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Women in Vision
& Eye Research Ireland

SJÖGRENS SYNDROME

The information leaflet briefly outlines our current understanding of the condition and provides advice from key specialists involved in the treatment and management of Sjögrens Syndrome.

WHAT IS SJÖGRENS SYNDROME?

Sjögren's Syndrome (SS) is an autoimmune disease which affects approximately 1 in 200 people in the developed world. In Sjögren's syndrome, specialised secretory glands that produce saliva and tears become inflamed and are badly damaged. As a result of this, these glands stop working leading to dry eyes and dry mouth.

In addition, many patients feel tired all the time and experience aches and pains which can be quite debilitating. The condition occurs when the body's immune system, which normally provides protection from infection, begins to attack parts of the body instead, particularly the tear and salivary glands. This causes many symptoms ranging from mild discomfort, irritation and vision distortion, to severe damage to the corneal surface.

In addition to dry eye and dry mouth some patients can also develop systemic complications which range in severity from fatigue, joint and muscle pain, skin rashes to pulmonary complications which range from mild reduction in lung function to severe complications such as idiopathic pulmonary fibrosis. At present there is no way to predict which patients will go on to develop these disease associated complications. Treatment options are reactive and limited, and the disease can have a significant impact patients' quality of life.

WHAT ARE TWO TYPES OF SJÖGRENS SYNDROME?

- ~ Primary Sjögren's Syndrome occurs by itself without any other illness.
- ~ Secondary Sjögren's Syndrome develops in a person who has another autoimmune condition, typically rheumatoid arthritis (RA) or systemic lupus erythematosus (SLE).



WOMENS HEALTH SPECIALIST

SJÖGREN'S SYNDROME, THE MENOPAUSE AND AND HORMONE REPLACEMENT THERAPY

The Menopause is an inevitable part of aging for most women. The symptoms may be mild or they can be so severe that significant disruption to the quality of life may result.

Women with Sjögren's Syndrome often already have some of the classic menopausal symptoms such as dryness of the eyes, mouth, vagina and vulva. Menopausal dryness may exacerbate symptoms of dryness, especially Vaginal Dryness, which may persist for many years.

Other genito-urinary symptoms such as leaking of urine or bladder irritation can also be linked to Sjögren's Syndrome and these may be exacerbated as natural estrogen levels deplete after the Menopause.

Pain and discomfort during sex may become worse.

Sjögren's Syndrome associated with disorders such as Lupus (but not primary Sjögren's Syndrome) can be linked to increase risk of Osteoporosis and Cardiovascular disease. Estrogen decline in the menopause also accelerates these conditions

HOWEVER, some doctors may hesitate to prescribe HRT for menopause symptom relief in women with Sjögren's Syndrome (esp secondary Sjögren's Syndrome) as certain types of HRT (oral/ equine estrogen mainly) are linked to thrombosis risk which can also be increased in other rheumatological conditions (e.g., SLE with anti-phospholipid antibodies).

There is good data to support the cautious use of low dose, transdermal or "body identical" estrogen and healthier progestagen in women with blood clot risk. Ideally, advice, prescriptions and management should at least be commenced by doctors with Menopause specialist training.

RHEUMATOLOGY

OUR ROLE

Investigate people to check if parts of the body beyond the glands in eyes and necks are affected and advise on treatment and help co-ordinate care across different specialists

ADVICE

1. Make sure you are attending your dentist regularly
2. Manage Fatigue with a regular exercise regime and good sleep hygiene
3. Stop smoking: HSE - Sign up and quit smoking <https://bit.ly/3oewoRf>
4. Thrush (candida) is more common in patients with Sjögren's and can be managed with either gels, drops or tablets
5. Inflammation in your joints can happen. Symptoms include swelling across your knuckles and hands as well as early morning stiffness that lasts more than one hour in the morning. If this happens treatment can include anti-inflammatory, steroid or hydroxychloroquine
6. If you develop new persistent swelling in a gland in your neck, armpit etc that lasts more than a few weeks tell your GP or rheumatologist
7. If you intend to become pregnant please let us know. Women with Sjögren's syndrome have a small risk of passing on antibodies which can cause slowing of the heart in the foetus. You will need to be seen more regularly in any pregnancy to make sure everything is okay.
8. Get support from Arthritis Ireland www.arthritisireland.ie/sjogrens-syndrome



