



Sjögren's Syndrome: A Clinical Overview

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Overview

- Pathophysiology and epidemiology
- Clinical manifestations and diagnostic criteria
- Treatment approaches and EULAR guidelines
- Disease burden and quality of life impact
- Lifestyle management and dietary recommendations

Epidemiology

- Female:male ratio of 9:1
- Most common in 40s and 50s

Epidemiology Insights (2021)



Gender-specific Prevalent Cases in the US



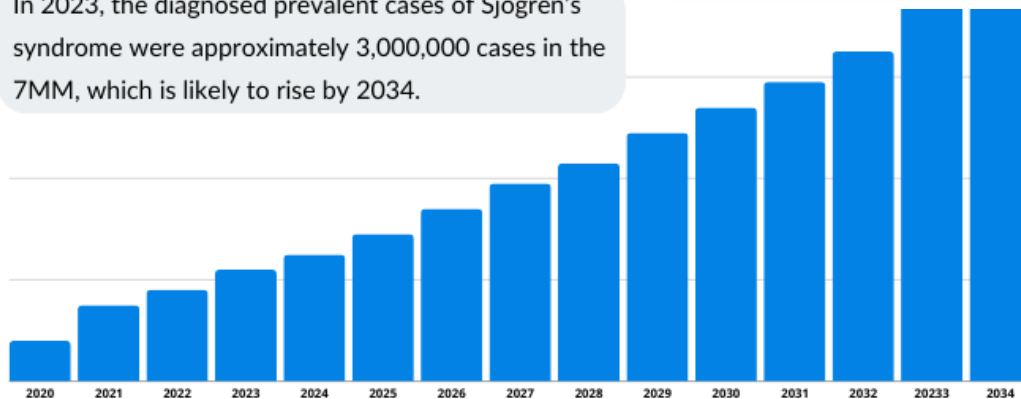
Males
174K



Females
1.5 million

NUMBER OF PATIENTS

In 2023, the diagnosed prevalent cases of Sjogren's syndrome were approximately 3,000,000 cases in the 7MM, which is likely to rise by 2034.



Reyes Jaraba CA, et al. *Revista Colombiana de Reumatología (English Edition)*. 2022;29(4):310-324. <https://www.delveinsight.com/report-store/sjogrens-syndrome-market-size>. Accessed December 24, 2024.

Burden of Disease

- Increased healthcare costs and utilization
- Annual costs rise 27% post-diagnosis
- Higher burden with extraglandular manifestations

What is Sjögren's Syndrome?

- Chronic autoimmune disorder
- The immune system attacks moisture-producing glands
- Primarily affects the eyes and mouth
- Can involve multiple organ systems

Etiology

Genetic Factors

A summary of a comprehensive literature review (1970-2012) found that disease susceptibility was associated with MHC class II alleles, mainly HLA-DR and HLA-DQ.⁶

Different ethnicities have different HLA susceptibilities.⁶

A worldwide meta-analysis found that DQA1*05:01 and DQA1*02:01 carry a significant risk of developing SS, while DQB1*05:01 is protective.⁶

First-degree relatives with SS have a higher relative risk of developing SS and other autoimmune diseases (eg, SLE, RA, T2DM), with siblings at highest risk.⁶

Environmental Factors

Viral infections (eg, EBV, HCV) are thought to promote autoantibody production and lymphocytic proliferation.⁷

Hormonal Factors

Low level of sex steroids is associated with SS.⁶

Estrogen deficiency's role in the development of SS is evident by the predominance of the disease in perimenopausal women.⁶ Low estrogen level is associated with dry eyes.³⁹

Serum DHEA and DHEA-S were found to be reduced in individuals with SS.⁶ Low androgens were associated with dry mouth.³⁹ A randomized, double-blind, controlled trial showed a statistically significant reduction in mouth dryness after 4 months of DHEA therapy.³⁹

Abbreviations: DHEA, dehydroepiandrosterone; DHEA-S, dehydroepiandrosterone sulfate; EBV, Epstein-Barr virus; HCV, hepatitis C virus; HLA, human leukocyte antigen; MHC, major histocompatibility complex; RA, rheumatoid arthritis; SLE, systemic lupus erythematosus; SS, Sjögren syndrome; T2DM, type 2 diabetes mellitus.

