Sjögren's: A Rare Disease with Serious Complications

A Series by:

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Sjögren's is a rare autoimmune disease that affects the entire body with symptoms that range from bothersome to worrisome.

Sjögren's: A Rare Disease with Serious Complications is a recently published series of articles that offers an inside look at the rare disease and one of its potentially serious complications.

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April is Sjögren's Awareness Month

This April, Real World Health Care is honored to shine a spotlight on a rare autoimmune disease that most people have probably never heard about. Sjögren's (SHOW-grins) is a systemic autoimmune disease that affects the entire body. Along with symptoms of extensive dryness of the eyes and mouth, other serious complications include profound fatigue, chronic pain, major organ involvement, neuropathies and lymphomas.

An estimated one to four million people in the United States currently live with Sjögren's, including tennis star Venus Williams. According to the Sjögren's Foundation, nine out of ten Sjögren's patients are women. The average age of diagnosis is late 40s, although it can occur in all age groups, including children, and in both sexes.

"Sjögren's is an incredibly complex disease, and it can be difficult to understand," said Janet Church, president and CEO, Sjögren's Foundation. "That is one of the reasons Sjögren's Foundation encourages patients to share their stories about what it's like to live with the disease."

Coming Together to Conquer Sjögren's

Sjögren's Awareness Month was first established in 1998 when late New York Congresswoman, Louise Slaughter, read it into the Congressional Record. Today, the Sjögren's Foundation works to keep the spirit of national awareness alive every April. This year, the Awareness Month theme is *Coming Together to Conquer Sjögren's*.

Janet Church

Throughout the month of April – and beyond – the Foundation will be highlighting the many Faces of Sjögren's and expanding its #ThisIs Sjögren's online campaign on Facebook, Twitter, and LinkedIn. Every day in April, the Foundation will highlight a different patient and share a glimpse into their life living with the disease. The goal is to not only raise awareness of Sjögren's, but also to share how patients are effectively coping with the disease and to offer advice for others.

"While each daily post will only give a small insight into Sjögren's, by the end of the month, we hope these 30 posts will showcase the complexity of the disease and the strength of our community," said Church.

Supporting the Sjögren's Community Through COVID

People living with Sjögren's have needed extra support throughout the COVID pandemic, said Church. Because many people with the disease take immunosuppressants to help manage their symptoms, they are more susceptible to catching colds, flus and the novel coronavirus than the average public.

"Staying at home and socially isolating are regular facts of life when you are taking immunosuppressants for an immune-deficiency disease like Sjögren's," Church said. "However, during the COVID pandemic, people with Sjögren's haven't been able to physically connect with their normal support systems and loved ones. That lack of in-person support leads to fear and stress, which can compound physical symptoms."

While some with Sjögren's may have considered suspending their use of immunosuppressants to keep their immune systems strong during the pandemic, the Sjögren's Foundation strongly recommends patients not stop the use of any drugs, including immunosuppressants, without first talking with their physicians, as some drugs need explicit steps to be taken when decreasing or stopping medication.

Another COVID-related factor driving fear and stress among Sjögren's patients is the lung complications that can co-occur with the disease, which can occur in up to two-thirds or more of patients. According to Church, patients and health care providers may not be attuned to which pulmonary symptoms are related to their Sjögren's and which may be attributed to COVID.

To help the Sjögren's patient community throughout the COVID pandemic, the Sjögren's Foundation has offered support and guidance through an online FAQ and vaccine position statement, as well as through virtual support groups, online Town Halls, and a virtual National Patient Conference.

"Shifting to online formats for support groups, our national conference and other events has opened up accessibility and allowed us to reach more people," Church said.

The Foundation also has been active in addressing the shortage of Hydroxychloroquine (HCQ), a medication taken by about 40 percent of Sjögren's patients, and which was anecdotally discussed as being effective for the treatment of COVID-19. While HCQ is not an immunosuppressant, it does improve the way Sjögren's patients' immune systems work and thereby improves symptoms. It works similarly in other immune diseases like rheumatoid arthritis and lupus.

"Shortages of HCQ early in the pandemic created havoc among patients, who were worried that they may not be able to access their needed medications," Church said. "We have been working with government groups, insurance commissioners, industry associations and boards of pharmacy to enlighten them about the shortage issue and work toward increasing supply of this critical drug for autoimmune patients."

