

LUPUS

A Guide for Patients



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This factsheet gives general information about lupus, what it is, some common symptoms, diagnosis and treatments.

What is lupus?

Systemic Lupus Erythematosus: Is a disease where the immune system becomes overactive and starts making antibodies against its own cells and is therefore called an auto-immune disease. When the disease is active, immune responses can cause inflammation in cells which can then affect one or many tissues of the body: skin, joints, muscles, blood vessels, blood cells, brain and nerves, other organs such as lungs, heart, kidneys, gastrointestinal tract and/or the linings around internal organs. Patients have a variable course with the disease lasting many years in the majority.

Discoid Lupus (DLE): In general DLE is a disease just affecting the skin and rarely affects the internal organs, i.e. rarely becomes systemic. Most studies suggest that approximately 5% of patients with discoid lupus at some stage may suffer a generalised flare of the disease, involving joints, kidneys and autoantibody production and may progress to developing Systemic Lupus Erythematosus. If left untreated Discoid Lupus may leave a scar.

Drug-induced Lupus (DILE): can occur during the administration of certain drugs in susceptible individuals. Hydralazine, Phenytoin, anti-TNF drugs, procainamide and minocycline can lead to lupus like symptoms which usually resolve with time after the medication is stopped by the doctor.

Who gets lupus?

Lupus affects women nine times as often as men, usually occurring in women in the childbearing years. It can, however, occur in children or post-menopause. Afro-Caribbean and South Asian races are more likely to develop lupus.

How does lupus affect the body?

Lupus is a highly variable disease which may be affected by the interaction of genetic, environmental and hormonal factors. Potentially, it can affect any organ or tissue of the body. In any individual patient, however, only some organs may ever be affected; the tissues and systems involved and the intensity of involvement may vary with time. The disease is very much an individual illness differing from person to person.



What are the features of lupus?

Fatigue, joint and muscle pain and muscle weakness, flu-like illness, skin rashes (including the classical “butterfly” rash on the cheeks and nose), hair-loss and mouth ulcers are the most common symptoms. However internal organ involvement can include chest wall pain (pleurisy), kidney disease and brain inflammation. Some patients with lupus have a clotting tendency and this can present, for instance, as a thrombosis (blood clot) in the vein or an artery.



How serious is lupus?

Lupus is different for each person. Many will experience a mild flare which could affect various joints, skin and cause fatigue. With time, treatment may help improve these symptoms and this is when the disease is said to be in remission. Some patients only experience one flare, however, others may experience periodic increased disease activity followed by periods of remission. While these milder flares often disrupt normal activities and patients may need to make certain adaptations to their lifestyle (to make life easier at these times) they do not cause a threat to internal organs. However, despite treatment some patients may still only feel some improvement and activity of the disease continues at a low level. For a smaller group of patients lupus is more serious and despite the use of various treatments the disease remains difficult to control. Major kidney, central nervous system or vascular involvement of the disease require a more intensive medical follow up and treatment, and can have a serious impact on patients’ health. With new treatments and care pathways outcome results have improved.



How is lupus diagnosed?

- Lupus has a diverse presentation of symptoms often making it difficult to diagnose. Patient reporting of these symptoms to their doctor is the first step.
- Thorough medical history and physical examination provide most of the data required for diagnosis along with laboratory tests.
- Referral should be made to a specialist centre for early diagnosis and appropriate treatment can then be started to treat the disease and minimise any potential for organ damage as kidney involvement can be serious.

Recommended blood tests for diagnosis and monitoring include:

Positive anti-nuclear antibody - ANA.

Raised double stranded DNA (dsDNA) and Extractable nuclear Antigens (ENA) such as Sm, Ro, RNP and La.

Full Blood Count (FBC), Complement (C3 and C4) and Blood Chemistry Tests.

Inflammatory markers such as C reactive protein (CRP) and Erythrocyte Sedimentation Rate (ESR).

Urine testing - Urine dipstick at diagnosis and clinic visits to check for blood and protein.

Urine albumin creatinine ratio if protein is present in the urine sample to evaluate kidney involvement.

Other - Skin Biopsy of a rash or a kidney biopsy if the doctor is concerned that these organs may be affected by lupus.

Chest X-ray and ECG (Electrocardiogram).

Blood pressure.

Echocardiogram, Brain Scan, Lung Functions Tests, Abdominal CT Scan.