Pathophysiology

- **Immune Mechanisms:**
 - Autoimmune attack on exocrine glands.
 - Role of B-cell hyperactivity and T-cell infiltration
- **Key Autoantibodies:**
 - Anti-SSA (Ro) and anti-SSB (La)

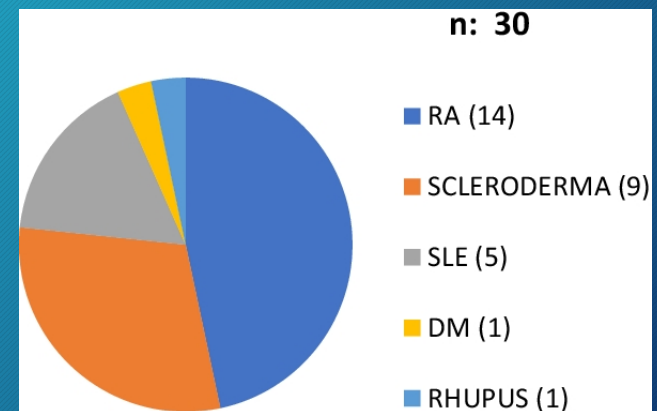
Types of Sjögren's Syndrome

- **Primary Sjögren's Syndrome:**
 - Occurs alone without other autoimmune diseases
- **Secondary Sjögren's Syndrome:**
 - Associated with other autoimmune disorders (e.g., rheumatoid arthritis, lupus)

Risk Factors

- **Age:**
 - 40 or older
- **Gender:**
 - Women are 9-10 times more likely than men
- **Genetic predisposition**
- **Family history:**
 - First-degree relative with an autoimmune disease
- **Presence of other autoimmune diseases**
- **Environmental factors:**
 - Certain environmental triggers, possibly including viral infections, may contribute to the development of the disease

Proportion of Other ARD Developed



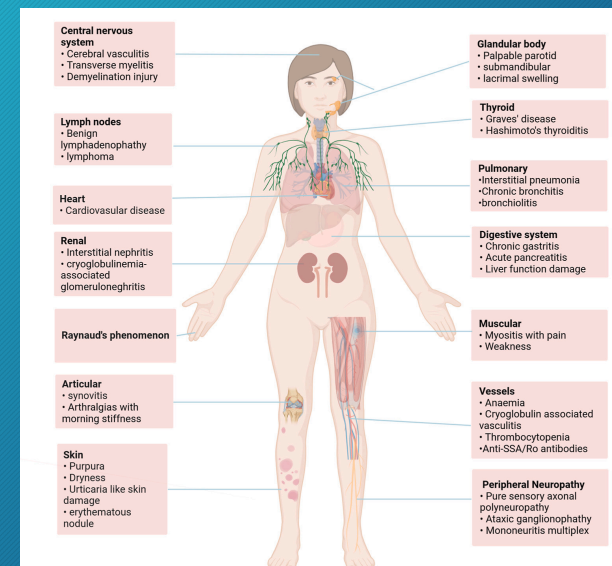
Early Warning Signs

- Persistent dry eyes and mouth (>3 months)
- Difficulty swallowing or speaking
- Frequent dental issues
- Joint pain and fatigue
- Swollen salivary glands

Common Symptoms

- **Classic Symptoms:**
 - Dry eyes
 - Dry mouth
- **Systemic Manifestations:**
 - Fatigue, joint pain, skin dryness
 - Vasculitis, interstitial lung disease, neuropathy
 - Renal involvement

