope. An abnormal sound called a heart friction rub or pleural friction rub may be heard. A nervous system exam will also be done.

Tests used to diagnose SLE may include: Antibody tests, including antinuclear antibody (ANA) panel, CBC, Chest x-ray, Kidney biopsy, Urinalysis.

This disease may also alter the results of the following tests: Antithyroglobulin antibody, Antithyroid microsomal antibody, Complement components (C3 and C4), Coombs' test - direct, Cryoglobulins, ESR, Kidney function blood tests, Liver function blood tests, Rheumatoid factor.

Treatment:

There is no cure for SLE. The goal of treatment is to control symptoms.

Mild disease may be treated with: Nonsteroidal anti-inflammatory medications (NSAIDs) treat arthritis and pleurisy, Corticosteroid creams to treat skin rashes, An antimalarial drug (hydroxychloroquine) and low-dose corticosteroids for skin and arthritis symptoms.

Use of corticosteroid to treat various lupus manifestations:

Clinical feature initial	Initial Dose of prednisolone
Arthritis (poorly responding to NSAIDs)	20-30mg/d, reducing by about 5mg/wk if symptoms abate
Haemolytic anemia	1mg/kg/d for about 1M
Thrombocytopenia	reduce by 10mg/d if blood tests improve
Neuropsychiatric	controversial! 1-2mg/kg/d
Nephritis	0.5-1g/d

You should wear protective clothing, sunglasses, and sunscreen when in the sun. Severe or life-threatening symptoms (such as hemolytic anemia, extensive heart or lung involvement, kidney disease, or central nervous system involvement) often require more aggressive treatment by doctor specialists.

Treatment for more severe lupus may include:

- High-dose corticosteroids or medications to decrease the immune system response
- Cytotoxic drugs (drugs that block cell growth) if you do not get better with corticosteroids, or whose symptoms get worse when the stop taking them. These medicine have serious, severe side effects. You should be closely monitored by your doctor.

If you have lupus, it is also important to have: Preventive heart care, Up-to-date immunizations, Tests to screen for thinning of the bones (osteoporosis). Talk therapy and support groups may help relieve depression and mood changes that may occur in patients with this disease.

Expectations (prognosis):

How well a person does depends on the severity of the disease. The outcome for people with SLE has improved in recent years. Many people with SLE have mild symptoms. Women with SLE who become pregnant are often able to carry safely to term and deliver a healthy infant, as long as they do not have severe kidney or heart disease and the SLE is being treated appropriately. However, the presence of SLE antibodies may increase the risk of pregnancy loss.

Complications:

Some people with SLE have abnormal deposits in the kidney cells. This leads to a condition called lupus nephritis. Patients with this condition may eventually develop kidney failure and need dialysis or a kidney transplant. SLE causes damage to many different parts of the body, including: Blood clots in the legs (deep vein thrombosis) or lungs (pulmonary embolism), Destruction of red blood cells (hemolytic anemia) or anemia of chronic disease, Fluid around the heart (pericarditis), endocarditis, or inflammation of the heart (myocarditis), Fluid around the lungs (pleural effusions) and damage to lung tissue, Pregnancy complications, including miscarriage, Stroke, Severely low blood platelets (thrombocytopenia), Inflammation of the blood vessels^{4,21}.

Calling your health care provider:

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


CONCLUSION:

An appreciation of the many facets of SLE is essential, including recognition of the current limit of our knowledge about the disease and its management. A better understanding of the pathogenesis of SLE promises to provide much information about the nature and the role of the immune response in this and other diseases.

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