

A Short Guide to Lupus and Visiting your Doctor



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LUPUS UK is the national charity caring for those with systemic lupus erythematosus (SLE) and discoid lupus erythematosus (DLE), supporting people as they develop the symptoms prior to diagnosis and those already diagnosed.

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



About this Booklet

This booklet is intended to provide a short guide to lupus and to offer helpful advice to the patient with lupus when visiting a doctor.

This booklet does not constitute or replace medical advice and should be treated as such.

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1. 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.

What does the immune system do?

The immune system functions to protect the body against foreign invaders i.e. abnormal molecules, bacteria, viruses, parasites, foreign tissues including transfused blood cells, organ transplants and deranged cells such as cancer cells. It does so by a complex series of cellular interactions culminating in:

- i. The formation of antibodies to foreign bodies (outsiders) or antigen which then combines with it.
- ii. Generation of cells, which interact directly with the outsider both of which facilitate damage and removal of the foreign invaders.

Implicit in proper functioning of this system is the ability to differentiate between foreign material (non-self) and the body's own tissue (self). In SLE that recognition system is somehow flawed and the body's immune system generates antibodies and cells hostile to its own tissue.

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, Asian and Eastern races are more likely to have lupus.



2. 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. Lupus can affect almost any part of the body including the skin, joints, blood vessels and organs such as the heart and kidneys. 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.

People with lupus have symptoms that can range from mild to severe, and may come and go over time. 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.

When lupus affects the kidneys, it is usually referred to as ‘lupus nephritis’. Most often this causes no symptoms and is detected by testing the urine or blood for abnormalities. It is important to recognize, since in severe cases it can lead to permanent kidney damage. If lupus nephritis causes very large amounts of protein to leak from the kidneys, swelling in the legs or more generally (‘oedema’) may develop.

What causes lupus?

The causes of lupus are not fully understood but are likely to be due to a combination of genetic and environmental factors. Lupus causes periods of illness (‘flares’) and periods of feeling well with no symptoms or signs of disease (‘remission’). It is not infectious and cannot be caught from another person.

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.

Pregnancy

If planning pregnancy it is advisable to receive additional advice from a doctor who specialises in lupus and pregnancy. It may be necessary to perform further tests and investigations so that best advice can be given about possible risks to mother and baby. Lupus is more likely to be stable during the pregnancy if the disease was stable prior to conception. Some women do have flares of lupus during pregnancy and need careful observation. Lupus kidney disease needs particularly careful monitoring as there are no symptoms from this and, if uncontrolled, can seriously affect the health of mother and baby long-term.

Lupus and anti-phospholipid syndrome may affect the way the afterbirth (placenta) works. In early pregnancy if the placenta fails to work miscarriage will occur. Women with anti-phospholipid antibodies are particularly at risk of this complication as their blood is more “sticky”. Specific treatment is advised in this group of women and tests will be performed to look for these antibodies if a previous miscarriage or stillbirth has occurred. For further information see the LUPUS UK factsheet - ‘LUPUS and Pregnancy’.

3. 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.

Who looks after patients with lupus?

Many different doctors can be involved in looking after people with lupus but nephrologists (kidney doctors) and rheumatologists (arthritis specialists) are most often involved. Since lupus is a complicated condition, it is usually a good idea for you to see doctors who specialise in treating the disease. Your GP will also make an important contribution to looking after your lupus and your general health. You may see a specialist lupus nurse as well.

4. How is lupus treated?

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. 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).

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 taken 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 vaccines are reported as being safe and effective in lupus patients as benefits outweigh the risks. However, live vaccines should be avoided when lupus disease is active, and in general it is preferable to administer non-live vaccines during times when lupus is not 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.

In summary, SLE is a potentially serious disease, which can affect almost any system of the body. We do not know yet what causes it. The outlook for survival has improved in recent years and most patients with SLE will continue to have mild disease. With co-operation between the patient, family and doctor the problems associated with the disease can be significantly modified.

