



Sjögren's
FOUNDATION

Medical & Scientific Initiatives

Professional Education & Awareness

- *Sjögren's Quarterly*
- **Awareness Ambassadors**
- **CME Programming**
- **Professional Conferences**
- **National Coalitions, Boards, and Other Groups**
- **International Initiatives and Collaborations**

Foundation Resources

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LIVING WITH Sjögren's

SUMMARY OF MAJOR FINDINGS

Living with Sjögren's was conducted by Harris Poll on behalf of the Sjögren's Foundation, the only non-profit organization focused on increasing research, education and awareness for Sjögren's, an autoimmune disease that affects the entire body. The purpose of the survey was to gain an understanding from adults ages 18 and older who have been diagnosed with Sjögren's about the physical, emotional and financial impact of the disease on their lives.

Sjögren's Survey Patient Demographic Profile

The vast majority (96%) of Sjögren's patients who completed the survey were female. About one-third (33%) of respondents were 60 years of age or younger. On average, respondents said they were diagnosed with Sjögren's over a decade ago (12.3 years mean).



Most Common Symptoms Experienced

The vast majority of Sjögren's patients reported having experienced dry eyes (97%), dry mouth (97%), fatigue (94%), dry or itchy skin (93%), trouble sleeping (91%) and forgetfulness (90%) over the last year. Most said that their dry mouth (92%), dry eyes (92%), and fatigue (80%) symptoms occurred almost weekly or more frequently.

Patients 60 years of age and under said they were more likely than patients over age 60 to experience brain fog (i.e., confusion, forgetfulness, and lack of focus and mental clarity) (66% vs. 53%) and joint pain (67% vs. 62%) almost weekly or more frequently. Patients over 60 years of age reported that they were more likely than patients 60 years of age and under to experience dry nose (65% vs. 59%) and photosensitivity (sunlight) (64% vs. 56%) almost weekly or more frequently.

Half of Sjögren's patients with severe dryness (53%) also have severe fatigue.

LEARN MORE
sjogrens.org

Sjögren's Foundation Clinical Practice Guidelines

Systemic Manifestations in Sjögren's Patients

The Sjögren's Foundation has developed the first U.S. Rheumatology Clinical Practice Guidelines for Sjögren's to ensure quality and consistency of care for the assessment and management of patients by offering recommendations to clinicians for systemic disease management.

Previously, treatment guidelines for serious organ involvement from Sjögren's were borrowed from those used to treat Systemic Lupus Erythematosus (SLE) and Rheumatoid Arthritis (RA). Among the recommendations, the guidelines address the treatment of inflammatory, musculoskeletal pain in systemic Sjögren's, use of biologic agents and management of fatigue.

Rheumatology Guidelines Summary and Recommendations

For the development of the Sjögren's Foundation Rheumatology Guidelines, a highly rigorous and transparent process was employed with important guidance from the American College of Rheumatology and the Institute of Medicine. An extensive, systematic literature review by Topic Review Groups (TRG) was followed by data extraction and drafting of recommendations to be considered by separate consensus expert panels (CEP) consisting of academic and community practice clinicians, registered nurses and patients. Using a modified Delphi-type consensus process, the CEP reached consensus on eighteen recommendations with consensus set at a minimum of 75% agreement.

DMARDs for Musculoskeletal Pain

Recommendations regarding the use of disease-modifying anti-rheumatic drugs (DMARDs) to treat musculoskeletal (MSK) pain were presented as a decision tree with the American College of Rheumatology (ACR) as the first-line therapeutic approach. Although HCQ treatment failed to reach the primary endpoint for pain in a recent, randomized control trial, other studies have shown that following HCQ treatment, Sjögren's patients demonstrated improvement in inflammatory markers and MSK pain. The favorable safety profile of HCQ contributed to the 92% positive agreement of the Rheumatology Working Group. Thus, the recommendation for the use of HCQ received a moderate strength rating and is considered a best clinical practice first-line therapy.

