

Systemic Lupus Erythematosus

Systemic lupus erythematosus (SLE) can cause various symptoms, the most common being joint pains, skin rashes, and tiredness. Problems with kidneys and other organs can occur in severe cases. Treatment includes anti-inflammatory painkillers to ease joint pains. Steroids and/or other medication are needed to reduce inflammation in severe cases.

What is systemic lupus erythematosus?

Systemic lupus erythematosus is a chronic (persistent) disease that causes inflammation in various parts of the body. It is commonly just called SLE or 'lupus'. The severity of SLE ranges from mild to severe.

Who gets SLE?

SLE affects about 3 in 10,000 people in the US. It is 9 times more common in women than men. It most typically develops in women aged between 15 and 40. But, anyone at any age can be affected. It is more common in people from Afro-Caribbean, Asian, or Chinese origin.

What causes SLE?

SLE is an autoimmune disease. The immune system normally makes antibodies to attack bacteria, viruses, and other 'bugs'. In people with autoimmune diseases the immune system makes antibodies against part(s) of the body. This causes inflammation, and may damage the affected part(s) of the body.

There are many autoimmune diseases. The symptoms of each disease depend on which part(s) of the body are attacked by the immune system. In people with SLE, antibodies are made against tissues in various parts of the body. This is why SLE can cause many different symptoms in different parts of the body.

It is not known why SLE or other autoimmune diseases occur. Some factor may trigger the immune system to attack the body's own tissues. Possible triggers include: viruses, infection, medicines, sunlight, or other environmental factors. Hormone changes may play a role in SLE which may explain why it is much more common in women. Some people are more prone to

autoimmune diseases so there may be some genetic factor. However, SLE is not a hereditary disease. Children of people with SLE are not likely to develop it.

What are the symptoms of SLE?

Joint and muscle pains

These are the most common symptoms in SLE. Sometimes only a few joints are affected, sometimes many. The small joints of the hands and feet tend to be the ones affected most. The pains may 'flit' from joint to joint. Joint stiffness is common and is usually worse first thing in the morning. Mild joint swelling may occur but severe arthritis with joint damage is unusual. Serious arthritis similar to rheumatoid arthritis affects about 1 in 20 people with SLE.

Skin, mouth, and hair

A red rash that develops over the cheeks and nose is common (the 'butterfly rash'). Other areas of skin exposed to sunlight (hands, wrists etc) may also develop a rash. Exposure to sunlight tends to make the rash worse. Various other rashes may develop. The blood vessels just under the skin may also be affected and cause poor circulation to the fingers and toes (Raynaud's phenomenon). Mouth ulcers are more common in people with SLE.

Some hair may fall out (alopecia). Any hair loss tends to be minor and cause hair 'thinning' rather than bald patches. Sometimes hair loss is severe.

Tiredness

This is common and sometimes becomes severe and difficult to cope with.

Blood and lymph

A mild anemia is common. Other blood problems such as reduced numbers of white blood cells or platelets (the particles that help the blood to clot) are less common. A tendency to form blood clots is an uncommon complication. Some lymph glands may swell.

Heart and lungs

The tissues that cover the heart and lung (the pleura and pericardium) may become inflamed. This can cause pleurisy (pains in the side of the chest) or pericarditis (central chest pains). The actual heart or lung tissue is less commonly affected.

Kidneys

Inflammation of the kidneys may cause protein and blood to leak into the urine. This is not usually severe, but occasionally the kidneys can become badly affected. Kidney failure is an uncommon complication.

Brain

Inflammation of the brain may occur but is uncommon. It can lead to mental health problems or seizures. Anxiety and depression are common but these are not due to the inflammation. They are usually due to difficulties that some people have coping with the disease.

Other

There are various other uncommon symptoms.

How does SLE progress?

In some cases the symptoms develop quite slowly. At first they may be confused with other problems as there are many possible causes of joint pains and tiredness. Sometimes several symptoms occur together. Sometimes different symptoms follow each other. Symptoms range from mild to severe. For example:

- **Mild SLE.** Many people with SLE just have joint and/or skin symptoms with tiredness. These are unpleasant but are not serious or life threatening.
- **Moderate SLE.** This includes some inflammation of other parts of the body apart from joints and skin. This may include pleurisy, pericarditis, or mild kidney inflammation. The symptoms of moderate SLE are unpleasant, but are not likely to cause much 'damage'.
- **Severe SLE.** In some cases, severe inflammation develops which can cause damage to organs such as the heart, lung, brain, or kidneys. This can be life threatening.

SLE is a chronic (persistent) disease. However, typically there are times when the disease flares up (relapses) and symptoms become worse for a few weeks or so, sometimes longer. These relapses tend to alternate with times when symptoms settle (remission). However, once you have SLE it is unusual for symptoms to completely go away. The reason why symptoms flare-up or settle down is not understood.

How is SLE diagnosed?

If symptoms suggest SLE, your doctor will usually do some blood tests. Most people with SLE have an antibody called antinuclear antibody in their blood. Various other antibodies are also associated with SLE. These tests are not foolproof as these antibodies may also occur in people with other conditions. Sometimes they occur in perfectly well people. However, typical symptoms combined with high levels of certain antibodies usually indicate that you have developed SLE.