OPHTHALMOLOGY

OCULAR SYMPTOMS OF SJÖGREN'S SYNDROME:

- ~ Dry eyes
- ~ Blurry vision
- ~ Burning or stinging eyes
- ~ Gritty sensation
- ~ Light sensitivity
- ~ Heavy or tired eyes
- ~ Eyelid inflammation

Ocular lubricants are the backbone of treatment for dry eye disease and are often sufficient in managing mild to moderate dry eye. These are available over the counter. If your symptoms are particularly bothersome during the night, an ointment can be helpful. These also come in preparation with vitamin A, which can aid ocular healing. However, these may blur your vision as they have a high viscosity so are most useful just before bed.

In moderate to severe cases, autologous serum drops, topical steroids, or topical immunomodulatory solutions may also be used. Surgical cautery or temporary plugs can be used to seal tear ducts, preventing your drops and what little tears the lacrimal gland is still producing from draining away.

ADVICE:

- ~ Use your drops frequently
- ~ Manage your environment, avoiding air conditioning where possible
- ~ Ensure your drops are preservative free, as they have less ocular surface toxicity
- ~ Some oral medications may also add to your dry eye symptoms, such as blood pressure medications, acne medications, anti-depressants etc. If this is the case, it is important to manage medication choices in conjunction with your prescribing physician and ophthalmologist.

DERMATOLOGY

ALWAYS USE HIGH FACTOR SUNSCREEN FROM MARCH TO SEPTEMBER

The sun emits Ultraviolet light which can trigger auto immune conditions such as Sjögren's. It is important to use an SPF 30 or 50 every day between the 1st of March to the end of September, whether in Ireland or abroad, to prevent flares and also to ensure protection against skin cancer

SWAP OUT YOUR USUAL SHOWER GELS AND SOAPS TO FRAGRANCE AND SOAP FREE CLEANSERS

Eliminating soap and fragranced products from your routine will prevent your skin from drying out. Use a soap and fragrance free shampoo, conditioner and shower wash.

MOISTURISE YOUR SKIN REGULARLY

Hydrating your skin from the outside in is very important to maintain its integrity and to rehydrate dry sensitive skin. Choose a product that you like to use and feels comfortable on your skin. Again this should be a fragrance free, low preservative moisturiser.

CHECK IN WITH YOUR DERMATOLOGIST

If you are experiencing skin symptoms which are difficult to control, ask your G.P. for a referral to your local dermatologist, who has experience with auto-immune conditions which affect the skin and can help resolve ongoing skin issues.

GET SUPPORT FROM THE IRISH SKIN FOUNDATION

The Irish Skin foundation (www.irishskin.ie), are a fantastic resource for all things skin related. The website has a free helpline, where you can connect with dermatology nurses for advice and guidance on all skin conditions.





DENTAL ADVICE

FOR PATIENTS WITH DRY MOUTH (XEROSTOMIA)

Sjögren's is a complex condition, with a wide range of clinical presentations requiring multidisciplinary care. The impact of dry mouth on quality of life and on the hard and soft tissues of the mouth is often under appreciated. Adequate oral care can be further compromised by fatigue, joint pain/stiffness and the medications required to manage various aspects of the disease. It is important to actively engage regularly with a supportive, experienced dental team, make them aware of your issues and be pro-active regarding self-management. They should provide you with expert advice and a personalized oral homecare plan to increase comfort and reduce your risk of rampant decay, gum disease and oral infections such as thrush.

