

Sjögren's syndrome

This is the lay version of the EULAR recommendations for the use of topical and systemic treatments in people with Sjögren's syndrome. The original publication can be downloaded from the EULAR website: www.eular.org.

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Introduction

EULAR recommendations give advice to doctors, health professionals – such as nurses, occupational therapists, physiotherapists or psychologists – and patients about the best way to treat and manage diseases.

EULAR has developed new recommendations for the use of topical and systemic treatments in people with Sjögren's syndrome. Systemic medicines are applied by mouth or as an injection or infusion and spread through your body in your blood, reaching all cells. This is different to topical treatments, which are usually applied to the skin to treat a local problem. Doctors, health professionals and patients have worked together in a taskforce to develop these recommendations. The patients working on the team ensured that the patient point of view was integrated in the recommendations.

What do we already know?

Sjögren's syndrome is a systemic autoimmune disease. Autoimmune disease happens when a person's immune system attacks its own cells and organs; in Sjögren's syndrome the glands that secrete fluids, such as saliva or tears, are attacked. One of the main features of Sjögren's syndrome is dryness in the eyes and mouth (called sicca symptoms), as well as fatigue and pain, but it can also affect the internal organs. Sjögren's syndrome is much more common in women – only one in ten people affected are men.

The treatment for people with Sjögren's syndrome has not changed much in recent years, and there are no specific treatment targets beyond relieving symptoms.

What do the recommendations say?

Overall, there are three over-arching principles and 12 recommendations. The principles say that people with Sjögren's syndrome should be looked after at expert centers that have doctors from different specialties working together in a multi-disciplinary team. They also emphasize that the first treatment for dryness should be symptomatic relief using topical therapies, but that systemic therapies can be considered for people with active systemic disease.

Each recommendation is based on available scientific evidence and expert opinion. The more stars a recommendation has the stronger the evidence is and the more important it is that you and your doctor should follow it.

One star (*) means there is very limited or no scientific evidence, but significant expert opinion.

Two stars (**) means it is a weak recommendation with some scientific evidence and significant expert opinion.

Three stars (***) means it is a strong recommendation with quite a lot of scientific evidence and expert opinion.

Four stars (****) means it is a strong recommendation supported with a lot of scientific evidence and expert opinion.

- **The function of your salivary glands should be checked before starting treatment for oral dryness.***

Oral dryness can be very bothersome, and people's reporting of the symptoms can be subjective. How well the salivary glands are working should be tested using a clinical tool that can measure the flow of saliva.

- **The first treatment for oral dryness depends on how badly your salivary glands are working.******

People with mild impairment may have non-drug stimulation treatment, such as sweets, cough drops (lozenges) or chewing gum to help generate saliva in the mouth. For people with moderate oral dryness, drugs can be used, but these can cause you to sweat a lot, among other side effects. People whose salivary glands do not work at all need saliva substitutes. These are given as sprays, gels or mouth washes.

- **Artificial tears and eye gels or ointments should be the first treatment for eye dryness.******

If you have Sjögren's syndrome and dry eyes, you should use artificial tears at least twice a day, and as often as once an hour if needed. Ointments can be used at bedtime for relief through the night.

- **Drops containing immunosuppressives or autologous serum can be only prescribed for people with complicated eye dryness.*****

If you have a complicated ocular course, your ophthalmologist may prescribe you drops containing steroids or drugs that dampen down your immune system (immunosuppressives) or a type of serum made from your own blood that you can use in your eyes. If those options do not work for you, you may need plugs inserted in your tear ducts to stop any fluid in your eyes from draining away.

- **People with fatigue or pain should be checked for other diseases, and severity scored by their doctor using specific tools.***

People with Sjögren's syndrome often say that the worst symptoms are joint or muscle pain, and feeling very tired. The healthcare team looking after you should check to make sure that you do not have any other underlying conditions that could be causing these.

- **You may be prescribed painkillers to relieve muscle or joint pain.****

If you have muscle or joint pain, you can use painkillers. Your doctor may prescribe you short courses of tablets, or you can use pain-relief gels. For very bad pain, stronger options might be used, but you should not use any opioids.

- **Treatment of underlying disease should take into account the organs that are affected.****

The management of systemic Sjögren must be tailored to the specific organ involved and the severity evaluated using a tool called ESSDAI. This stands for the 'EULAR Sjögren's Syndrome Disease Activity Index', and is a way to measure joint pain and inflammation. Any treatment you receive should be appropriate for your type of disease and the parts of your body that are affected.

- **Steroids should be used at the lowest dose and for the shortest time needed to get your disease under control.****

Steroids are a good treatment option for people with systemic disease features, but they can have a lot of side effects, so they should be used at the lowest dose (5 mg a day or less) and for the shortest time necessary to control your disease.

- **Immunosuppressive drugs can be used to reduce the need for steroids.****

Some people might need long-term treatment, especially those with severe organ damage. To help

reduce the use of steroids, other types of drugs that damp down or suppress the immune system can be used. These may include azathioprine, mycophenolate, cyclophosphamide, methotrexate or leflunomide.

- **Biological drugs that target B-cells can be considered in people with severe and recurring systemic disease.******

Biologicals are a type of medicine made from proteins. These target particular cells or molecules in the body and help to reduce inflammation. Drugs such as rituximab or belimumab can be tried in people with severe Sjögren's syndrome symptoms that keep coming back despite other treatments.

- **As a general rule, people with systemic disease should be treated first with steroids, then immunosuppressive agents, and then biologics. Some medicines can be combined.***

If you have Sjögren's syndrome, you will probably be treated first with steroids – a type of anti-inflammatory medicine. Steroids should be used at the lowest possible dose for the shortest possible time. If steroids do not work, other immunosuppressive agents should be tried next, and finally the biologics.

- **If you have B-cell lymphoma, treatment should be tailored to you as an individual, depending on your specific disease subtype and stage.****

Lymphoma is a rare but very serious complication associated with Sjögren's syndrome. If you have B-cell lymphoma, your treatment should be tailored to the specific type, and you should be looked after by a hematologist or an oncologist.

Summary

Overall, these recommendations highlight how to best use topical and systemic treatments in people with Sjögren's syndrome. These recommendations provide you guidance on what to expect from healthcare providers, and what support may be offered.

Recommendations with one or two stars are based on expert opinion and less rigorous study designs and cannot be backed up by effectiveness studies. However, these may be as important as those with three or four stars, which are backed up by appropriate effectiveness studies. It is important to understand that the absence of evidence for effectiveness does not imply that a statement is not applicable.

If you have any questions or concerns about your disease or its treatment, you should speak to a healthcare provider involved in your care.