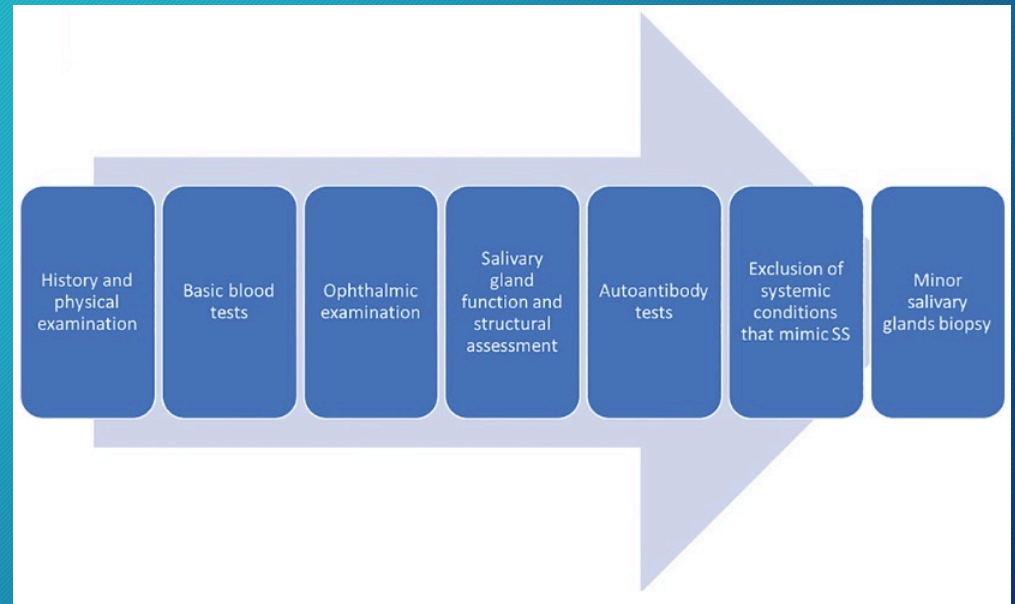
Systemic Complications of Sjögren's Syndrome



<https://www.niams.nih.gov/health-topics/sjogrens-disease>. Accessed December 24, 2024. https://www.physio-pedia.com/Sjogren's_Syndrome. Accessed December 24, 2024. Zhao T, et al. *Heliyon*. 2024;10(17):e36220.

Diagnosis

- **Clinical Evaluation:**
 - Symptom history and physical examination
- **Diagnostic Tests:**
 - Schirmer's test for tear production
 - Salivary gland biopsy
 - Blood tests for ANA, RF, anti-SSA/Ro, anti-SSB/La
- **Classification Criteria:**
 - ACR-EULAR classification criteria

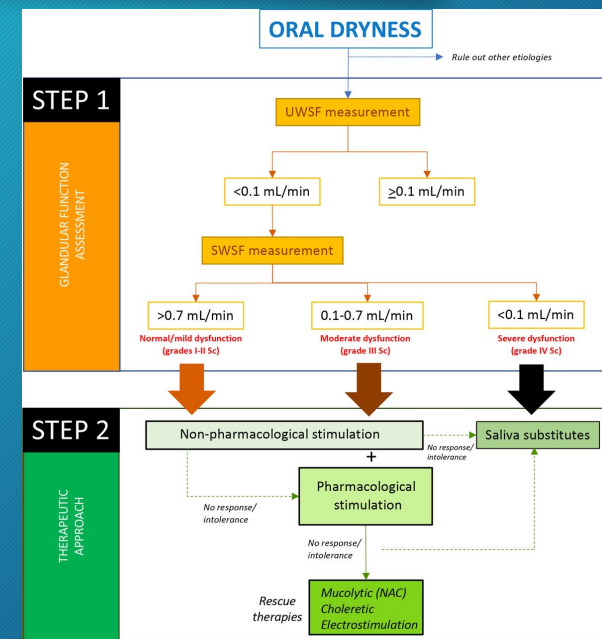


Differential Diagnosis

- Other autoimmune diseases
- Medication-induced sicca symptoms
- Chronic fatigue syndrome
- Fibromyalgia

EULAR Management Guidelines

- Logical sequence of symptom management
- Focus on dryness, fatigue, and pain
- Systemic manifestation treatment
- Regular multidisciplinary follow-ups