Biological Medications

Biological therapies such as rituximab will become increasingly important in the management of Sjögren's patients and are best used in Sjögren's patients with serious organ manifestations who fail more conservative treatments. There was strong consensus that TNF- α inhibitors not be used to treat sicca symptoms in patients with Sjögren's. This recommendation was qualified by the consideration that clinicians should not withhold TNF- α inhibitor treatment if a patient also suffers from another condition for which such treatment would be indicated.

Fatigue

Fatigue is most effectively managed with self-care measures and exercise. Exercise provides similar benefit to reduce fatigue in Sjögren's patients as was seen for those with RA, SLE or Multiple Sclerosis.

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Patient Education Sheet Muscle and Joint Pain in Sjögren's

The Foundation thanks Alan Boer, MD for authoring this Patient Education Sheet. Dr. Boer is an Associate Professor of Medicine, Director, Jerome L. Green Sjögren's Center, Johns Hopkins University School of Medicine.

Joint and muscle pain in Sjögren's may result from a variety of causes including inflammation, fibromyalgia, age-related osteoarthritis, vitamin D deficiency, hypothyroidism etc. Work with your rheumatologist to identify the specific cause(s) of your pain and find the best treatment regimen for you. Maintain a positive attitude and be an active partner in the management of your pain. The tips below will also help.

- ▶ Become knowledgeable about your medications.
- ▶ Get a good night's sleep
 - Maintain a regular sleep schedule.
 - Set aside an hour before bedtime for relaxation. Listen to soothing music. Consider taking a warm bath before going to bed.
 - Make your bedroom as quiet and comfortable as possible.
 - Avoid caffeine and alcohol late in the day.
 - Avoid long naps during the day.
- ▶ Exercise regularly with the goals of improving your overall fitness and keeping your joints moving, the muscles around your joints strong, and your bones strong and healthy.
 - A physical therapist, occupational therapist, or your health-care provider can prescribe an exercise regimen appropriate for your joint or muscle problem.
 - Start with a few exercises and slowly add more.
 - Make your exercise program enjoyable. Do it with your spouse or a friend. Include recreational activities, such as dancing, walking, and miniature golf.
 - Try different forms of exercise, such as Tai chi, yoga, and water aerobics.
- ▶ Balance rest and activity.
 - Pace yourself during the day, alternating heavy and light activities and taking short breaks to rest.
- ▶ Control your weight.
- ▶ Protect your joints and muscles.
 - Use proper methods for bending, lifting, and reaching.
 - Use assistive devices, such as jar openers, reach extenders, and kitchen and garden tools with large rubber grips that put less stress on affected joints.
- ▶ Use various therapeutic modalities that can relieve joint and muscle pain.
 - Use heat (heating pads, warm shower or bath, paraffin wax) to relax your muscles and relieve joint stiffness.
 - Use cold packs to numb sore joints and muscles and reduce inflammation and swelling of a joint.
 - Consider massage therapy.
 - Practice relaxation techniques, such as guided imagery, prayer, and self-hypnosis.

For more information on Sjögren's, visit the Foundation website at www.sjogrens.org, call 301-530-4420, email info@sjogrens.org, or write to the Sjögren's Foundation, 10701 Parkridge Blvd, Ste 170, Reston, VA 20191

Clinicians: Please make multiple copies of this Patient Education Sheet and distribute to your patients.

Sjögren's Quarterly

The Professionals' Resource on Sjögren's

Vol. 15, Issue 1 – Winter 2020

Do No Harm: Why You Should Avoid Benzalkonium Chloride-Containing Artificial Tears

An editorial by John L. Shells, PhD, FARGO Professor of Biology, Emory University, Grant Sigbee, MD, Michael Christensen, OD, PhD, FARGO Professor and Director of Clinical Research, Southern College of Optometry, Memphis, TN, J. Daniel Nelson, MD, FACS, FARGO Senior Fellow, HealthPartners Institute, Bloomington, MN

