



Sjögren's Foundation Update

Conquering Sjögren's Together

Janet E. Church, President & CEO

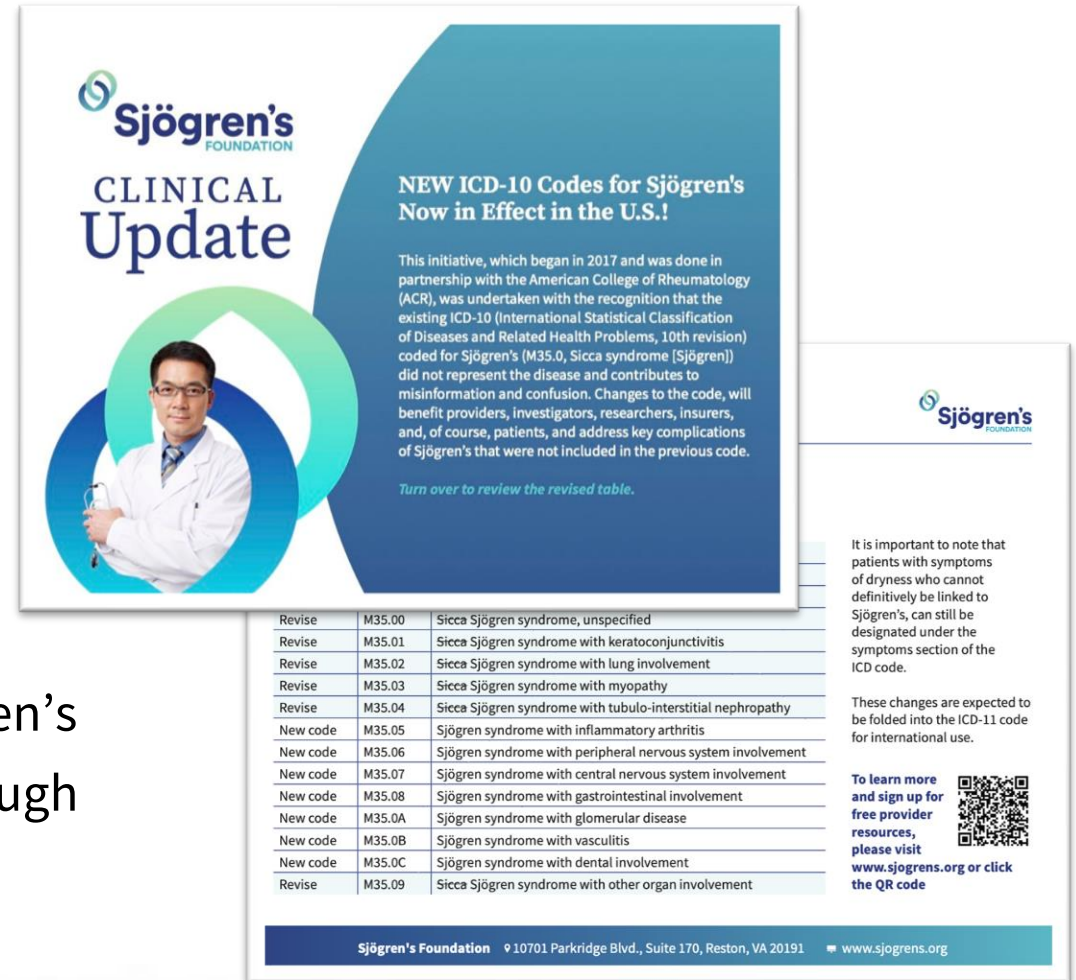
ICD-10 Code Changes and ACR

ICD-10 Code Changes for Sjögren's

- Launched October 1st in U.S.
- Download at Sjogrens.org/resources

ACR November Conference

- Two symposia on Sjögren's
- Many abstracts and posters, and two abstracts that received our Abstract of the Year Award
- Childhood Sjögren's workgroup
- presentation on Pulmonary Complications in Sjögren's
- Foundation Meeting- Understanding Sjögren's through COVID-19 Dysautonomia



Sjögren's FOUNDATION
CLINICAL Update

NEW ICD-10 Codes for Sjögren's Now in Effect in the U.S.!

This initiative, which began in 2017 and was done in partnership with the American College of Rheumatology (ACR), 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 [Sjögren]) did not represent the disease and contributes to misinformation and confusion. Changes to the code, will benefit providers, investigators, researchers, insurers, and, of course, patients, and address key complications of Sjögren's that were not included in the previous code.

Turn over to review the revised table.

Revise	M35.00	Sicca Sjögren syndrome, unspecified
Revise	M35.01	Sicca Sjögren syndrome with keratoconjunctivitis
Revise	M35.02	Sicca Sjögren syndrome with lung involvement
Revise	M35.03	Sicca Sjögren syndrome with myopathy
Revise	M35.04	Sicca Sjögren syndrome with tubulo-interstitial nephropathy
New code	M35.05	Sjögren syndrome with inflammatory arthritis
New code	M35.06	Sjögren syndrome with peripheral nervous system involvement
New code	M35.07	Sjögren syndrome with central nervous system involvement
New code	M35.08	Sjögren syndrome with gastrointestinal involvement
New code	M35.0A	Sjögren syndrome with glomerular disease
New code	M35.0B	Sjögren syndrome with vasculitis
New code	M35.0C	Sjögren syndrome with dental involvement
Revise	M35.09	Sicca Sjögren syndrome with other organ involvement

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

These changes are expected to be folded into the ICD-11 code for international use.

