

# SLE · Systemic Lupus Erythematosus

A plain-English guide for you and your family

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## WHAT IS HAPPENING

Normally, your immune system has a built-in safety switch called tolerance that stops it from attacking your own cells. Special checkpoint cells in the thymus (a small organ behind your breastbone) and bone marrow screen out any immune cells that might recognize 'you' as an enemy. At the same time, chemicals called type I interferons (messengers that usually fight viruses) stay quiet unless a real germ shows up. When lupus begins, that safety switch fails. Your body starts making autoantibodies — immune proteins that grab onto your own DNA, cell surfaces, and other normal parts of you. These antibodies glue together into clumps called immune complexes that lodge in small blood vessels, especially in your skin, kidneys, joints, and the thin sacs around your heart and lungs. The clumps call in swarms of white cells that cause swelling and scarring. At the same time, your cells pour out too much type I interferon, which keeps the whole immune system revved up and angry. Triggers like sunlight (UV rays), the Epstein-Barr virus (the germ that causes mono), cigarette smoke, and hormones (especially estrogen) can flip the switch in someone whose genes already loaded the risk. Once it starts, lupus moves from organ to organ over months and years. You might see a butterfly rash across the cheeks, joint pain without bone damage, protein spilling into the urine as the kidney filters break, chest pain from inflamed heart or lung linings, dropping blood counts, seizures or confusion, and dangerous clots if antiphospholipid antibodies appear. Each flare-up is the immune attack landing somewhere new. The body tries to heal the damage, but the antibodies keep coming, so scar tissue slowly builds up. None of this is random — every symptom traces back to that one lost brake on self-recognition.

## What you might feel

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- Malar rash (butterfly rash across the nose and cheeks)
- Joint pain and swelling (arthritis), especially in hands, wrists, and knees
- Extreme tiredness (fatigue) that does not go away with rest
- Photosensitivity (rashes or feeling sick after sun exposure)
- Fever without infection
- Mouth or nose sores (usually painless)

- Kidney problems (proteinuria, blood in urine, or rising creatinine) with little or no pain
- Seizures, headache, confusion, or stroke-like symptoms
- Chest pain that gets worse with deep breathing (pleuritis or pericarditis)
- Low blood counts (anemia, low white cells, or low platelets)

## **Call the doctor right away if...**

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- New seizure, severe headache, confusion, or sudden weakness on one side of the body (possible brain lupus or stroke)
- Sudden shortness of breath, chest pain, or coughing up blood (possible blood clot in the lung, heart inflammation, or lung bleeding)
- Swelling of the legs and face with foamy urine or much less urine than usual (kidney failure may be starting)
- Fever over 100.4°F (38°C) — could be infection (lupus suppresses immunity and treatments do too) or a severe lupus flare
- Severe abdominal pain, especially with nausea or vomiting (possible inflammation of blood vessels in the gut, which can cause bleeding or tissue death)

## **What the care team will do**

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- Check urine dipstick for protein and blood, and monitor daily weights
- Assess for signs of infection before assuming symptoms are a lupus flare
- Monitor blood pressure every 4 hours and report systolic over 140 or diastolic over 90
- Check complete blood count daily for low white cells, platelets, or hemoglobin
- Ensure hydroxychloroquine is given daily and teach the patient it prevents flares
- Teach sun protection with SPF 50 sunscreen, hats, and long sleeves every day
- Monitor for signs of serositis including sharp chest pain, friction rub on lung sounds, or muffled heart sounds
- Screen for depression and teach that brain fog and mood changes are part of lupus

## **Medicines you may be given — and why**

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### **Hydroxychloroquine**

Hydroxychloroquine calms the overactive immune system that attacks the body's own tissues in lupus. It works by changing the pH (acid level) inside certain immune cells, which slows down the cells that present 'targets' to attack and quiets the release of inflammatory signals.

Think of it like turning down the volume on an alarm system that's going off too much. This medicine reduces flares, protects organs, and helps people with lupus live longer.

### **Prednisone (or other corticosteroids)**

Prednisone is a powerful brake on inflammation. It mimics cortisol, a hormone the adrenal glands make to shut down the immune system's attack response. Prednisone enters cells and turns on genes that reduce inflammation and turns off genes that make inflammatory chemicals. It is used for flares or when organs are actively inflamed, but because long-term use causes serious side effects, doctors aim to use the lowest dose for the shortest time.

### **Mycophenolate mofetil (or mycophenolic acid)**

Mycophenolate slows down the multiplication of B cells and T cells (the immune cells that make antibodies and attack tissues). It does this by blocking a key enzyme those cells need to make DNA building blocks. Because B and T cells rely heavily on this pathway, mycophenolate selectively suppresses the overactive immune response in lupus without shutting down the whole immune system as much as steroids do. It is a mainstay for lupus kidney inflammation (lupus nephritis) and is used for maintenance therapy to keep the disease quiet.

