## What is Lupus?

Lupus is a chronic autoimmune disorder that occurs when the body's immune system attacks its own tissues and organs. Inflammation caused by lupus can affect many different body systems, including the joints, skin, kidneys, blood cells, brain, heart, lungs and intestines.

Under normal circumstances, the body's immune system protects the body against "foreign" invaders like viruses, bacteria and parasites. In lupus, the immune system malfunctions and produces antibodies and cells which attack its own organs – hence lupus is known as an "auto-immune" disease. The reason why this happens is not known but lupus flares are commonly triggered by a combination of factors such as exposure to sunlight, stress or infection. There is also a genetic link since lupus occasionally runs in families.

## Who gets Lupus?

Lupus occurs more frequently in women than in men. Asians and Afro-Americans are more prone to develop lupus than Caucasians and the disorder is more severe in these ethnic groups.

Four types of Lupus exist:

a. **Systemic Lupus Erythematosus (SLE**) – the generalised and most common form
b. **Discoid Lupus Erythematosus** – only affecting the skin
c. **Drug-induced Lupus** – Lupus caused by drugs
d. **Neonatal Lupus** – Lupus in babies born to mothers with SLE

The outlook for people with lupus was once grim, but diagnosis and treatment of the disorder has improved tremendously in recent years. With early diagnosis and treatment, most people with lupus can lead active lives.

## What are the signs and symptoms of Lupus?

No two cases of lupus are exactly alike. Signs and symptoms may come on suddenly or develop slowly, may be mild or severe and may be temporary or last for a long period of time. Most people with lupus experience episodes – called flares – of worsening signs and symptoms that eventually improve or even disappear completely for a time with treatment. The course of the disorder is unpredictable, hence long-term treatment and follow-up is essential. The signs and symptoms of lupus will depend on which body systems are affected by the disorder.

In general, SLE signs and symptoms include the following:

- Fever, fatigue and weight loss.
- Joint pain, stiffness, swelling and diffuse muscle aches.
- Butterfly-shaped rash on the face that covers the cheeks and bridge of the nose and worsens with sun-exposure.
- Hair loss, mouth ulcers and easy bruising.
- Chest pain on breathing and shortness of breath.
- Swelling around the eye-lids, swelling of the feet and legs and decrease in urine output.
- When lupus affects the nervous system, patients can present with headache, memory or behavioural changes, drowsiness, stroke, muscle weakness, paralysis or fits.

## How is Lupus diagnosed?

Diagnosing lupus is difficult because the disease varies considerably from person to person and the signs and symptoms come and go unpredictably and overlap with many other diseases. Therefore, doctors may not initially consider lupus until the signs and symptoms become more definite.

Even then, diagnosis can often be challenging and a number of laboratory tests are necessary to confirm the diagnosis.

These tests include the following:

**Full blood count** - This test measures the levels of haemoglobin, number of red blood cells, white blood cells and platelets. Results may indicate the presence of anaemia, or low white blood cell or platelet count. An extremely low platelet count can result in spontaneous bleeding in the skin (bruises), in the stomach or even in the brain.

**Erythrocyte Sedimentation Rate (ESR)** – the ESR is raised in many disorders including lupus. It is sometimes a good measure of disorder activity and as your disorder improves your ESR may drop.

**Urine examination** - An examination of your urine may show an increase in red blood cells or protein level. This can occur if lupus has affected your kidneys.

**Kidney function tests** - Blood tests can assess how well your kidneys are functioning.

**Antinuclear antibody (ANA)** - A positive test for these antibodies indicates a stimulated immune system which is common in lupus and other autoimmune diseases. However, a positive ANA test is not always indicative of lupus since certain infections or drugs can lead to a positive test. In fact, a small proportion of normal individuals can have a slightly raised ANA antibody test. Therefore this test needs to be interpreted in conjunction with a proper history and physical examination.

**Anti-dsDNA test** - This test is often done together with the ANA test. Patients with lupus and kidney involvement often have a raised anti-dsDNA test.

