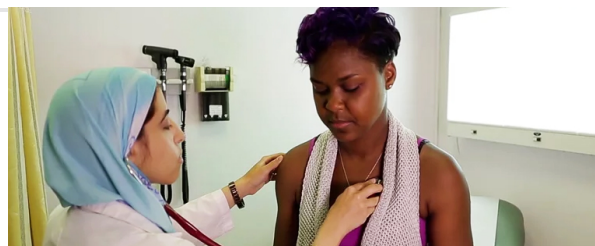


Lupus

Fast Facts

- Lupus occurs ten times more often in women than in men.
- Treatment depends on the organs involved .
- Involvement of the kidneys or/and the brain is the most serious manifestation of lupus.
- People can live well with lupus if they actively work toward good health.
- Sun exposure can lead to lupus flares.
- Carefully plan your pregnancies; lupus can flare during pregnancy and can affect its outcome.



Systemic lupus erythematosus, referred to as SLE or lupus, is a chronic (long-term) disease that causes systemic inflammation which affects multiple organs.

In addition to affecting the skin and joints, it can affect other organs in the body such as the kidneys, the tissue lining the lungs (pleura), heart (pericardium), and brain. Many patients experience fatigue, weight loss, and fever.

Lupus flares vary from mild to serious. Most patients have times when the disease is active, followed by times when the disease is mostly quiet - referred to as a remission. Yet, there is much reason for hope. Improvements in treatment have greatly improved these patients' quality of life and increased their lifespan.

⊕ What causes lupus?

When healthy, our immune system protects the body from foreign germs and cancers. With lupus, the immune system misfires and attacks “self”, the patient’s own tissues, in a process called autoimmunity or “loss of self-tolerance”.

In lupus as the attack goes on, all the branches of the immune system join the fight. This leads to significant and intense inflammation. The cause of Lupus is unknown, as well as what drives its diverse presentation. We know that multiple factors are required, including: the “right” genetic makeup, environmental exposures, and organ specific characteristics. People with lupus may also have an impaired process for clearing old and damaged cells from the body, which in turn provides continuous stimuli to the immune system and leads to abnormal immune response.

Most often, lupus starts in young females in their fertility age. The disease is more common in some ethnic groups, mainly blacks and Asians, and tends to be worse in these groups.

+ How is lupus diagnosed?

Lupus can be hard to detect because it is a complex disease that has many symptoms, and they can come on slowly. As experts in diagnosing and treating autoimmune diseases such as lupus, rheumatologists can best determine whether a patient has lupus and advise them about treatment options.

People with lupus often have symptoms that are not specific to lupus. These include fever, fatigue, weight loss, blood clots, and hair loss in spots or around the hairline. They may also have heartburn, stomach pain, and poor circulation to the fingers and toes. Pregnant women can have miscarriages.

The American College of Rheumatology has a list of symptoms and other measures that doctors can use as a guide to decide if a patient with symptoms has lupus.

- Rashes:
 - butterfly-shaped rash over the cheeks - referred to as malar rash
 - red rash with raised round or oval patches - known as discoid rash
 - rash on skin exposed to the sun
- Mouth sores: sores in the mouth or nose lasting from a few days to more than a month
- Arthritis: tenderness and swelling lasting for a few weeks in two or more joints
- Lung or heart inflammation: swelling of the tissue lining the lungs (referred to as pleurisy or pleuritis) or the heart (pericarditis), which can cause chest pain when breathing deeply
- Kidney problem: blood or protein in the urine, or tests that suggest poor kidney function
- Neurologic problem: seizures, strokes, or psychosis (a mental health problem)
- Abnormal blood tests such as:
 - low blood cell counts: anemia, low white blood cells, or low platelets
 - positive antinuclear antibodies (ANA) result: antibodies that can cause the body to begin attacking itself that are present in nearly all lupus patients
 - certain abnormal antibodies: anti-double-strand DNA (called anti-dsDNA), anti-Smith (referred to as anti-Sm), or antiphospholipid antibodies

If your doctor suspects you have lupus based on your symptoms, a series of blood tests will be done in order to confirm the diagnosis. The most important blood screening test is ANA. If ANA is negative, you don't have lupus. However, if ANA is positive, you might have lupus and will need more specific tests. These blood tests include antibodies to anti-dsDNA and anti-Sm, which are specific to the diagnosis of lupus.

The presence of antiphospholipid antibodies signals a raised risk for certain complications such as miscarriage or blood clots. Doctors also may measure levels of certain complement proteins (a part of the immune system) in the blood, to help detect the disease and follow its progress.

+ How is lupus treated?

Lupus is a chronic disease. The treatment objective is to induce remission. Treatment depends on the type of symptoms you have and how serious they are.