Clinical Trials and Patient Outcomes

Throughout the COVID pandemic, the Sjögren's Foundation has not lost sight of its commitment to the development of new therapeutics for the disease.

The Foundation encourages patients to participate in clinical trials and acts as a conduit to the pharmaceutical industry to get companies interested and engaged in developing treatments for the entire disease, not just one symptom.

"There is an unmet medical need among Sjögren's patients," Church said. "They are clamoring for treatments to improve their quality of life. We need to make sure their voices are heard when companies are evaluating development priorities and researchers are designing trials."

To that end, the Foundation has connected researchers with Foundation staff and Sjögren's patients to offer insights on trial design, participate in focus groups and interviews, and review marketing materials. These input sessions have addressed issues including clinical trial eligibility, timing and requirements around stopping or modifying current treatments to participate.

"We're also helping researchers develop better outcome measures for their trials," Church added, noting that the standard EULAR Sjögren's Syndrome Disease Activity Index (ESSDAI) used to measure disease activity doesn't address quality-of-life concerns such as dryness and fatigue. "We're pushing for more patient reported outcome measures to reflect how symptoms affect quality of daily life. What does a specific ESSDAI score mean in terms of how the patient feels, whether they could go to the store that week or put in a productive day at work?"

The Foundation also supports research to uncover biomarkers for Sjögren's and has joined the Foundation for the National Institutes of Health (FNIH) biomarkers consortium, a public-private biomedical research partnership that endeavors to discover, develop and seek regulatory approval for biomarkers to support new drug development, preventative medicine and medical diagnostics.

"Diagnosing Sjögren's typically requires a number of objective and subjective tests," Church concluded. "Wouldn't it be great if instead, physicians could administer a simple blood, saliva or tear test that would effectively diagnose the disease as well as its severity and projected course? These questions are the type of innovative, out-of-the-box, and meritorious research projects we want to fund to make real impacts on patient quality of life."

Sjögren's Foundation and **Nancy Carteron, MD, FACR, University of California**

| Jul 7, 2021

Pulmonary Complications in Sjögren's

Lung diseases are a potentially serious complication of Sjögren's and often go unrecognized by health care providers. To help improve the early identification, evaluation and management of Sjögren's-related pulmonary complications, the Sjögren's Foundation led development of a new set of clinical practice guidelines for physicians, rheumatologists and pulmonologists.

Real World Health Care recently discussed the guidelines with two of their key architects: Katherine Morland Hammitt, MA, vice president of Medical and Scientific Affairs, Sjögren's Foundation, and Nancy Carteron, MD, FACR, professor of Medicine and Health Sciences with dual appointments at the University of California – San Francisco and Berkeley. Dr. Carteron also serves as Rheumatology chair of the Sjögren's Foundation's Clinical Practice Guidelines Program, Berkeley Sjogren's Clinic team member and as a board member of the HealthWell Foundation, sponsor of *Real World Health Care*.

An Under-Appreciated, Underlying Threat

Real World Health Care: Why are these guidelines significant? What critical need did they address?

Katherine Hammitt: Pulmonary manifestations are among the biggest complications that can co-occur with Sjögren's. An estimated 16 percent of Sjögren's patients demonstrate pulmonary complications, with increased mortality and lower quality of life – an estimate that may be a bit low, due to gaps in awareness and education among patients and providers: Rheumatologists who treat Sjögren's patients may not know to look for these complications, and pulmonologists treating lung disease may not recognize signs of Sjögren's in their patients.

Katherine M. Hammitt

Sjögren's is a systemic, serious and prevalent disease. Because some physicians haven't been attuned to symptoms that can co-occur between Sjögren's and pulmonary disease, they may have come across to their patients as being dismissive. Now, providers have the tools they need to appropriately recognize, diagnose and treat these manifestations.

These latest guidelines join other clinical practice guidelines we've worked to develop, including guidelines for other systemic manifestations, oral management, and ocular management. We're also developing guidelines on topics including the peripheral nervous system, central nervous system, lymphoma and other blood cancers, and vasculitis.

Dr. Nancy Carteron

Nancy Carteron: Pulmonary involvement in Sjögren's has been under-appreciated by clinicians, leading to delays in diagnosis and management. Also, diagnoses of asthma, GERD and post-nasal drip are made without consideration that Sjögren's could be the underlying driver of the patient's symptoms. These guidelines will improve early identification, evaluation and uniformity of care by primary care clinicians, rheumatologists, pulmonologists and other subspecialists. In addition, the guidelines identified key knowledge gaps and opportunities for research.