Focus on disease prevention and increasing oral function and comfort. As dry mouth is multifactorial, it is important to review all relevant aspects, including: your overall fluid intake (hydration), use of oral lubrication, salivary stimulation, limit caffeine and alcohol intake; review all medication with your doctors, as many products increase dryness and alternatives may be available. Heat produced by smoking and vaping causes significant dryness in mouth/throat and smoking greatly increases your risks of other diseases including mouth, head and neck cancer. Consider smoking cessation, alcohol reduction, diet modification, stress management, choice of oral hygiene products, and denture care.

HYDRATION: Aim is 2 litres decaffeinated, non-fizzy, non-alcoholic fluids daily, if kidney function normal

ORAL LUBRICATION: Use salivary replacement therapies to replace missing saliva. Ask your pharmacy /dentist for samples of products such as BioXtra, Biotene, Xerostom, Oralieve, MouthKote etc. and see what you like. Best results when use full set: tooth paste + gel + rinse. Avoid toothpastes with SLS as this foaming agent/soap makes the mouth much drier. Sips of water and ice chips are soothing; some like chamomile or sage tea, a little coconut oil, butter– according to personal preference.

SALIVARY STIMULATION: Sucking sugar-free sweets can be helpful however sugar-free gum is more effective and better for oral hygiene. Avoid harsh flavours, gentle fruit flavours are best. Sucking fruit slices – lemon, melon, pineapple etc may help but some find it too harsh. Best to use in small amounts as the fruit acid can soften tooth enamel.

Other options include xylitol tablets, and drugs such as Pilocarpine and Bethanechol which can stimulate salivary flow, but side-effects may be unpleasant. Acupuncture does not appear to help in Sjögren's related dry mouth. Trials of novel agents seem promising but early days.

DECAY PREVENTION: Brush teeth and remove and clean any denture after every snack/meal. Food tends to stick to teeth when the mouth is dry. Floss at least once a day – otherwise you are only cleaning 60% of tooth surfaces. Use Teepee brushes to clean between teeth. Rinse with an alcohol free mouthwash containing chlorhexidine after brushing and flossing to reduce plaque 'stickiness'. Use a lubricating gentle toothpaste 3-4 times/day and a high fluoride toothpaste at night (Colgate Durophat 2,800 or 5,000) to strengthen teeth, on prescription only. Dentists can also apply fluoride varnish and gels. GC Paste Plus also helps strengthen teeth with extra calcium and phosphate. Avoid sugar and fizzy drinks. Dry mouth increases the risk of gum (periodontal) disease; careful, frequent, daily oral home care and regular contact with your dental team is the key to prevention and management.

BEHAVIOUR MODIFICATION: Diet modification increases comfort and taste. Avoid dry, spicy, acidic foods like bread, crackers, biscuits, chilli. Opt for moist food with sauces, gravy, stews, casseroles, eggs, smoothies, yogurt, custard, mousse, etc. Slow cooker very useful. Avoid sucking sugar containing sweets / drinks to reduce oral infection risk. Carry a small bottle of water with you always. Focus on the foods that you CAN eat rather than what is difficult to eat and try to stay positive despite the challenges of this condition. E45 Cream, Neutrogena hand / lip cream are all helpful if lip/skin dry.





BRAIN HEALTH ADVICE

Many patients with Sjögrens report brain fog. In some cases cognitive difficulties are the first clinical manifestations, preceding diagnosis of primary Sjögrens by an average of two years.

Brain fog is a general term that describes a collection of symptoms. The most common symptoms are:

- ~ **Loss of mental clarity**
- ~ **Inability to focus or concentrate**
- ~ **Problems with learning and remembering, slow thinking**
- ~ **Issues with language or word finding and**
- ~ **Trouble navigating spaces, which many people would describe as clumsiness**
- ~ **Mental fatigue**

When you have brain fog your symptoms are persistent, occur regularly and interfere with the quality of your life, your relationships, and your work.