Despite many years of basic and clinical research, treatment of dry eye, a multifactorial disease of the ocular surface characterized by a loss of homeostasis of the tear film,¹ remains challenging. While the U.S. Food and Drug Administration (FDA) has approved two prescription drugs which target the inflammatory aspects of dry eye, the mainstay of treatment remains the palliative application of lubricant eye drops, commonly known as artificial tears. The use of artificial tears is included in each step of the treatment recommendations of the recent TFOS-DEWS II Management and Therapy report.²

The concern addressed in this editorial is the continued use of benzalkonium chloride (BAK) as a preservative in "store brand" lubricant eye drops. Step 1 for treatment of dry eye, as recommended in the TFOS-DEWS II Management and Therapy report,² includes the use of "ocular lubricants of various types." Although the toxicity of BAK is addressed in the report, this non-specific recommendation allows wide interpretation by eye care professionals and patients, and as we will show, may lead to use of products preserved with BAK that can cause additional harm to the already compromised ocular surface.

FDA regulations require that multi-dose topical ophthalmic formulations, including prescription drugs, contact lens wetting solutions and lubricant eye drops contain a preservative to prevent contamination by bacteria and fungi. As recently reviewed in the TFOS-DEWS II Iatrogenic Dry Eye report³ and by Walsh and Jones,⁴ BAK is a very effective

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Recap: Sjögren's at the 2019 American College of Rheumatology Annual Meeting

The American College of Rheumatology (ACR) 2019 Annual Meeting, which plays host to an estimated 15,000 attendees, was held November 9-13 in Atlanta, GA. This meeting is one of the largest rheumatology-focused meetings in the world, with an international audience making up a significant portion of attendees.

For the Foundation, this is one of the year's top opportunities to interact, educate and raise awareness among the rheumatology community on the many things happening both at the Sjögren's Foundation and in the Sjögren's community at-large. Staff also use this gathering as an opportunity to meet with industry partners, attend educational sessions and foster new and existing relationships. This article covers some of the highlights from this past year's meeting.

Exhibit Booth

Once again, the Foundation set-up a booth in the exhibit hall to help educate and raise awareness on Sjögren's and how attendees could get involved with our work. This year's display featured a variety of issues of Sjögren's Quarterly, clinical practice guidelines, referral materials for a clinician's office as well as the newly designed guide for parents on the symptoms of Sjögren's in children.

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Town Hall Agenda

- **Clinical Trials Consortium**
- **ICD Coding Change**
- **Clinical Practice Guidelines**
- **Research Funding & Support**

International Clinical Trials Consortium

Why form a Clinical Trials Consortium?

- Patients need a systemic therapy for Sjögren's, and we recognized that industry was not interested in developing one!

Foundation's Response:

- Organized an expert, international group to identify barriers to successful clinical trials and strategize together about how to overcome them.
 - Companies fear entering Sjögren's, because no clear path has been established for FDA approval
 - Critical elements are missing for successful clinical trials



International Clinical Trials Consortium

Actions Taken:

- Launched a dialogue with Industry and Key Opinion Leaders in Sjögren's
- Led international community to develop one set of criteria to identify Sjögren's patients for use in clinical trials
- Forged a relationship with the U.S. Food & Drug Administration (FDA)
- Created an online training program on Sjögren's outcome measures (Sjögren's Training and Education Platform, or STEP)
- Encourage and support initiatives to identify better outcome measures and find new biomarkers



International Clinical Trials Consortium

Next Steps:

- Keep regular discussions going with industry and international leaders in Sjögren's to keep everyone engaged and focused on successful clinical trials
- Continue to focus on key questions in clinical trial design
- Expand STEP to include multiple outcome measures and testing done in Sjögren's
- Take leadership roles on new outcome measures (OMERACT and NECESSITY)
- Partner with the Foundation for the NIH (FNIH) on finding new biomarkers
- Match clinical trial sites with industry needs

Change the ICD Code!

What is the ICD-10 Code?

- A coding system for diagnosis used around the world by healthcare providers

SUCCESS!

