

The management of Lupus

This is the lay version of the EULAR recommendations for the management of people with Lupus. The original publication can be downloaded from the EULAR website: www.eular.org.

Bertsias G, et al. EULAR recommendations for the management of systemic lupus erythematosus (SLE): Report of a Task Force of the European Standing Committee for International Clinical Studies Including Therapeutics (ESCISIT). *Ann Rheum Dis* 2008;67(2):195–205. [doi:10.1136/ard.2007.070367](https://doi.org/10.1136/ard.2007.070367)

Introduction

Recommendations give advice to doctors and patients about the best way to treat and manage diseases. EULAR has written recommendations on the management of people with systemic lupus erythematosus (also called SLE or Lupus for short). Lupus is an autoimmune disease where the immune system attacks the body's own tissues and causes inflammation. Lupus is a complicated disease. It has many different symptoms, including joint pain, fatigue (tiredness) and skin rash. Lupus can also affect internal organs such as the kidneys and cause neurological problems.

The recommendations were written by doctors. They looked at the evidence on the management of people with Lupus. They also discussed their expert opinion to achieve a level of agreement.

More recently EULAR has created additional recommendations for specific aspects in the care of people with Lupus, namely Neuropsychiatric Lupus, Lupus Nephritis, and fertility and pregnancy in Lupus (see 'other recommendations' at the end of this document). The recommendations summarised here look at Lupus in general.

What do we already know?

Lupus is a complicated disease, and can be hard to manage. Often the disease comes and goes, sometimes called 'waxing and waning' or flares and remission. Because of the variety of different symptoms and parts of the body that can be affected, many different doctors are involved in the care of people with Lupus. Lupus is most common in women in their child-bearing years.

What do the recommendations say?

The recommendations fall into five categories: general management, pregnancy in Lupus, blood clotting and miscarriage (antiphospholipid syndrome), Lupus affecting the brain (Neuropsychiatric Lupus), and Lupus affecting the kidneys (Lupus Nephritis).

Overall, there are 12 statements or recommendations. Each recommendation is based on available scientific evidence or expert opinion. The more stars a recommendation has the stronger the evidence is and the more important it is that you and your doctor follow it.

One star (*) means it is a weak recommendation with limited evidence.

Two stars (**) means it is a weak recommendation with some evidence.

Three stars (***) means it is a strong recommendation with some evidence.

Four stars (****) means it is a strong recommendation with a lot of evidence.

1. General management

- **Symptoms and tests should be considered when evaluating a person with Lupus and deciding on their prognosis, or how well they are likely to do in the future.****

Symptoms and the results of laboratory tests may predict a person's disease outcome, and can tell us which major organs are affected by the Lupus. Lupus patients should receive the same tests as anyone else going to their doctor with neuropsychiatric symptoms. These might include a physical examination, laboratory tests and imaging such as MRI (magnetic resonance imaging).

- **New symptoms and changes may be used to diagnose flares and monitor disease activity.****

The appearance of new symptoms or changes in laboratory test results can be used to monitor disease. They can also help your doctor to diagnose and recognise flares or changes in the disease.

- **People with Lupus are more likely to have other diseases and need to be carefully managed and treated.****

People with Lupus have a higher than normal risk of having infections and other diseases such as diabetes, high blood pressure, osteoporosis (brittle bones), cancer and heart disease. If you have Lupus you should try to minimise your risks. Doctors need to diagnose and quickly treat any other diseases that their Lupus patients have.

- **Drug choice will depend on the symptoms of the disease and how severe it is.**/****

For people with no major organs affected, antimalarial drugs or steroids may be used. Non-steroidal anti-inflammatory drugs (NSAIDs) may be used for short periods to reduce pain and swelling. Immunosuppressive drugs may be used if you cannot take steroids.

- **Add-on (adjunct) treatments may be used in some people.**/****

If you have Lupus and suffer from skin rashes, take steps to protect yourself from the sun. General lifestyle modifications will help to keep you healthy, such as stopping smoking, controlling your weight, and taking exercise. Some people may need to take drugs to control their blood pressure or dietary supplements that contain calcium and vitamin D. Oral contraceptives should be used carefully if you have Lupus.

2. Neuropsychiatric Lupus (that affecting the brain and cognition)

- **Diagnosis should be similar to that for anyone else with neuropsychiatric symptoms.****

Lupus patients should receive the same tests as anyone else going to their doctor with neuropsychiatric symptoms. These may include a physical examination, laboratory tests and imaging such as MRI (magnetic resonance imaging).