To learn more and sign up for free provider resources, please visit www.sjogrens.org or click the QR code

Sjögren's Foundation 910701 Parkridge Blvd., Suite 170, Reston, VA 20191 www.sjogrens.org

Clinical Practice Guidelines & PRI-MED CME Course

Pulmonary guidelines

- Created with Consensus Panel of 82 people
- Published in *CHEST*
- Published in *Sjögren's Quarterly* and highlighted in *Conquering Sjögren's*
- Downloads available on website

Peripheral Nervous System guidelines

- Working on it now *with* Topic Review Group
- Equal number of Rheumatologists and Neurologists as well as patient representatives

Pri-Med Session February 2022

- *Spotting Sjögren's: Keys to Successful Diagnosis for PCPs.*

Sjögren's Foundation Clinical Practice Guidelines

Pulmonary Manifestations in Sjögren's

Pulmonary complications occur much more frequently in Sjögren's than is often recognized by healthcare providers and are a potentially serious complication of Sjögren's. Clinical practice guidelines for pulmonary manifestations in Sjögren's were developed under the leadership of the Sjögren's Foundation to improve early identification, evaluation and consistency of care by primary care physicians, rheumatologists and pulmonologists. Of note:

- Approximately 15% of Sjögren's patients demonstrate pulmonary complications with increased mortality and lower quality of life. This figure may be low due to gaps in awareness and education.
- In all, 52 recommendations are provided for patient evaluation (by rheumatologists and primary care physicians for pulmonary symptoms and by pulmonologists for potential Sjögren's), airways disorders, interstitial lung disease (ILD) and lymphoproliferative disease.
- As many as 65% of asymptomatic Sjögren's patients will have abnormal pulmonary imaging, highlighting the need for awareness of pulmonary manifestations in Sjögren's.
- A baseline chest x-ray should be considered for all Sjögren's patients, and if concern is high for lung involvement, a high-resolution computed tomography (HRCT) scan may be preferred.
- Airway disorders in Sjögren's are associated with a wide range of symptoms, including a dry nonproductive cough, dry trachea, reflux, vocal cord lesions, bronchiectasis (characterized by narrowed airways) and respiratory inflammation.
- Nearly 40% of Sjögren's patients will have a chronic cough, which should be investigated and the cause identified.
- ILD symptoms can include shortness of breath, cough, sputum production or chest pain. Onset of ILD in Sjögren's may increase with time following diagnosis of Sjögren's.
- Approximately 6% of Sjögren's-associated lymphomas may directly involve the lungs.
- A multidisciplinary approach for pulmonary complications is encouraged and should include a rheumatologist, primary care physician, pulmonologist, pathologist, radiologist, and, when appropriate, an oncologist.

	Strength of Evidence	Strength of Recommendation
Imaging with bronchovascular bundle (BAL) done by case basis and limited to special	LOW	STRONG
Low sensitive spirometry	LOW	STRONG
HRCT in patients with chronic cough	LOW	STRONG
HRCT as surveillance	INSUFFICIENT	STRONG
HRCT in patients with endobronchovascular emphysema	INSUFFICIENT	STRONG

Sjögren's Patient

- With pulmonary symptoms
- Complete PFTs + HRCT
- Normal PFTs and HRCT
- Abnormal PFTs or HRCT
- Manage according to the abnormality detected

Consider alternative etiologies:

- When appropriate, consider testing for possible asthma/obstructive hyperreactivity, or adjunct tests such as fractional exhaled nitric oxide (FeNO) or exhaled nitric oxide (eNO) in patients with chronic cough.
- Consider airway involvement (bronchiectasis). Consider repeating HRCT with expiratory views, if expiratory views were not obtained previously.
- Consider etiologies by patients with dyspnea. Consider echocardiogram or cardiopulmonary exercise testing.
- When cardiac, non-pulmonary disorders. Consider reflux disease, upper airway disorders, anemia, thyroid dysfunction, side-effects of medications, other organ involvement or systemic disorders.

HRCT for any of the following:

1. Restriction
2. Abnormal DLCO
3. Abnormal CXR

Consider HRCT in obstructive defect if concerned about small airway defect.

1 The benefit of obtaining baseline PFTs in asymptomatic Sjögren's patients with regards to long-term outcomes is not clear. The paucity of evidence and potential costs of the test should be taken into account and discussed with individual patients prior to proceeding with PFTs.
Complete PFTs includes spirometry, DLCO, Lung volumes, ideally measured by body plethysmography.
Abbreviations: CXR = chest x-ray; HRCT = high-resolution computed tomography; PFTs = pulmonary function tests; DLCO = diffusing capacity of lung for carbon monoxide.

Active Clinical Trials

15 Companies in Clinical Trials for Sjögren's

- With 21 systemic therapies
- More companies and therapies for Dry eye and Dry Mouth

Many of these companies are part of our Clinical Trials Consortium

- Focus on understanding patients
- Company's have patient panels to contribute to trial process
- Dedicated to making a difference for us

Clinical Trials slowed during Covid/ But Now re-engaging

- Look for trials on our website and sign up
- <https://www.sjogrens.org/living-with-sjogrens/clinical-trials/clinical-trial-locations>

2021 *Living with Sjögren's* Survey

- Survey attracted over 3,600 respondents. THANK YOU!
- Updated, comprehensive insight from the Sjögren's community
- Allow for comparisons and identification of trends and changes
- Insight into new questions – such as Dysautonomia symptoms – since last survey



Biomarker Research in Sjögren's

Biomarkers will make “precision medicine” possible by

- Leading to better and faster diagnoses
- Allowing classification of patients into subgroups (for management & clinical trials)
- Ultimately leading to greater success of clinical trials so we get a new therapy
- Identifying which patients will benefit from which therapies
- Creating a new-age definition of Sjögren's!

Foundation Research Grants Awarded

- 2 Impact Grants and 1 Pilot Grant focus on biomarkers

New 5-Year Biomarker Research Project