## How is Lupus treated?

Specialised clinics may result in better outcomes. Treatment of lupus depends on the signs and symptoms and which organs are involved. Determining what medications to use requires a careful discussion of the benefits and risks with a rheumatologist. For quick control of lupus, a rheumatologist may recommend more powerful drugs in higher doses initially but as the disease flare subsides, the dosage can usually be tapered off slowly and carefully. More aggressive lupus usually requires more powerful drugs.

In general, when first diagnosed with lupus, your doctor may recommend the following medications:

a. **Non-steroidal anti-inflammatory drugs (NSAIDs)**
These include diclofenac acid (Voltaren) and the COX 2 inhibitors (Celebrex and Arcoxia). They are effective in controlling fever, muscle aches, joint pains and swelling.

b. **Antimalarial drugs**
Although there is no known relationship between malaria and lupus, these medications have proved useful especially in patients with mild lupus and those with skin involvement. Hydroxychloroquine (Plaquenil) is the most commonly prescribed antimalarial drug.

c. **Corticosteroids**
These drugs counter the inflammation of lupus and are very effective. They can have serious long term side-effects including weight gain, easy bruising, high blood pressure, diabetes, thinning of the bones (osteoporosis) and an increased risk of infection. Very often, doctors need to prescribe corticosteroids (eg. prednisolone) in order to prevent permanent organ damage such as kidney failure or even death. To help reduce the side-effects, your doctor will try to find the lowest dose that controls the disease and prescribe the medicine for the shortest possible period of time.

d. **Immunosuppressive drugs**
These drugs suppress the immune system and may be useful in

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serious cases of lupus including patients with severe kidney or brain involvement. The most often used immunosuppressive drugs are cyclophosphamide and azathioprine. Cyclophosphamide is often given by injection into the veins. Side-effects include an increased risk of infection, low white cell count, liver damage and infertility. These drugs should only be used under close supervision by a rheumatologist or a nephrologist (kidney specialist).

e. **Mycophenolate Mofetil (cellcept)**
This is a relatively new immunosuppressive drug which has proven to be effective in lupus affecting the kidneys and other major organs. It has fewer side-effects than corticosteroids and is proving to be an effective alternative to cyclophosphamide.

## Lifestyle changes

Lifestyle changes are needed to reduce the incidence of relapses.

- Get adequate rest and sleep
- Be sun smart – avoid sun-bathing and stay out of the sun entirely when it is the strongest i.e. 8am – 6pm.
- Use sunblock and wear protective clothing
- Get regular exercise but do not overstrain your body
- Maintain a healthy diet
- Do not smoke and avoid excessive alcohol intake

## Special problems in Lupus patients

**Pregnancy and Lupus**
Doctors used to recommend that patients with lupus should avoid pregnancy. However, this is no longer the case as more effective treatment strategies are available. Pregnancy is usually safe in patients with mild lupus. In severe lupus, pregnancy is best avoided during the active phase of the disorder. Always discuss this problem with your doctor before embarking on pregnancy. Certain drugs used to treat lupus can affect the unborn baby and are best avoided in lupus patients who intend to become pregnant.

**Neonatal Lupus (Lupus in newborn babies)**
In rare cases, babies born to mothers with lupus can develop a condition called neonatal lupus. This is usually a benign condition and the baby will have some rash which subsides in a few weeks' time. Very occasionally, such babies will have a disturbance in heart rate which will require treatment by a neonatal cardiologist.

To optimise care, it is best to have a team of rheumatologist, obstetrician and paediatrician working together.

**Coping and support**
Coping with lupus can be stressful. Patients with lupus often experience anxiety, depression and frustration. Strategies for coping with lupus include :

- learning all you can about lupus
- gathering support among your friends and family
- connecting with others who have lupus

## In Summary

Lupus can be very mild or can be life-threatening. The diagnosis of lupus however is not a catastrophe or a death sentence. Many effective treatments are now available.

As with all illnesses, early diagnosis and treatment are important. Never stop or reduce medications on your own or try new medications without the knowledge of your doctor. As with all chronic diseases, you will need to be under medication and observation for long periods of time. However, once the acute symptoms are under control, you can return to your work and former lifestyle with minor modifications.

Establish a good working relationship with your doctor and nurse and keep updated on new developments of the disease by attending regular talks and seminars. Always have a positive outlook and attitude.

*Information modified from National Arthritis Foundation, Singapore*

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