- ICD-10 Code changes will take effect in October 2020
- Changes will continue in the next update for the ICD-11 Code

The CDC and American College of Rheumatology (ACR) joined us in this effort.

Change the ICD Code!

Why change the ICD Coding for Sjögren's?

- **There was no dedicated code for Sjögren's!** We currently fall under "Sicca."
 - Sicca is a symptom and not a disease
 - Sicca is not Sjögren's! Sicca simply means dryness and can result from multiple causes
- The current coding is **inaccurate** and doesn't reflect the disease.
- Improper coding **contributes to delayed diagnosis and misdiagnosis.**
- Adding manifestations to the list **opens physicians' eyes** to the fact that these occur in Sjögren's and they need to look for them!
- Proper coding will **help researchers** and **aid in understanding the true prevalence and health-related costs of Sjögren's.**



Change the ICD Code!

- **Sjögren's is now the main heading!**
- **7 new complications were added, with checklist now including:**
 - Inflammatory arthritis
 - Myopathy (muscle pain)
 - Peripheral nervous system involvement
 - Gastrointestinal involvement
 - Lung involvement
 - Tubulo-interstitial nephropathy (kidney disease)
 - Glomerular disease (kidney disease)
 - Central nervous system involvement
 - Vasculitis
 - Ocular involvement (Keratoconjunctivitis or KCS)
 - Dental involvement
 - Other organ involvement

Clinical Practice Guidelines

Why Clinical Practice Guidelines?

- **Patients** need expert, consistent care
- **Physicians** need to know how to best manage and treat patients. They need to understand Sjögren's, take it seriously, and appreciate the potential severity and all the ways it can affect the body
- **Insurance companies** need published guidance for reimbursement
- **Researchers** need research gaps identified
- **Multiple medical specialists** need to recognize Sjögren's

Clinical Practice Guidelines

Why do Guidelines take so long?

- To be used by the medical community, guideline development requires a highly rigorous, well-defined, **complicated**, unbiased, and transparent process.
- This takes many, many hours, over a long period of time, gathering the right people, and is a **huge effort** on the part of Foundation staff and the healthcare providers who **voluntarily** contribute their time.

Clinical Practice Guidelines

Pulmonary Guidelines just completed!

Journal submission contained:

- Guidance on screening, upper and lower airway disorders, interstitial lung disease, and lymphoproliferative disease
- 51 Recommendations with a Clinical Rationale for each
- 5 Tables and 3 Figures to guide the physician
- Online Supplementary Materials with 15 Appendices totaling 128 pages
- 164 References

Clinical Practice Guidelines

Completed Guidelines:

- Fatigue
- Inflammatory Musculoskeletal Pain
- Use of Biologics
- Caries Prevention
- Ocular Management & Treatment
- Pulmonary – *submitted for publication*

Next Steps? Complete Guidelines on:

- Peripheral Neuropathy (incl autonomic nervous system)
- Central Nervous System
- Lymphoma
- Vasculitis
- Gastrointestinal complications
- Gynecological complications
- Caries Restoration; Mucosal Management & Symptom Relief
- Use of Secretagogues; Parotid Gland Swelling

Research Program

To increase interest in Sjögren's research and accelerate scientific and clinical knowledge about Sjögren's, the Foundation:

- Awards Sjögren's Foundation Research Grants
- Recognizes junior investigators in rheumatology by providing Sjögren's Foundation Abstract Travel Awards
- Encourages greater focus on Sjögren's by the NIH through its research and grants
- Brings researchers together to spark enthusiasm and collaboration
- Executes and analyzes surveys that add to our knowledge about Sjögren's and catalyzes greater interest by industry



Future Advocacy & Research Issues

And we are always ready to take action on issues and opportunities as they arise!

For example:

COVID-19 and the many needs of our patients, including addressing the hydroxychloroquine/Plaquenil® shortage for Sjögren's patients



Our Vision

To create a community where patients, healthcare professionals, and researchers, come together to conquer the complexities of Sjogren's!

