

## Updated: SLE recommendations

This is the lay version of the EULAR recommendations for managing systemic lupus erythematosus. The original publication can be downloaded from the EULAR website: [www.eular.org](http://www.eular.org).

[Fanouriakis A, Kostopoulou M, Andersen J, et al EULAR recommendations for the management of systemic lupus erythematosus: 2023 update Annals of the Rheumatic Diseases Published Online First: 12 October 2023. doi: 10.1136/ard-2023-224762](#)

### Introduction

EULAR – the European Alliance of Associations for Rheumatology – gives advice to doctors, nurses, and patients about the best way to treat and manage diseases. In 2023, EULAR updated its recommendations on systemic lupus erythematosus (often shortened to SLE). Doctors, nurses, other health professionals, and patients worked together to develop this new advice. The patients in the team ensured that the patient point of view was included.

### What do we already know?

SLE is an inflammatory autoimmune disease, where the immune system mistakenly attacks tissues in the body. SLE does not look the same in every patient, but it can affect all organs or tissues in the body, including the skin, joints, kidneys, nervous system, lungs, heart, and blood cells. People with SLE can have different symptoms, including fatigue, rash, and joint pain or swelling. There are also a number of different antibodies associated with SLE, which might be detected in a blood test. These include antinuclear antibodies (ANA) and antiphospholipid antibodies (aPL), among others.

EULAR first published recommendations around the management of SLE in 2008. Traditionally, there have not been many treatment options for this disease. However, since the last update in 2019, new drugs have been approved and options are beginning to expand.

Of note, the previous recommendations included 33 statements across four topics. This update has only 13 statements. This structural change is in line with current EULAR practice to streamline the number of recommendation statements in any one publication. Because of this change, all overarching principles and individual recommendations are considered to be new, even where the essence has remained unchanged from the previous version.

### What do the recommendations say?

In total, there are 5 overarching principles and 13 recommendations. The principles say that people with SLE require multidisciplinary, individualized management based on patient education and shared decision-making – and taking into consideration costs to both the patient and society. Disease activity should be assessed at each clinic visit, and organ damage checked at least annually. Non-pharmacological interventions, such as using sun protection, exercising, stopping smoking, and eating a healthy diet, as well as measures to promote bone health, are important to improve long-term outcomes. Drug treatments should be chosen based on each person's individual characteristics and preferences, as well as their disease type and severity of organ involvement. Early diagnosis, regular organ screening, prompt treatment, and strict adherence are essential to prevent flares and organ damage, as well as to improve the long-term outcome, and enhance quality of life.

Each recommendation is based on the best current knowledge from studies of scientific evidence or expert opinion. The more stars a recommendation has the stronger the evidence is. However, recommendations with limited scientific evidence may be important, because the experts can have a strong opinion even when the published evidence may be lacking.

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

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

Three stars (\*\*\*) means it is a recommendation with quite a lot of scientific evidence.

Four stars (\*\*\*\*) means it is a recommendation supported with a lot of scientific evidence.

## Recommendations

- **Hydroxychloroquine is the mainstay of treatment for SLE.\*\*\***

An antimalarial drug called hydroxychloroquine is recommended for everyone with SLE, unless there is a specific reason why you cannot take it (a contraindication). The target dose is based on your weight, and is 5 mg/kg per day. However, this advice should be tailored based on each person's individual risk for flare and eye toxicity.

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- **If you need glucocorticoids, the dose will depend on your organ involvement, but should be low and temporary. Pulses of intravenous methylprednisolone can be considered in some people.\*\***

Glucocorticoids are a type of steroid drugs. The dose should be based on the type and severity of your organ involvement. The dose should be reduced to maintenance dose of  $\leq 5$  mg/day (prednisone equivalent) and, when possible, withdrawn, because long-term therapy with glucocorticoids is associated with adverse effects. If you have moderate-to-severe disease, pulses of intravenous methylprednisolone (125–1000 mg/day for 1–3 days) can be considered.

- **If you do not respond to hydroxychloroquine or are unable to reduce your steroid dose, immunosuppressive\*\*\* and/or biologic agents\*\*\*\* should be considered.**

Some people will not respond to hydroxychloroquine, alone or in combination with glucocorticoids. Others might not be able to reduce their glucocorticoid doses low enough for it to be used for a long period. If this is the case, other drugs should be added. Your healthcare team might consider adding immunomodulating or immunosuppressive agents such as methotrexate, azathioprine, or mycophenolate, or they might consider adding belimumab or anifrolumab.

- **In people with organ- or life-threatening disease, intravenous cyclophosphamide should be considered; in refractory cases, rituximab may be considered.\*\***

Some people with SLE will have serious organ- or life-threatening disease that might need treatment with intravenous cyclophosphamide. Rituximab is also an option for people with refractory disease.