Once SLE is diagnosed, you will normally be advised to have regular checks and tests. For example, regular blood tests to check for anemia, urine tests to check for kidney problems, etc.

A blood test to measure a blood chemical called 'complement' (part of the immune system) can assess the activity of the disease. The level of this chemical reflects how 'active' the disease is.

Other tests, scans, x-rays, etc, may be advised to check on the function of the heart, kidneys, etc if the disease is thought to be affecting these organs.

What are the treatments for SLE?

There is no cure for SLE. However, the condition can be controlled and symptoms eased in most cases. A specialist who advises on treatment sees most people with SLE regularly. Treatments

and dosages may vary from time to time, depending on the severity of the disease or flare-up of symptoms, and which parts of the body are affected. You may even not need any treatment if you have very mild symptoms. Treatment options include the following.

Anti-inflammatory painkillers

One is commonly prescribed to ease any joint or muscle pains. For example, ibuprofen, naproxen, diclofenac, or piroxicam are commonly prescribed - but there are many others. The main possible side effects from these medicines are stomach and gut problems such as pain or bleeding in the stomach. If necessary, other medication can be prescribed to protect the stomach from these side effects.

Hydroxychloroquine

This medicine eases skin rashes, joint and muscle pains, and tiredness. It is not clear how it works. It may take 6-12 weeks for it to become fully effective. The dose is often reduced to a lower 'maintenance' dose once symptoms have eased. Many people with SLE take this medicine long term to keep symptoms away. Side effects are uncommon. For example, stomach upsets, headache, skin reactions, flu-like aches and tiredness occur in less than 1 in 10 treated people. These side effects tend to ease after a short while. Serious side effects are rare. The most serious is damage to the eye. If you are taking this medicine, report any new eye symptoms such as blurred vision to your doctor.

Steroid medication

Steroid tablets are usually advised if you develop severe SLE. Steroids reduce inflammation and have made a big impact in the treatment of SLE. A large dose may be needed at first to bring symptoms under control. The dose is then gradually reduced to the lowest dose that controls symptoms. Steroids may cause side effects if taken for long periods. These include thinning of the bones (osteoporosis), thinning of the skin, weight gain, muscle wasting, high blood pressure, and other problems. This is why the lowest possible dose is used.

Some people are able to stop steroid treatment from time to time if the symptoms settle. It can be started again if symptoms flare up again. However, you should never stop steroid medication suddenly. A gradual reduction of dose, monitored by a doctor, is needed.

Immunosuppressants

Medicines such as azothiaprine or cyclophosphamide may be advised if you have severe SLE. These work by suppressing the immune system. They are powerful medicines. If you take one of these you need to have regular blood and urine tests to look out for possible side effects.

An immunosuppressant is sometimes advised in addition to steroid medication. The two together tend to work better than either alone. Also, the dose of steroid that you need may then be less. This means that any side effects from steroids may be less severe.

Treating complications

Some people with severe SLE may also need treatment for complications that may occur. For example, treatment for high blood pressure, kidney damage, etc.

What is the outlook (prognosis) for people with SLE?

Older textbooks give a bleak picture for SLE and should be ignored. Mild forms of the disease are now recognized more often. This is due to antibody tests being done more routinely on people who would, in the past, have been labeled as having 'aches and pains' without a clear diagnosis. We now know that some of these people have mild SLE. Modern treatments are also more effective. So, the outlook is much better than in the past.

For many people with SLE, symptoms are mild or moderate with little risk to life. The joint and skin symptoms may persist, but can usually be eased with treatment.

For a few, SLE is severe and even life threatening. Severe inflammation of the kidneys leading to kidney failure is uncommon, but is the main reason why some people die from SLE. Severe brain involvement is also rare but can be very serious. However, modern immunosuppressive treatments have improved the outlook even for people with severe disease.

Some people find that symptoms settle in their middle age and they can come off all treatment.

Some other points about SLE

- **Avoid the sun** Strong sunlight can aggravate symptoms (not just the skin symptoms). Long sleeved clothing and wide brimmed hats are best in sunny weather. On hot sunny days you should wear a sun block on exposed skin with a protection factor of 25 or above that protects against UVA and UVB.
- **Try to avoid infections.** If you have SLE you are more prone to infection, particularly if you take steroids or immunosuppressant medication. Avoid contact with people who have infections.
- **Pregnancy.** Some women with SLE have a higher chance of miscarriage. This depends on a type of antibody that sometimes develops called antiphospholipid antibody. Women who have badly inflamed kidneys due to SLE may have high blood pressure in pregnancy. However, most women with mild or well-controlled SLE at the start of pregnancy are likely to go through pregnancy with few problems.
- **Some contraceptive pills** may not be advised depending on disease severity. A doctor or nurse will advise on the best method of contraception.
- **Other autoimmune diseases** such as Sjögren's syndrome and thyroid problems are more common than average if you have SLE. (For example, about 1 in 10 people with SLE also have a thyroid problem.)