



Updates Slated for ICD-10 Code for Sjögren's!

It is with great excitement to announce that the Sjögren's Foundation (SF), in partnership with the American College of Rheumatology (ACR), has led a successful effort to revise the U.S. ICD-10 code for Sjögren's – a major victory for the Sjögren's community. These changes are expected to be folded into the ICD-11 Code for international use.

This initiative, which began in 2017, was undertaken with the recognition that the existing ICD-10 (International Statistical Classification of Diseases and Related Health Problems, 10th revision) code for Sjögren's (M35.0, Sicca syndrome [Sjogren]) does not represent the disease and contributes to misinformation and confusion. Changes to the code, which are mentioned in detail later in this article, will benefit providers, investigators, researchers, insurers, and, of course, patients, and address key complications of Sjogren's that were not included in the current code.

Rationale for this Initiative

The current code uses "Sicca syndrome" and "Sjögren's" synonymously. While at one point this may have been a prevailing thought, we know that using these terms interchangeably is inaccurate for a variety of reasons.

Sicca is a symptom and not a disease, while Sjögren's is a distinct systemic autoimmune, rheumatic disease that can affect multiple organs and body systems. Dryness certainly occurs in Sjögren's, but dryness alone does not represent the disease and the many other symptoms involved. Furthermore, sicca includes many non-Sjögren's patients who may have dryness symptoms for numerous reasons, including radiation for head and neck cancers, graft-versus-host disease and as a side effect of certain medications.

Further justification can be found when looking at classification criteria. Sjögren's classification criteria has never used either "sicca" nor "sicca syndrome," and no criteria exist for "sicca syndrome." Using the 2002 American European Consensus Criteria (AECC) and the 2016 ACR-EULAR criteria as examples, we can see that these criteria rely primarily on serology and biopsy, and in no way indicate that Sjögren's be defined by sicca.

The current ICD-10 code creates confusion, contributes to misinformation and is potentially detrimental to patients, whose diagnosis carries an incorrect label, which can influence subsequent difficulty with insurance reimbursement, clinicians, who may be unsure of how to best code the disease, study investigators and researchers, who may have trouble accurately identifying Sjögren's patients for clinical trials and data acquisition, and insurers, who grapple with inaccuracy, which can influence reimbursement to both patients and providers.

What's Changing?

The following changes are slated to take effect in October, 2020:

TABULAR MODIFICATIONS

	M35	Other systemic involvement of connective tissue
Revise	M35.0	Sicca syndrome [Sjogren] <u>Sjogren syndrome</u>
Add		Sicca syndrome
Add		Excludes1: Dry mouth, unspecified (R68.2)
Revise	M35.00	Sicca <u>Sjogren</u> syndrome, unspecified
Revise	M35.01	Sicca <u>Sjogren</u> syndrome with keratoconjunctivitis
Revise	M35.02	Sicca <u>Sjogren</u> syndrome with lung involvement
Revise	M35.03	Sicca <u>Sjogren</u> syndrome with myopathy
Revise	M35.04	Sicca <u>Sjogren</u> syndrome with tubulo-interstitial nephropathy
New code	M35.05	Sjogren syndrome with inflammatory arthritis
New code	M35.06	Sjogren syndrome with peripheral nervous system involvement
New code	M35.07	Sjogren syndrome with central nervous system involvement
New code	M35.08	Sjogren syndrome with gastrointestinal involvement
New code	M35.0A	Sjogren syndrome with glomerular disease
New code	M35.0B	Sjogren syndrome with vasculitis
New code	M35.0C	Sjogren syndrome with dental involvement
Revise	M35.09	Sicca <u>Sjogren</u> syndrome with other organ involvement

It is important to note that patients with symptoms of dryness who cannot definitively be linked to Sjogren's, can still be designated under the Symptoms section of the ICD Code.