- **If you have active skin disease, initial treatment should include topical agents,\*\*\* antimalarials,\*\*\*\* and/or glucocorticoids.\*\* Immunosuppressives or biologics can be considered as second-line.**

The first-line treatment for active skin disease is topical glucocorticoids or calcineurin inhibitors, antimalarials (hydroxychloroquine, chloroquine), and/or systemic glucocorticoids. Around 40% of people will not respond to first-line treatments, and they can then try methotrexate,\*\*\* mycophenolate,\*\* anifrolumab,\*\*\*\* or belimumab\*\*\* as second-line therapies.

- **If you have active neuropsychiatric disease caused by SLE, you might be offered glucocorticoids and immunosuppressives,\*\*\*\* or antiplatelet agents/anticoagulants\*\* – this will depend on your specific neuropsychiatric symptom(s).**

Some people with SLE will have neuropsychiatric symptoms. It can be difficult to link any symptoms to your SLE. Depending on your specific problems, you might be offered glucocorticoids and immunosuppressives agents for inflammatory manifestations such as myelopathy or an acute confusional state. Or you might be given antiplatelet agents or anticoagulants if you have cerebrovascular disease or a stroke.

- **If you have acute severe autoimmune thrombocytopenia, you might need high-dose glucocorticoids\*\*\* with or without immunoglobulin G,\*\* rituximab,\*\*\* and cyclophosphamide\*\* followed by maintenance therapy.**

If you have severe autoimmune thrombocytopenia, you might be treated with high-dose glucocorticoids (including pulses of intravenous methylprednisolone) – with or without intravenous immunoglobulin G, rituximab, or high-dose intravenous cyclophosphamide. This initial treatment might be followed by maintenance with rituximab, azathioprine, mycophenolate, or cyclosporine.

- **People with active proliferative lupus nephritis should receive low-dose cyclophosphamide or mycophenolate and glucocorticoids. Combination therapy should be considered.\*\*\*\***

Lupus nephritis is the term used when the disease affects the kidneys. People with active proliferative lupus nephritis should receive low-dose intravenous cyclophosphamide or mycophenolate and pulses of intravenous methylprednisolone followed by lower oral doses. Your healthcare team may also consider using combination therapy, for example, belimumab plus cyclophosphamide or mycophenolate, or calcineurin inhibitors such as voclosporin or tacrolimus plus mycophenolate.

- **After renal response, lupus nephritis treatment should continue for at least 3 years.\*\*\***

If your kidneys respond to therapy, your lupus nephritis treatment should continue for at least 3 years. If you were initially treated with mycophenolate alone or in combination with belimumab or a calcineurin inhibitor, you should remain on these drugs. If you were initially treated with cyclophosphamide – either on its own or in combination with belimumab – then cyclophosphamide should be replaced with azathioprine or mycophenolate.

- **In you are at high-risk for renal failure, you may receive high-dose cyclophosphamide in combination with pulse methylprednisolone.\*\*\*\***

Being at high risk of renal (kidney) failure is defined as having a reduced function called GFR (glomerular filtration rate), presence in your kidney biopsy specimen of cellular crescents or fibrinoid necrosis, or severe interstitial inflammation, at the onset of nephritis. If this is your case, your healthcare team may consider using high-dose intravenous cyclophosphamide in combination with pulse intravenous methylprednisolone.

- **If you achieve sustained remission, you might be able to gradually reduce your treatment. Glucocorticoids should be stopped first.\*\*\***

Remission is when you have no signs or symptoms of your disease. If this happens and is sustained for a period of time, you may be able to gradually taper (reduce) your treatment. Any glucocorticoids should be withdrawn first, before trying to taper any other treatments.

- **If you have thrombotic antiphospholipid syndrome and have a blood clot, you will need long-term vitamin K antagonists. Low dose aspirin should also be considered in people with a high-risk antiphospholipid antibody profile.\*\*\***

SLE associated with thrombotic antiphospholipid syndrome (APS) should be managed with long-term vitamin K antagonists, such as warfarin, after the first arterial or unprovoked venous thrombotic event (blood clot). Low-dose aspirin (75–100 mg/day) should be considered in people with SLE without a thrombotic event, but who have a high-risk antiphospholipid antibody profile.

- **You can take care of your health by keeping up-to-date with vaccinations, managing bone health, kidney and cardiovascular risks, and attending screening for malignancies.\***

People with rheumatic diseases such as SLE can be at a higher risk of infections than other people. Vaccines (immunizations) can help prevent infections. You should consider getting vaccinated against herpes zoster virus, human papillomavirus, influenza, COVID-19, and pneumococcus. You can also

take steps to manage the health of your bones, kidneys, and heart. Screening for malignancies should also be performed.

### Summary

Overall, these recommendations give guidance to health professionals and patients about the management and treatment of people with SLE. You should work with your healthcare team to make an informed decision about your disease and its treatment. These recommendations focus on pharmacological options, but there are also non-pharmacological treatments that might help your disease. Separate recommendations have been published, and you can read them [here](#).

Recommendations with just one or two stars are based mainly on expert opinion and not backed up by studies, but these may be as important as those with three or four stars.

If you have any questions or concerns about your disease or your medication, you should speak to a health professional involved in your care.