

# HOSPITAL PHYSICIAN®

## RHEUMATOLOGY BOARD REVIEW MANUAL

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## Sjögren's Syndrome

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### Sjögren's Syndrome

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#### INTRODUCTION

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Sjögren's syndrome is a chronic systemic inflammatory disorder characterized by dry eyes and dry mouth. It is also known as Mikulicz's disease, Gougerot's syndrome, sicca syndrome, and autoimmune exocrinopathy. In the late 1800s, Mikulicz described a patient with bilateral enlargement of the lacrimal and salivary glands with significant round cell inflammation. In 1933 Henrik Sjögren wrote his monograph associating dry eyes with arthritis. In 1953 Castleman and Moore described the histopathology of the exocrine glands and made the association that Mikulicz's disease and Sjögren's syndrome were identical.

Sjögren's syndrome is one of the most common autoimmune diseases, second only to rheumatoid arthritis in prevalence (0.1% to 1%). It is estimated that 1 to 2 million people have Sjögren's syndrome, but most are undiagnosed. The full spectrum of complications is often not appreciated in this disease. The oral and ocular symptoms of Sjögren's result from lymphocytic infiltration of the exocrine glands. In addition to decreased salivary and lacrimal gland activity, patients may have extraglandular manifestations, which commonly include constitutional symptoms, joint pain, and Raynaud's phenomenon. This manual reviews the diagnosis and management of primary and secondary Sjögren's syndrome.

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#### PRIMARY SJÖGREN'S SYNDROME

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##### INITIAL PRESENTATION

A 43-year-old white woman is referred to a rheumatologist by her ophthalmologist for evaluation of dry eyes and a complaint of arthritis. As part of the evaluation of the patient, the ophthalmologist ordered an

antinuclear antibody (ANA) test; the results of the test were positive.

##### HISTORY

The patient has no significant past medical history other than recurrent urinary tract infections that have not responded to multiple courses of antibiotics. She has had fatigue and arthralgias for a number of years, which she relates to stress. The patient reports having dry eyes with a gritty sensation and some photophobia. She cannot eat a cracker without drinking fluid simultaneously and occasionally has dysphagia. She denies chest pain or shortness of breath. She reports having nasal congestion but denies abdominal pain, nausea, vomiting, fever, or chills. She has no muscle weakness but does complain of morning stiffness, joint pain, and swelling. She has not noticed any joint deformity. She reports noticing swollen, tender parotid glands a few times per year, which she attributes to infection. The swelling and pain improve after a few weeks. She has experienced Raynaud's phenomenon for more than 6 years. She denies oral ulcers, headaches, or alopecia. There is no history of malar rash, photosensitivity, headaches, or seizures. She has no history of blood clots. She complains of vaginal dryness and multiple candida infections. She says that she has trouble sleeping and has nonrestorative sleep.

The patient reports no past surgeries. She takes no oral medications currently but requires eye drops due to dry eyes. The patient is not taking any over-the-counter medications known to cause decreased secretions. She has no allergies to any medications. She does not drink alcohol, smoke tobacco, or use illicit drugs. There is no significant family history.

##### PHYSICAL EXAMINATION

The patient appears tired on physical examination. Vital signs include blood pressure of 120/80 mm Hg, pulse rate of 85 bpm, respiratory rate of 14 breaths/min,