Brain fog is not a disease, disorder or diagnosis rather it is a sign or a symptom of:

- ~ An underlying health condition (e.g. auto-immune conditions, inflammatory diseases, chronic pain, most chronic conditions, depression, anxiety, type 2 diabetes, and some cancers)
- ~ A side-effect of medication (chemotherapy, medications for pain, auto-immune diseases, pain and depression, anti-histamines, anti-nausea tablets)
- ~ The result of hormonal changes (e.g. PMS, pregnancy, menopause, thyroid dysfunction, etc)
- ~ Nutritional or dietary deficiency (e.g. vitamin B12, iron, folate or Omega-3 deficiency)
- ~ Lifestyle factors (poor sleep, chronic stress, lack of exercise, lack of mental stimulations)

LONG TERM CHANGES

For most people multiple factors contribute until a critical threshold is reached and brain fog is experienced. Making the lifestyle adjustments listed below can make a huge difference by boosting the health of your brain and freeing up cognitive resources.

PRIORITISE SLEEP - keep to a regular routine, manage exposure to light and develop a wind-down routine. Manage stress - smile more, laugh more, have fun, set aside time each day to do something that you love - find your joy.

EXERCISE YOUR BODY AND YOUR BRAIN - both are critical for brain health

NOURISH YOUR BRAIN - The food you eat is the fuel your brain needs to function properly - rubbish in rubbish out. Shop for your brain, best evidence is for a Mediterranean diet.

SHORT TERM STRATEGIES

Avoid multi-tasking, it's a myth. The brain is task switching and that comes at a cost, you take longer make more errors. Do one thing at a time.

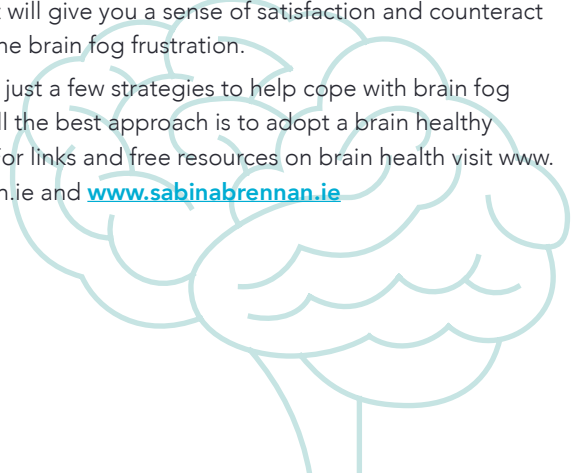
DECLUTTER, YOUR BRAIN - (write worries down, make to do lists), your work space, your computer. Your brain has to process billions of bits of information and filter out what's not relevant – why waste valuable brain resources on clutter.

ESTABLISH ROUTINES - Your brain constantly scans your behaviour for routines it can automate – because and behaviour that is repeated at regular intervals can be handed over to an unconscious part of the brain (basal ganglia) which uses up less energy than your thinking brain thus freeing up brain energy for the more complex stuff you struggle with when you have brain fog. Regularise all of your mundane tasks so your brain can automate them.

STOCKPILE BORING TASKS FOR THE BAD DAYS

Nothing is more frustrating than losing an entire day to brain fog because your brain can't rise to the challenge of your job. There are days when you have to accept that your brain won't do what you want it to but rather than lose the entire day you can still be productive by turning your attention to that stockpile of boring 'no brain' tasks – ticking them off your 'to do' list will give you a sense of satisfaction and counteract some of the brain fog frustration.

These are just a few strategies to help cope with brain fog but overall the best approach is to adopt a brain healthy lifestyle. For links and free resources on brain health visit www.superbrain.ie and www.sabinabrennan.ie





RESEARCH

OUR AIM

To increase understanding about what is leading to the uncontrolled inflammation that is seen in Sjögren's Syndrome. Increasing our understanding through research could help find ways to control this inflammation and potentially lead to improved diagnosis and better treatment options.