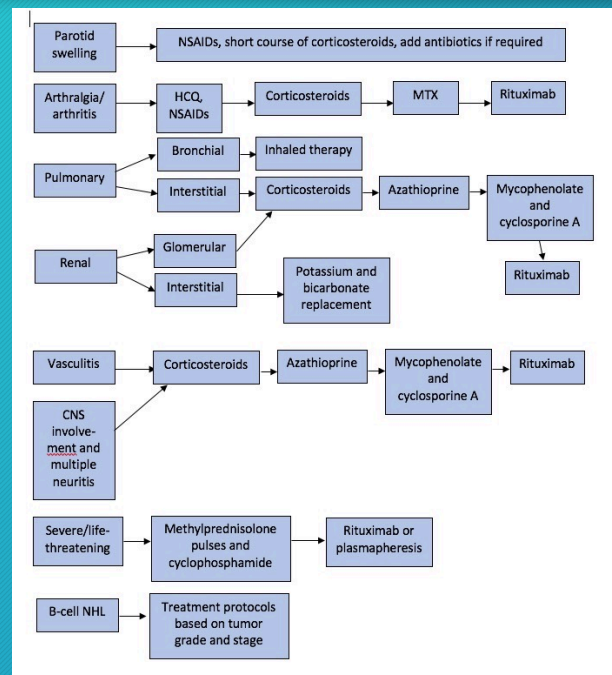


Algorithm of glandular function assessment and therapeutic approach in patients with primary SjS presenting with oral dryness

Treatment and Management

- **Symptom Relief:**
 - Artificial tears and saliva substitutes
 - Pilocarpine or cevimeline to stimulate glandular secretion
- **Systemic Treatments:**
 - Corticosteroids, immunosuppressants (e.g., hydroxychloroquine, methotrexate)
- **Lifestyle Modifications:**
 - Staying hydrated, using humidifiers, regular dental care
- **Multidisciplinary care**

Proposed Therapy for the Main Extraglandular Manifestations



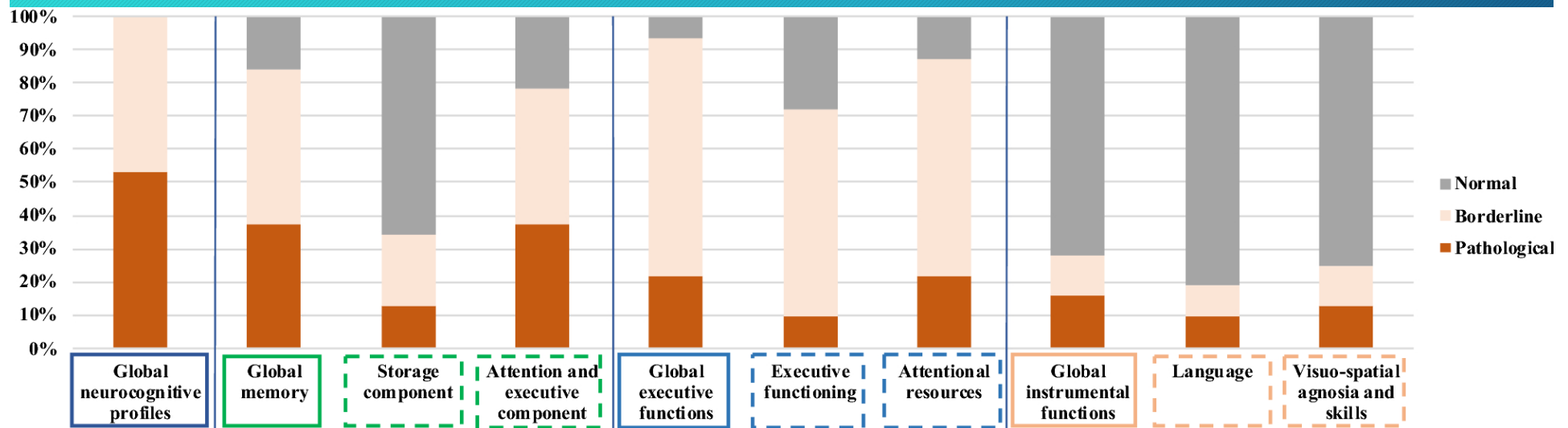
Recommended Pharmacologic and Nonpharmacologic Interventions Used in the Clinical Treatment