It's important to note that from the start, development of the guidelines was conducted in partnership between subspecialists in pulmonology, rheumatology and oncology, along with patients and their family members. This multi-disciplinary approach resulted in guidelines that are highly relevant to patients' clinical care.

RWHC: Why are people with Sjögren's at risk for developing pulmonary disease? Is there anything they can do to help prevent it or lessen its effects?

NC: It's not currently known why. In addition to the disease pathogenesis, which involves genetic risk, triggering events and gender risk, I suspect a key component is the epithelial cell – immune system interphase in the airways and lung air sacs (alveoli).

In terms of prevention and mitigation, people with Sjögren's should avoid smoking, minimize exposure to air pollution and lessen the frequency of respiratory infections. Influenza virus and pneumococcal bacteria vaccines can help prevent respiratory infections.

Spreading the Word to the Sjögren's Community

RWHC: How is the Sjögren's Foundation promoting the new guidelines among health care providers?

KH: The guidelines were published in *CHEST*, a prestigious journal in the field of pulmonary medicine and research. One of the team's members presented an oral abstract on the guidelines at the American Thoracic Society's annual meeting. We've also published articles about the guidelines in Foundation newsletters, including our *Conquering Sjögren's* newsletter for patients and our *Sjögren's Quarterly* publication for the medical and scientific community. Additionally, we've created a 12-page handout summarizing key points and recommendations for management and treatment that we'll continue to distribute broadly.

This fall, we were invited to hold a virtual symposium at the American College of Rheumatology annual conference, where a pulmonologist will highlight recommendations and Dr. Carteron will report on the importance of multiple specialists working together, followed by a live Q & A.

RWHC: What should people with Sjögren's know about the guidelines? How can they use the guidelines to be part of decision-making with their care team?

KH: Patients play a critical role in their care. When they are part of the decision-making process, they are more likely to play a more active role and comply with treatments. We encourage patients to learn about potential pulmonary symptoms so they can watch out for them and report them to their doctor. Patients are encouraged to share the handout with their doctors, to help inform their care as well as the care of other Sjögren's and pulmonary patients.

We also invite patients to learn more and become advocates for themselves and the entire Sjögren's community by taking advantage of our support groups and Town Hall programs and spreading awareness through social media. The Foundation runs an Awareness Ambassadors program designed to help conquer the complexity of Sjögren's as well.

World Sjögren's Day

RWHC: July 23 is World Sjögren's Day. How can people recognize and celebrate this important observance?

KH: World Sjögren's Day honors the birthday of Dr. Henrik Sjögren, the Swedish ophthalmologist who first identified the disease in 1933 in a rheumatology patient who had dry eyes and a dry mouth.

In addition to honoring Dr. Sjögren, World Sjögren's Day gives everyone touched by the disease a vehicle to reach out and educate those close to them about Sjögren's. It is meant to put a face on the millions of people who live with this disease and is the ideal opportunity for people living with Sjögren's to have their voice heard.

Leading up to World Sjögren's Day, and especially on the day itself, we encourage patients to talk about Sjögren's with the people in their life – to share their story and educate others. By doing so, they are helping spread the message that Sjögren's is a serious and complex disease that deserves to be recognized.

The Future of Sjögren's Management

RWHC: What is next on the radar in terms of Sjögren's research and treatment?

NC: Beginning in 2015 with the stewardship of the Sjögren's Foundation, the biopharma industry began to explore drug development for systemic disease in Sjögren's. There are several mature clinical development programs which are poised to potentially lead to an FDA-approved medication for Sjögren's. Similar to drug development in lupus, outcome measures and biomarkers have needed to be developed and continue to be refined. This remains a work in progress.

I believe the most important developments will result from the current revolution in understanding the basic pathways of the innate and adaptive immune system and their regulation in autoimmunity and cancer biology. Various checkpoints in immune pathways are key toggles between autoimmunity and cancer. The risk of one type of cancer, lymphoma, is significantly increased in Sjögren's. Current biotechnology developments are poised to help the scientific and medical community understand the basic biology and thus, why this lymphoma link is so robust in Sjögren's. Further understanding causes of the disease process will shed light on options for mitigation.

There is a lot of optimism for patients and for understanding the mechanisms of the disease at this point in time.