- ~ In trying to understand what is happening at the ocular surface we have begun to look at a network of regulators which control all of the processes within our cells. These regulators or controllers are small pieces of genetic material termed micro RNAs.
- ~ We found that patients with autoimmune dry eye disease had different amounts and types of regulatory micro RNAs or controllers in their cells compared to eye cells from healthy controls.
- ~ This suggested that changes in the amounts of these regulators could be causing the increased production of inflammation-triggering messenger molecules from our immune cells ultimately leading to dry eye disease in Sjögren's Syndrome patients.
- ~ Our research is a first step toward a potential new treatment, and much more pre-clinical testing is needed before we can develop it into something that is ready for patients. However, our research provides the opportunity to possibly treat the root cause of the disease rather than just the symptoms.
- ~ You can find out more about the research the Ocular Immunology Research Group (OIRG) are currently doing in RCSI on the Sjögrens Research Ireland webpage: <https://sjogrensresearch-ireland.eu/>
- ~ If you are interested in participating in a research study (by completing questionnaires or providing samples) or being involved in designing future webinars or patient material please go to our Sjögrens Research Ireland webpage and complete the expression of interest form on the link below: sjogrensresearch-ireland.eu/information-for-patients/
- ~ For those interested in finding out more about the Sjögrens patient advocacy group please Email Sjögrens Ireland at the following email: sjogrensireland@gmail.com





ADVICE FROM SJÖGRENS PATIENTS TO SOMEONE RECENTLY DIAGNOSED WITH SJÖGRENS SYNDROME

BECOME YOUR OWN ADVOCATE - DO YOUR OWN RESEARCH

- ~ There is a lack of awareness of Sjögrens Syndrome among the general public and within the healthcare setting.
- ~ Educate yourself so that you can educate your friends, family and healthcare professionals involved in your care.
- ~ Don't read all about Sjögrens online and don't panic. Not everyone gets all the symptoms.
- ~ Do your research using selective reputable sources e.g. BSSA, Arthritis Ireland etc
- ~ Inform yourself as to how Sjögrens can affect different parts of your body.

MAINTAIN POSITIVE COMMUNICATION WITH YOUR GP

- ~ Make sure you that you have a good GP and you maintain positive communication with them. Seek an alternative GP if you are unhappy with your treatment too often Sjögren symptoms are mistaken for depression. It is important to see a doctor who specialises in women's health, hormones and menopause etc.
- ~ Expect some frustration as it can take time see the benefits of medication. You will have more bad days than good days initially.
- ~ Keep a log of your symptoms and note when, how often they occur. Include a description of pain or dryness level to assist/support when you visit health professionals.

PRIORITISE YOUR HEALTH

Be proactive in seeking referrals for the different parts of your body that are affected eg eyes and teeth. Take particular care of your teeth and improve your dental care routine. Moisturise your mouth, eyes and skin.

Stay physically active, attend a physiotherapist to help with a graduated exercise program. Exercise can help alleviate some fatigue and pain, so don't be afraid to get active, but remember to be kind to yourself and take it one day at a time.

Do not overdo things. Learn to pace yourself and spread work and housework evenly throughout the week and rest when symptoms flare.

SUPPORT

YOU ARE NOT ALONE –

Join local/national and international organisations and chat rooms that focus on autoimmune diseases such as Healthunlocked, British Sjögrens Support Group (www.bssa.uk.net), Chronic Pain Ireland (www.chronicpain.ie) and Arthritis Ireland (www.arthritisireland.ie) etc.

Talk to as many Sjögrens sufferers as possible each will have new tips and advice. Don't be afraid to ask for help when needed. Seek specialist help if your disease is not in control.





Women in Vision
& Eye Research Ireland

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USEFUL LINKS:

sjogrensresearch-ireland.eu

www.arthritisireland.ie/sjogrens-syndrome

www.irishskin.ie

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Disclaimer Every effort has been made to ensure that the information in this leaflet is accurate before going to print. This information does not replace medical advice, diagnosis or treatment. If you have questions or concerns or need more information as your GP or specialist healthcare provider.