Common treatment options include:

- **Nonsteroidal anti-inflammatory drugs (NSAIDs):** NSAIDs decrease joint swelling, joint pain, fever, and inflammation of the heart and lung linings. These drugs include ibuprofen (brand names Motrin, Advil) and naproxen (Naprosyn, Aleve). Some of these NSAIDs can cause serious side effects like stomach bleeding or kidney damage. Always check with your doctor before taking any medications that are over the counter (without a prescription) for your lupus.
- **Antimalarial drugs:** Hydroxychloroquine (Plaquenil), recommended in every patient with lupus. Hydroxychloroquine was used in the past to prevent and treat malaria and found, during World War 2, to be effective for lupus related arthritis, fatigue, rashes, and mouth sores.
- **Corticosteroids and immune suppressants:** Patients with serious or life-threatening problems such as kidney inflammation, lung or heart involvement, and central nervous system symptoms need more “aggressive” (stronger) treatment. This may include high-dose corticosteroids such as prednisone (Deltasone and others) and drugs that suppress the immune system. Immune suppressants include azathioprine (Imuran), cyclophosphamide (Cytoxan), and cyclosporine (Neoral, Sandimmune). Recently mycophenolate mofetil has been used to treat severe kidney disease in lupus – referred to as lupus nephritis.
- **Biologics:** In 2011, the FDA approved a biologic, belimumab (Benlysta), for the treatment of active SLE in adult patients. Benlysta has shown to be effective in mild forms of Lupus and it is the first new drug approved for lupus since 1955.
- **Combination treatment:** Health care providers may combine a few medications to control lupus and prevent tissue damage. Each treatment has risks and benefits. Most immune-suppressing medications may cause side effects and require close monitoring. Side effects of these drugs may include a raised risk of infections as well as nausea, vomiting, hair loss, diarrhea, high blood pressure, and osteoporosis (weak bones). Rheumatologists may lower the dose of a drug or stop a medicine because of side effects or when the disease goes into remission. As a result, it is important to receive careful and frequent health exams and lab tests to track your symptoms and change your treatment as needed.

+ Broader health impact of lupus

Lupus disease, especially when active, could lead to accelerated atherosclerosis (clogging of the arteries) which can develop in young women and could also lead to heart attacks, heart failure, and strokes. Thus, it is vital that patients with lupus, in addition to controlling their disease, exercise and lower other risk factors for heart disease, such as smoking, high blood pressure, and high cholesterol.

Renal inflammation is one of the common and most serious manifestations of lupus. It could go undetected and can lead to renal failure and dialysis. You can help prevent these serious outcomes by seeking treatment at the first signs of kidney disease. These signs include:

- High blood pressure
- Swollen feet and hands

- Puffiness around your eyes
 - Changes in urination (blood or foam in the urine, going to the bathroom more often at night, or pain or trouble urinating)
-

+ Living with lupus

Most people with lupus can live normal lives. Treatment of lupus has improved, and people with the disease are living longer. Here are a couple of tips that may help you when living with lupus:

- Form a support system. A good doctor-patient relationship and support from family and friends can help you cope with this chronic and often unpredictable illness.
- Get involved in your care. Learn as much as you can about lupus, your medications, and what kind of progress to expect. Take all your medications as your doctor prescribes, and visit your rheumatologist often to prevent serious problems. This lets your doctor keep track of your disease and change your treatment as needed. If you do not live near a rheumatologist, you may need to have your primary care doctor manage your lupus with the help of a rheumatologist.
- Stay active. Exercise helps keep joints flexible and may prevent heart disease and strokes. This does not mean overdoing it. Switch off doing light to moderate exercise with times of rest.
- Avoid excess sun exposure. Sunlight can cause a lupus rash to flare and may even trigger a serious flare of the disease itself. When outdoors on a sunny day, wear protective clothing (long sleeves, a big-brimmed hat) and use lots of sunscreen.

If you are a young woman with lupus and wish to have a baby, carefully plan your pregnancy. With your doctor's guidance, time your pregnancy for when your lupus activity is low. While pregnant, avoid medications that can harm your baby. These include cyclophosphamide, cyclosporine, and mycophenolate mofetil. If you must take any of these medicines, or your disease is very active, use birth control. For more information, see *Pregnancy and Rheumatic Disease*.

Rheumatologists have long been concerned that the female hormone estrogen or treatment with estrogen may cause or worsen lupus. Recent research showed that estrogen therapy can trigger some mild or moderate flares of lupus, but does not cause symptoms to get much worse. Yet, estrogen can raise the risk of blood clots. Thus, you should not take estrogen if your blood tests show antiphospholipid antibodies (meaning you already have a high risk of blood clots).

Updated March 2017 by Ziv Paz, MD and reviewed by the American College of Rheumatology Committee on Communications and Marketing.

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