- **Immunosuppressive drugs may be used in people with neuropsychiatric lupus.***

If neuropsychiatric symptoms are thought to be due to inflammation in the eyes, brain or nervous system, immunosuppressive drugs can be used.

For additional information on Neuropsychiatric Lupus, you could look at reference 1 in the list below.

3. Pregnancy

- **Women with Lupus are more at risk of developing complications during pregnancy.****

Women with Lupus are just as fertile as those without the disease. However, pregnancy may cause your Lupus to worsen. Women with Lupus are more likely to develop a complication called pre-eclampsia during pregnancy, and should be carefully monitored until they give birth.

- **The babies of women with Lupus may be more at risk of birth complications, and some drugs should be avoided during pregnancy.******

Women with Lupus have a higher risk of miscarriage, stillbirth and premature birth. The babies of mothers with Lupus may also have complications with their growth and heart development.

Some drugs for Lupus can be used during pregnancy – for example:

- Prelone, Orapred, Predicort or Milliepred (a steroid, also called prednisolone)
- Imuran (also called azathioprine)
- Plaquenil (also called hydroxychloroquine).
- Low-dose aspirin.

Other drugs for Lupus must be avoided during pregnancy – including:

- CellCept (also called mycophenolate mofetil)
- Cytoxan, Endoxan, Noesar, Procytox, Revimmune or Cycloblastin (also called cyclophosphamide)
- Trexall or Rheumatrex (also called methotrexate).

For additional information on fertility and pregnancy in people with Lupus, you could look at reference 3 in the list below.

4. Antiphospholipid syndrome

- **Low-dose aspirin may be used to prevent antiphospholipid syndrome, which may cause blood clotting and miscarriage.*/****

In people with Lupus and antiphospholipid syndrome, anticoagulant drugs can be used to prevent thrombosis. In pregnant women with Lupus and antiphospholipid syndrome, heparin and aspirin can be used to reduce the chances of miscarriage.

5. Lupus nephritis (affecting the kidneys)

- **Kidney tests may help to predict how well drugs will work in people with lupus nephritis.****

Tests such as renal biopsy, urine sediment analysis, proteinuria, and kidney function may predict outcomes of therapy of lupus nephritis, but the results need to be interpreted carefully. Changes in immunological tests have a lower ability to predict the response to treatment and should be used only for additional information.

- **Drug therapy can slow the progression to end-stage kidney disease.******

In patients with proliferative lupus nephritis, glucocorticoids in combination with immunosuppressive drugs or CellCept (also called mycophenolate mofetil) can slow the progression to end-stage disease. Cytoxan, Endoxan, Noesar, Procytox, Revimmune or Cycloblastin (also called cyclophosphamide) also work, but can cause side effects. Some people have flares of their disease again after remission, so your doctor should monitor you carefully.

- **Dialysis or kidney transplant may be needed in some people with lupus nephritis.*****

Dialysis and kidney transplant both work well in people with Lupus who have end-stage kidney disease, although transplantation is preferred.

For additional information on Lupus Nephritis, you could look at references 2 and 3 in the list below.

Summary

Overall, the recommendations say that it is important for you and your doctor to work together to monitor and manage your disease and particular symptoms, and to get the best possible results from treatment. If you have Lupus these recommendations will give you tips about what to expect from your doctor.

If you have any questions or concerns about your disease or your medication, speak to your doctor.

Other recommendations and further reading

1. Bertsias G, et al. EULAR recommendations for the management of systemic lupus erythematosus with neuropsychiatric manifestations: report of a task force of the EULAR standing committee for clinical affairs. *Ann Rheum Dis* 2010; 69(12): 2074–82. [doi: 10.1136/ard.2010.130476](https://doi.org/10.1136/ard.2010.130476)
2. Bertsias G, et al. Joint European League Against Rheumatism and European Renal Association-European Dialysis and Transplant Association (EULAR/ERA-EDTA) recommendations for the management of adult and paediatric lupus nephritis. *Ann Rheum Dis* 2012;71(11): 1771–82. [doi: 10.1136/annrheumdis-2012-201940](https://doi.org/10.1136/annrheumdis-2012-201940)
3. Andreoli L, et al. EULAR recommendations for women's health and the management of family planning, assisted reproduction, pregnancy and menopause in patients with systemic lupus erythematosus and/or antiphospholipid syndrome. *Ann Rheum Dis* 2016 Jul 25. [doi: 10.1136/annrheumdis-2016-209770](https://doi.org/10.1136/annrheumdis-2016-209770) [Epub ahead of print]