

# Sjögren's: the patients' perspective

K.M. Hammitt

*Vice President of Medical and Scientific Affairs, Sjögren's Foundation, and Sjögren's Patient, Reston, VA, USA.*

*Katherine Morland Hammitt, MA*

*Please address correspondence to:*

*Katherine Morland Hammitt,  
Sjögren's Foundation,  
10701 Parkridge Blvd, Ste 170,  
Reston, VA 20191, USA.*

*E-mail: khammitt@sjogrens.org*

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Why is the patient's perspective important? First and foremost, the patient is the ultimate beneficiary of the important work done by clinicians and researchers. A better understanding of Sjögren's patients will lead to greater success in managing patients and finding new treatments. I and millions of Sjögren's patients thank all of those who are working to find better biomarkers, learn more about risks and complications, improve clinical trial endpoints, and engage in basic scientific research that will elucidate the disease process and provide better future therapies.

Patients are grateful to clinicians who want to learn about the difficulty of living with Sjögren's, thereby increasing one's empathy and understanding; about what is most important to patients, so clinical time with patients is well spent and researchers focus on developing therapies that are most meaningful; about how to run clinical trials in a way that will entice more patients to participate; about patients, overall, so we better understand elements such as which symptoms are most prevalent and which are most bothersome, who is more susceptible to specific complications so we are monitored for those, and who is affected by the disease so we can better diagnose Sjögren's and learn to recognise it in populations that were not traditionally diagnosed.

Sjögren's has long taken a back seat to other related diseases such as rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE), but as a Sjögren's patient myself and in a leadership role with a major organisation that represents those with Sjögren's and works with other organisations around the world, we can no longer remain silent about the devastation wrought by this common, yet complex, disease. And, yes, it is common and much more prevalent than most in healthcare recognise. A 2008 study by the U.S. Centers for

Disease Control, National Institutes of Health, and Arthritis Foundation found a range of 0.4 to 3.1 million in the U.S. alone for those with Sjögren's and who did not have another major autoimmune rheumatic disease (1). Compare this to their finding of 161,000-322,000 with SLE and 1.3 million with RA, and Sjögren's is at least tied with RA numbers, if not surpassing it.

We need to work together to increase awareness of Sjögren's. Sjögren's remains under-recognised and misdiagnosed. While recent Sjögren's Foundation efforts were successful in reducing the time to diagnosis from 6 to 2.8 years, too many Sjögren's patients remain undiagnosed and untreated. Patients not yet diagnosed with Sjögren's are being seen in pulmonary, oncology, gastroenterology and neurology clinics and not being referred for diagnosis and treatment of a systemic disease. They are seen for repercussions of dry eye by ophthalmologists, dry mouth by dentists, vaginal dryness by gynaecologists, interstitial cystitis by urologists or family practitioners and not getting a proper diagnosis of Sjögren's.

Some patients suffer from symptoms but are not managed medically by anyone. Fatigue, cognitive dysfunction, musculoskeletal pain, ocular and oral symptoms, and peripheral neuropathies can be dismissed as vague, benign, and/or not seen as part of one disease. Patients and healthcare providers can use different terminology for symptoms. For example, unless patients understand what is meant by "dry eye" or "dry mouth," patients will not describe their symptoms as dryness, and a diagnosis of Sjögren's might be missed. Instead, we need to learn to ask patients if they have frequent ocular or oral pain and/or irritation, difficulty focusing clearly, frequent eye infections, rampant caries or chipping or cracking of teeth, difficulty swallowing without

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liquids, genital pain or pain with intercourse.

We must start doing a better job of recognising Sjögren's in younger patients and in men. When we focus on Sjögren's as a disease of middle-aged and post-menopausal women and one that is marked primarily by dryness, we miss important sectors of our population that might have Sjögren's. We miss the chance to treat patients earlier in the disease process and the ability to learn more about the disease overall, such as how the disease might start, how it progresses over time, and about symptoms and complications with earlier onset.