5. Some helpful advice for you, the patient,

When visiting your doctor

The most important way you can stay healthy is to be an active member of your own health care team. One way to get high-quality health care is to find and use information and take an active role in all the decisions made about your care.

Research has shown that patients who have good relationships with their doctors tend to be more satisfied with their care – and to have better results.

Around 80% of the information your doctor needs to make a correct diagnosis comes from what you say to him/her. The remaining pieces of the puzzle are found when you are examined and have tests carried out.

This verbal information, ‘the history’, is very important and is mostly obtained in two ways – the first is when you answer your doctor’s questions, the second is when you say things without being asked.

Never be frightened to give your doctor information; something you think is insignificant may be just the piece that cracks the diagnosis.

It can be easy in everyday life to misunderstand something; in medicine it can be frighteningly easy.

Patients and doctors are assumed to speak the same language but, sometimes, they don’t. Doctors often use technical language and some words, which may mean one thing to him/her, may mean something completely different to you.

This is why it’s important to spell things out and not to be embarrassed if you don’t understand something. No one will think you’re stupid. It makes everyone’s life easier and can avoid problems. Similarly, if your doctor asks you what you mean when you say something, then try to explain it in a different way so that he/she understands.

Here are some tips to help you and your doctor become partners in improving your health care.

Give Information. Don’t wait to be asked!

- * You know important things about your symptoms and your health history. Tell your doctor what you think he/she needs to know. Doctors are human too and may forget to ask you about certain things.
- * It is important to tell your doctor personal information – even if it makes you feel embarrassed or uncomfortable.
- * Have a “health history” with you – remember to keep this updated.



- * Have any medicines you are taking – or a list of your medicines – include when and how often you take them and what strength.
- * Tell your doctor about any allergies or reactions you may have had to your medicines.
- * Tell your doctor about any herbal/natural products you use or alternative medicines or treatments you receive.

Get Information

- * Ask questions. If you don't, your doctor may think you understand everything that has been said.
- * Write down your questions beforehand. List the most important ones first to make sure they get asked and answered.
- * You might want to have someone with you. This person can also help ask questions or help you understand and/or remember the answers.
- * If it will help, ask your doctor to draw pictures.
- * Take notes; your doctor won't mind if it helps you to understand/remember.
- * Let your doctor know if you need more time. If there is not time at this visit, ask if the practice nurse can help you or make another appointment.

Check that your doctor has washed his/her hands before examining you – don't be shy of doing this! Research shows that hand washing can prevent the spread of infections.

Take Information Home

- * Ask your doctor for written instructions, particularly if being prescribed new medication.
- * The surgery/hospital may have leaflets that can help you. If not, ask how you can obtain these.
- * Ask for details of patient support groups.

When you leave the Surgery/Consulting Room

- * If you have problems with your medicine or your symptoms worsen, make contact with your doctor.
- * If you have had tests and are expecting to hear from your doctor but do not, ring the surgery or consultant's secretary.
- * If your doctor said you need special tests, check that these are booked.
- * If your doctor wants you to see a specialist, make sure you are referred.
- * Read any information leaflets you may have been given and keep them while you are taking the treatment.
- * Make contact with a patient support group.

Remember

Be honest when you tell your doctor how you feel, say not only where you are hurting but how long you have been hurting.

Tell him/her how you feel about your condition and treatment. If you don't want to take a treatment it is better to say so and discuss other possibilities than to go away and not bother with it.

Your doctor wants to help you to be as well as possible but he/she cannot do it on his/her own.

Your care will be much more successful if your doctor knows how you are feeling. You don't have to say you're "fine"!

If you are upset about your condition, talk to your doctor. He/she may be able to recommend a counsellor or psychologist who can help you deal with the emotions you are feeling and any decisions you may want to make.

It is your body that is being treated – be responsible for it and make your doctor/patient relationship a partnership.



Publicity materials, leaflets, posters, a dvd for the newly diagnosed, media releases and more are always available from the charity's National Office for better awareness about lupus in clinics, hospitals and public places.