

# Systemic Lupus Erythematosus

## What is Systemic Lupus Erythematosus?

Systemic Lupus Erythematosus (SLE) is a chronic disorder of the immune system, which normally functions to protect against foreign invaders, becomes hyperactive, forming antibodies that attack normal tissues and organs, causing chronic inflammation and damage. It can affect virtually any organ of the body. The most common organs affected include joints, skin and kidneys. Lupus is characterized by periods of illness, called flares, and periods of wellness, or remission.

There are two other types of lupus:

1. Discoid lupus erythematosus, which mainly affects the skin. The symptoms of this form of lupus include a rash on the face, scalp, or elsewhere. The rash may last for days or years, and can recur.
2. Drug-induced lupus, which is triggered by certain medications, e.g. procainamide, hydralazine, phenytoin, etc. Symptoms usually disappear once the drugs are discontinued. The symptoms in this form of lupus may be milder.

## What Causes Systemic Lupus Erythematosus?

To date, there is no known cause or cure. Etiology is likely multifactorial:

1. Genetic predisposition: those individuals with HLA-DR2 or HLA -DR3 genes and complement deficiency especially C1q or C4 deficiencies have higher risk of developing SLE. Those with positive family history of SLE also have a higher chance of disease development.
2. Environmental factors: ultraviolet light, stress, emotion, surgery, trauma, tobacco, certain chemicals could induce or exacerbate lupus.
3. Oral contraceptive pills: to date, evidence still suggests avoidance of estrogen containing pills in lupus patient, and believes it could worsen disease condition.

## Who Gets Systemic Lupus Erythematosus?

Ninety percent of lupus victims are women, and the onset of the disease usually occurs at child bearing age. Men, elderly and children can rarely be affected. Lupus is also more common among Latino, Asian, and Native American women.

## What are the Symptoms of Systemic Lupus Erythematosus?

Lupus is unpredictable and highly individualized. Most people with lupus have mild symptoms in only a few organs; others may face serious, sometimes life-threatening problems.

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Common symptoms include:

1. Butterfly-shaped photosensitive rash across cheeks and nose
2. Arthralgia, especially over small hand joints
3. Recurrent oral or nasal ulcers (often painless)
4. Unusual hair loss, anaemia, low white cell count or low platelet count
5. In severe case, brain, central nervous system, kidneys and heart could be affected

## The Diagnosis of Systemic Lupus Erythematosus

With reference to the 1998 revised criteria by the American College of Rheumatology, if any 4 of the following 11 symptoms or specific laboratory findings occur consecutively or simultaneously, one individual is likely to suffer from lupus.

1. Malar rash: butterfly-shaped rash across cheeks and nose
2. Discoid skin rash: raised red scaly, disk-shaped sores on the face, neck and/or chest
3. Photosensitivity: skin rash as a result of unusual reaction to sunlight
4. Recurrent mouth or nose ulcers: usually painless
5. Non-erosive arthritis: in 2 or more joints with tenderness, swelling, or effusion
6. Cardio-pulmonary involvement: inflammation of the lining around the heart (pericarditis) and/or lungs (pleuritis)
7. Neurological disorder: seizures and/or psychosis/cognitive dysfunction
8. Renal (kidney) disorder: excessive protein in the urine, or cellular casts in the urine, or biopsy proven lupus nephritis
9. Hematological (blood) disorder: hemolytic anemia ( $< 10$  g/dl), low white blood cell count ( $< 4.0 \times 10^9/L$ ), or low platelet count ( $< 100 \times 10^9/L$ )
10. Immunological disorder: antibodies to double stranded DNA or antibodies to anticardiolipin antibody
11. Antinuclear antibodies (ANA): positive test in absence of drugs known to induce it

## Medication

1. NSAIDS, nonsteroidal anti-inflammatory drugs, are used to relieve achy joints and arthritis in mild lupus when pain is limited and organs are not affected. The newer generation of NSAID, COX-II specific NSAID, could reduce the side effect on gastrointestinal tract; however, its adverse effect on cardiovascular system, limit its use in selected individuals.
2. Antimalarial drugs, such as hydroxychloroquine, are often prescribed for arthritis or skin problems. It could also help to stabilize the disease and keep it “quiet”.

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3. Corticosteroids, such as prednisolone, are used for major organ involvement. The dosage prescribed will depend on the type of organ involvement, symptoms, and blood-test results. When the disease is under control, physicians will tail down and even tail off the drug to reduce its long term side effects. In long term user, steroid could not be stopped abruptly or adrenal insufficiency could occur.
4. Immunosuppressive agents, such as azathioprine (Imuran), methotrexate, cyclophosphamide, cyclosporine, and mycophenolate mofetil (CellCept), are potent drugs that help control the overactive but misdirected immune system in lupus patients. They help limit damage to major organs and are closely monitored to counter the potentially serious side effects and complications.

## **Treatment**

While there is no cure for lupus, there are treatments. Early diagnosis and appropriate treatment can help manage the symptoms of lupus and lessen the chance of permanent damage to organs or tissue. Drug compliance and communication with physicians is of utmost importance in the treatment of lupus.

Once a lupus diagnosis is established, patients are assessed for damage to major organs (central nervous system, kidneys, heart, or lungs). Treatment depends on the activity and extent of the disease, and can range from over the counter pain relievers and anti-inflammatories to prescription medication, therapy, dietary changes, and lifestyle changes such as staying out of the sun, wearing sun block, and avoiding stress.

## **What is the outlook?**

While there is not yet a cure, research uncovers promising new findings each year. Over the last two decades, better diagnostic techniques and treatment methods have led to more effective management of lupus and its complications. Now with earlier diagnosis, the latest therapies and careful monitoring, the 10 year survival rate is more than 90% and most people with lupus can look forward to a normal lifespan. Majority of patients could enjoy normal life style and work.

## **Suggestions**

1. During a flare, get plenty of rest. Comply with treatment.
2. When in remission: exercise to increase joint flexibility and muscle strength.
3. Use sunscreen and sun-protective clothing, and avoid the sun.
4. For fever over 38 degree Celsius: seek medical advice.

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5. Attend regular follow-ups: to monitor disease activity; these usually include blood and urine tests.
6. Ask questions when in doubt.
7. Report any side effects or new symptoms promptly: help your doctor know when a change in therapy might be needed.
8. Safe contraceptive method: condom or sterilization is the best to you, better to avoid intra-uterine device which could increase risk of infection.
9. Relieve stress: support groups, counseling, talking with friends, family, and doctors can be helpful.