How is lupus treated?

Symptom control is the goal for non-organ-threatening lupus (e.g. skin and joints) whereas organ preservation is the aim for organ threatening lupus (e.g. kidney).

Medication - the type of medication is determined by the clinical manifestations and severity of symptoms. Patients may need to try various treatments before finding the one that is both effective in treating the symptoms but also has the fewest side-effects.

Anti-inflammatory agents are used for mild disease and painful symptoms - joints, muscle pain, pleurisy, headaches, etc. Non-steroidal anti-inflammatories should be avoided in patients with kidney disease.

Hydroxychloroquine is a drug originally developed for malaria that also helps patients with lupus symptoms. It is a useful first line treatment in mild to moderate disease and can often control joint symptoms, pleurisy and skin involvement. Benefit usually occurs gradually over several months.

Steroids - Generally reserved for more serious manifestations of the disease (vasculitis, central nervous system, kidney, etc.). It is sometimes necessary to use steroids for non-organ threatening manifestations such as arthritis, pleurisy, but side effects are common and doses are kept as low as possible and for as short a time as possible.

Immunosuppressive drugs - These are reserved for moderate to severe disease either not responsive to first line treatments or requiring prolonged use of unacceptably high doses of steroids. Examples include: Cyclophosphamide,

Azathioprine, Mycophenolate Mofetil and Myfortic. Regular careful monitoring by the specialist centre and General Practitioner is required to identify any side-effects early before they become dangerous.

Biological agents - There are two biologic drugs used to treat lupus. Rituximab and Belimumab are newer therapies used to treat moderate to severe disease. Rituximab is selective because it works by targeting and depleting cells which play a role in autoantibody production. Belimumab is a monoclonal antibody which works by inhibiting B-Lymphocyte stimulator (BLyS) which stimulates antibody production.

More information regarding medication can be obtained from the 'LUPUS and Medication' factsheet available from National Office.

Non-medication measures

- Plan restful periods during the day, pacing of daily activities with regular meal breaks, adequate sleep and you may need to consider making further changes during time of flare.
- Minimising stress: by considering lifestyle modification that makes living with lupus more manageable. Developing good communication links and support from your medical team, family members and employer is helpful in keeping the disease under control. Regular gentle exercise also helps improve fatigue and reduce stress.
- Avoid provoking factors - minimise sun exposure and excessive heat and treat infections early. Non live vaccinations are reported as being safe and effective in lupus as benefits outweigh the risks. However, vaccines should be avoided when lupus disease is active.
- Patient awareness of early symptoms, which may signal a flare and prompt reporting of these to their physician allowing earlier treatment.
- Stop smoking as it reduces the effects of Hydroxychloroquine.
- Access to personalised treatment at expert clinics, specialising in lupus management.

What is the outlook?

In summary, SLE is a potentially serious disease, which can affect almost any system of the body. We do not yet know what causes it. The outlook for survival has improved in recent years and most patients with SLE will continue to have mild disease. If diagnosed early and treated appropriately at an early stage, lupus may settle and ultimately go into remission - i.e. the patient requires no, or minimal, medication. With good partnership, support and co-operation between the patient, family and doctor, the problems associated with the disease can be significantly modified.

The LUPUS UK Range of Factsheets

A range of factsheets are available as follows:

1. LUPUS Incidence within the Community
2. LUPUS A Guide for Patients
3. LUPUS The Symptoms and Diagnosis
4. LUPUS The Joints and Muscles
5. LUPUS The Skin and Hair
6. LUPUS Fatigue and your Lifestyle
7. LUPUS and Pregnancy
8. LUPUS and Blood Disorders
9. LUPUS and Medication
10. LUPUS and the Kidneys
11. LUPUS and Associated Conditions
12. LUPUS and the Brain
13. LUPUS The Heart and Lungs
14. LUPUS The Mouth, Nose and Eyes
15. LUPUS and Light Sensitivity
16. LUPUS and the Feet
17. LUPUS and Men
18. LUPUS and Mixed Connective Tissue Disease

LUPUS UK is the registered national charity caring for people with lupus and has over 5,500 members who are supported by the Regional Groups.

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Please contact our National Office should you require further information about the sources used in the production of this factsheet or for further information about lupus. LUPUS UK will be pleased to provide a booklist and details of membership.

LUPUS UK is certified under the requirements of the Information Standard.



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