Nonpharmacologic interventions for SS: initial approach	
Patient education and behavioral modifications	Avoid factors that trigger fatigue
	Proper sleep hygiene to decrease fatigue
	Air humidification
	Adequate hydration
	Tobacco cessation
Dental caries prevention	Maintenance of good oral hygiene
	Use of fluoride-containing toothpaste
Elimination of anticholinergic drugs	Sicca symptoms are well-known adverse effects
Exercise programs	Aerobic endurance training is effective against fatigue
Pharmacologic interventions: sicca and extraglandular disease	
Dry eyes	Artificial tears or hyaluronate-containing eye drops for mild symptoms
	Topical cyclosporine A, 0.05%, for moderate to severe cases
	If these measures do not provide sufficient relief, the next step is a cholinergic agonist such as pilocarpine or cevimeline
Dry mouth	Salivary substitutes and/or salivary stimulation
	If these measures do not provide sufficient relief, the next step is a cholinergic agonist such as pilocarpine or cevimeline
General systemic symptoms	Best managed with hydroxychloroquine
Extraglandular manifestations	Organ-specific treatment, mainly corticosteroids and immunosuppressive medications

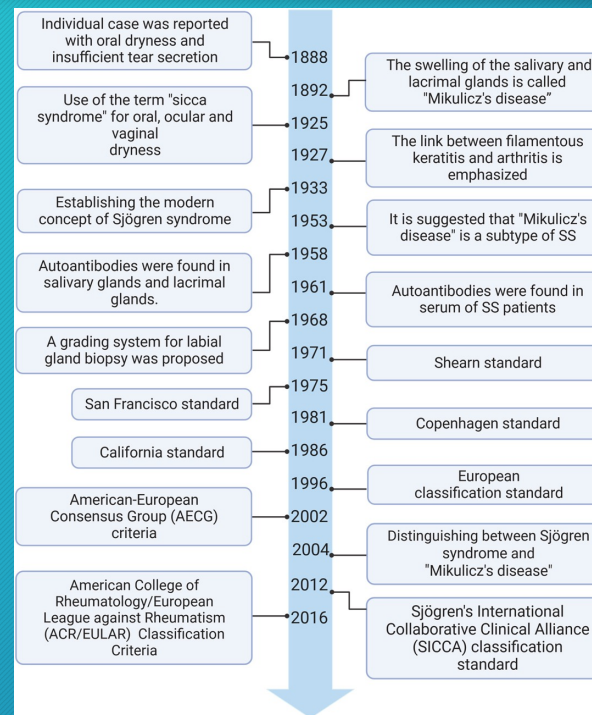
Quality of Life Impact

- Reduced health-related quality of life
- Lower SF-36 and WHOQOL-BREF scores
- Decreased employment rates
- Increased work disability

Interplay Between Cognition, Depression, Anxiety, and Sleep in Primary Sjogren's Syndrome Patients



Milestones in the History of Research



Future Directions

- Ongoing clinical trials
 - Novel therapies targeting B-cell activity (e.g., rituximab)
- Emerging biomarkers for early diagnosis
- Advances in understanding genetic predisposition

Key Takeaways

- Complex autoimmune disorder
- Multisystem involvement
- Significant impact on quality of life
- Importance of early diagnosis and multidisciplinary care

Thank You

Appendix

Lifestyle Management

- Regular use of artificial tears
- Increased fluid intake
- Good oral hygiene
- Humidifier use
- Smoking cessation
- Regular, gentle exercise

Dietary Recommendations

- Anti-inflammatory foods (berries, leafy greens)
- Fiber-rich foods
- Adequate hydration

Complications

- Dental issues:
 - Tooth decay and oral infection
- Eye infections or corneal damage
- Increased risk of lymphoma
Organ involvement (lungs, kidneys, liver)