The Process

From the onset, those involved knew this would be no small task. However, the importance of this initiative helped motivate a team of multi-disciplinary experts, represented by a non-profit, professional society, academia, industry, rheumatology, pediatric rheumatology, primary care, neurology, gastroenterology, pulmonology, nephrology, oncology, ophthalmology and oral medicine, to convene and collaborate on the best course forward.

After a series of meetings and discussions, a proposal was submitted to the ICD-10 Coordination and Maintenance Committee (C&M), a federal interdepartmental committee comprised of representatives from the Centers for Medicare and Medicaid Services (CMS) and the Centers for Disease Control and

Prevention's (CDC) National Center for Health Statistics (NCHS), with suggestions for modifications to the code for Sjögren's.

Excitingly, the proposal was accepted for presentation at the Fall C&M meeting taking place near Baltimore, MD, in September 2018. Here, Dr. Alan Baer represented the SF, ACR and the multi-disciplinary team who informed the proposal by expertly explaining the rationale for why changes to the Sjögren's code are needed. Dr. Baer's presentation was followed by a presentation by a CDC representative detailing the specific tabular changes that were being requested.

A period for public comment took place in the subsequent months after the meeting, during which few questions were raised. However, as the group learned, because questions had been raised, the proposed changes would need to be revised to address the questions and the proposal presented at the Spring C&M meeting in March, 2019.

This time, only a CDC representative was needed for the presentation of the proposal, and Kathy Hammitt, Vice President of Medical and Scientific Affairs, was on hand to provide perspective on the few questions that were raised. An additional public comment period was then held but resulted in no major issues being put forth.

A Team Effort

This effort was no-doubt strengthened by the multidisciplinary team who provided their time and expertise in guiding this project. The Sjögren's Foundation is sincerely grateful to the advisory team, who provided integral help throughout the entire process, and would like to recognize and thank the following individuals:

Alan N. Baer, MD:	Director, Jerome Greene Sjögren's Syndrome Clinic & Professor of Medicine, Johns Hopkins University School of Medicine (Rheumatology)
Theresa Lawrence Ford, MD:	CEO & Medical Director, North Georgia Rheumatology Group (Rheumatology)
Frederick Vivino, MD:	Director, Sjögren's Syndrome Center, Perelman School of Medicine at the University of Pennsylvania (Rheumatology)
Nancy Carteron, MD:	University of California San Francisco (Rheumatology)
Scott Lieberman, MD:	University of Iowa (Pediatric Rheumatology)
Judith Furlong, MD:	ProMedica Physicians Family Medicine (Primary Care)
Julius Birnbaum, MD:	Co-director, Jerome Greene Sjögren's Syndrome Clinic, Johns Hopkins University School of Medicine (Neurology)
Augustine Lee, MD:	Mayo Clinic, Jacksonville, FL (Pulmonology)
Marie Hogan, MD:	Mayo Clinic, Rochester, MN (Nephrology)
Lance Forstot, MD:	Corneal Consultants of Colorado (Ophthalmology)
Vidya Sankar, DMD:	Brigham and Women's Hospital (Oral Medicine)
Richard Ambinder, MD, PhD:	Johns Hopkins University School of Medicine (Oncology)
Katerina Shetler, MD:	Palo Alto Medical Foundation (Gastroenterology)
Jo Annah Jensen:	Novartis (Industry)
Steven Taylor:	Sjogren's Syndrome Foundation
Kathy Hammitt:	Sjogren's Syndrome Foundation
Antanya Chung:	American College of Rheumatology

A special thank you to Alan Baer, MD, for presenting, in-person, at CMS headquarters during the ICD-10 Coordination and Maintenance Committee meeting in September 2018. Dr. Baer's presence and presentation very eloquently made a case for the proposed changes and helped set us apart by being one of the only presenters outside of CMS and CDC staff during the event.

The SF would also like to recognize Antanya Chung and the American College of Rheumatology for their support and collaboration on this important initiative.