We must do a better job recognising the many systemic complications of Sjögren's. We cannot label Sjögren's as a disease that is solely one of dryness. While, according to the 2016 Sjögren's Foundation Living with Sjögren's national patient survey conducted by Harris Poll®, dryness is a top complaint and patients do want therapies to alleviate dryness, we also need to recognise the many other symptoms that cause suffering. For example, the same poll shows that 80% suffer from crippling fatigue. Up to 40% of Sjögren's patient will have neurological symptoms that antedate sicca manifestations, and in one recent study of 184 Sjögren's patients, an astounding 93% of patients were diagnosed after neurological symptoms appeared (2). Younger patients are more likely to suffer from chronically swollen salivary glands, musculoskeletal pain, fatigue and neuropathies than dryness. Those with Sjögren's, more than any other autoimmune disease, are susceptible to blood cancers (3). We suffer from cognitive dysfunction or "brain fog." We are susceptible to interstitial lung disease and interstitial nephritis, and mothers with Sjögren's are at higher risk of having babies with foetal heart block. Sjögren's is a disease that can affect any organ or bodily system and interferes with our ability to work, be the parent we wish we could be, socialise, and carry out daily activities.

We cannot remain dismissive of so-called "benign symptoms." Take fa-

tigue: when patients say they are tired, others often are dismissive and do not understand what this means for a patient. The U.S. National Library of Medicine defines fatigue as "a feeling of weariness, tiredness, or lack of energy." This does not begin to describe the fatigue endured by Sjögren's patients. Sjögren's fatigue is not simply a tiredness that can be alleviated by a nap or good night's sleep. It's not a tiredness brought on by working long hours, staying up late to study, or spending hours walking and shopping. It's not marked by laziness or an unwillingness to do things. Instead, we as patients have described our fatigue as overwhelming, engulfing, earthshattering, toxic, bone-tired, draining of life force, and feeling crumpled like a piece of laundry. We describe fatigue in terms of what we cannot do, such as get up out of bed or a chair, lift something, perform a basic household task, drive somewhere such as to the grocery store or a medical appointment, and if and when we can do these things, we are exhausted beyond what words can describe.

Words matter. We ask the broader medical and research communities to think about dropping terms such as "syndrome," "primary" and "secondary." Just because these terms were coined decades ago does not make them useful or even correct. The Sjögren's Foundation is leading a charge on behalf of patients and our medical advisors to change our terminology. "Syndrome" means a constellation of symptoms that tend to run together, while a "disease" is a condition that impairs normal functioning and is distinguished by specific signs and symptoms. Thanks to global research efforts, we have made amazing progress in our understanding of Sjögren's not simply as a group of symptoms but as a disease. While the terms "primary" and "secondary" might be useful shorthand for clinical trial inclusion, these terms become applied broadly. They are not applied to related diseases. Just as one either has or does not have RA or SLE, one either has Sjögren's or does not. In addition, today's clinician is not always familiar

with how to use these terms properly, leading to confusion for patients and clinicians alike. These terms are not helpful in light of what we now know about Sjögren's. Words matter. They especially matter to patients, who feel like they have something that is not as bad as a disease when told they have a "syndrome." Patients feel like their Sjögren's is "secondary" and not as important as their "primary" disease. Words should matter to our healthcare providers, too, and we are grateful to those who acknowledge that what was described 50 years ago may have evolved and differ from what we know today.

Sjögren's is a disease with a high burden of illness and severe impact on quality of life. While a diagnosis often brings a major and sudden transformation to our lives, it's the day-to-day struggles that we face that are most often devastating. Rather than the organ failure or major catastrophic event, the constant struggle without a break wears us down emotionally and physically. We crave validation from the healthcare and research communities as well as our friends and families. Many of our symptoms are hidden from sight, and a lack of acknowledgment denigrates our struggles and makes us feel invisible and misunderstood. While we as patients need to speak up more, tell our stories loudly and clearly, and not be fearful about telling others about our symptoms, it's hard to do this when others are dismissive about our unseen struggles.

As Sir William Osler once said, "If you listen to the patients, they will tell you what is wrong with them."

## References

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