### **Cyclophosphamide**

Cyclophosphamide is a powerful medicine that kills rapidly dividing cells, especially B cells and T cells that are multiplying and attacking the body. It works by damaging the DNA inside these cells so they cannot divide and eventually die off. This resets the immune system and is used for the most severe lupus, particularly life-threatening kidney inflammation (lupus nephritis) or brain/lung involvement. Because it is so strong and has serious side effects, it is used for induction (the first few months to get severe disease under control), then switched to a gentler medicine like mycophenolate for maintenance.

### **Azathioprine**

Azathioprine slows down the multiplication of B cells and T cells by interfering with the building blocks they need to copy their DNA. It is converted in the body into a fake DNA building block that gets incorporated into new DNA strands, causing errors and stopping cell division. It is used as maintenance therapy to keep lupus quiet after initial treatment, particularly for kidney disease, and for people who cannot tolerate mycophenolate.

### **Voclosporin**

Voclosporin blocks the activation of T cells, the immune cells that orchestrate the attack on kidneys and other organs in lupus. It works by inhibiting calcineurin, an enzyme inside T cells that turns on genes needed for the cells to spring into action and release inflammatory signals. By keeping T cells quiet, voclosporin reduces kidney inflammation. It is approved specifically

as an add-on to mycophenolate and low-dose steroids for active lupus nephritis (kidney inflammation from lupus).

### **Belimumab**

Belimumab targets B cells, the immune cells that make autoantibodies (self-attacking antibodies) in lupus. It works by blocking a protein called BLYS (B-lymphocyte stimulator), which B cells need to survive and mature. Without BLYS, excess B cells die off, reducing the number of cells making harmful antibodies. Belimumab is used for active lupus that is not controlled with standard medicines, and it is approved as an add-on for lupus nephritis (kidney inflammation).

### **Rituximab**

Rituximab wipes out B cells, the immune cells that make autoantibodies in lupus. It works by binding to a marker called CD20 on the surface of B cells, tagging them for destruction by the immune system and causing them to die. This resets the B-cell population, reducing harmful antibody production. Rituximab is used off-label (not FDA-approved for lupus, but widely used) for severe or refractory (stubborn) lupus, especially kidney disease that does not respond to standard treatment.

### **Anifrolumab**

Anifrolumab blocks interferon, a powerful inflammatory signal that is overactive in many people with lupus. Interferons are proteins that normally fight viruses, but in lupus, the immune system mistakenly makes too much interferon, which drives inflammation in skin, joints, and organs. Anifrolumab binds to the interferon receptor on cells, preventing interferon from delivering its inflammatory message. It is used for moderate to severe lupus that is not controlled with standard treatment.

### **Methotrexate**

Methotrexate calms inflammation in the joints and skin by interfering with folate (a B vitamin) metabolism inside immune cells. It blocks an enzyme called dihydrofolate reductase, which cells need to make DNA and proteins. At the low doses used for lupus, methotrexate reduces the activity of T cells and other inflammatory cells, lowering cytokine production and tissue inflammation. It is used mainly for joint pain (arthritis) and skin rashes in lupus.

### **Warfarin**

Warfarin prevents blood clots by blocking vitamin K, which the liver needs to make clotting factors (proteins that help blood clot). People with lupus and antiphospholipid syndrome (APS) have antibodies that make their blood clot too easily, leading to strokes, a clot in a deep leg vein (deep vein thrombosis), pulmonary embolism (a clot lodging in a lung artery), or pregnancy loss. Warfarin thins the blood just enough to prevent clots without causing too much

bleeding. The dose is carefully adjusted using a blood test called INR (international normalized ratio), with a target usually between 2 and 3 (or 3 to 4 for arterial clots like strokes).

### **NSAIDs (e.g., ibuprofen, naproxen)**

NSAIDs reduce pain, swelling, and fever by blocking enzymes called COX-1 and COX-2, which make prostaglandins (chemicals that cause inflammation and sensitize pain nerves). In lupus, NSAIDs are used for mild joint pain, muscle aches, and serositis (inflammation of the lining around the heart or lungs). They do NOT treat the underlying immune problem and should be used cautiously because people with lupus, especially those with kidney involvement, are at higher risk of kidney damage and stomach ulcers from NSAIDs.

### **Where this information comes from**

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- EULAR/ACR 2019 Classification Criteria for SLE
- American College of Rheumatology (ACR) SLE Guidelines
- Lupus Foundation of America
- UpToDate Clinical Overview (peer-reviewed synthesis)

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