

Sjögren's Syndrome: Challenges to Management

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Sjögren's syndrome is among the most common autoimmune diseases in the United States, affecting an estimated 1% of the population.¹ It is characterized by dry eyes and a dry mouth, which result from an autoimmune process in which mononuclear inflammatory cells invade the lacrimal and salivary glands. Sjögren's syndrome may have a substantial impact on a patient's quality of life. Dry eyes are uncomfortable, may be painful, cause blurred vision, and require the frequent installation of tear substitutes. Many patients will need to put artificial tear drops in their eyes every hour while awake, and sometimes even more frequently. A dry mouth makes it difficult to eat, increases dental decay, and many patients must carry water with them at all times.

Sjögren's syndrome may exist on its own, in which case it is termed primary Sjögren's syndrome, or in association with another connective tissue disease, in which case it is termed secondary Sjögren's syndrome. The 2 most frequent connective tissue diseases associated with secondary Sjögren's syndrome are rheumatoid arthritis and systemic lupus erythematosus. An estimated 10% to 15% of patients with rheumatoid arthritis and an estimated 20% of patients with lupus will have secondary Sjögren's syndrome.^{2,3} Among patients with primary Sjögren's syndrome, there is a subset who have systemic features including hypergammaglobulinemia, vasculitis, and neurologic disease.⁴

The diagnosis of Sjögren's syndrome often is difficult. The symptoms are non-specific and may be attributed to other causes. The symptoms of a dry eye may be caused by several other conditions, including contact lens overwear, blepharconjunctivitis as a result of meibomian gland dysfunction or ocular rosacea, aging changes resulting in aqueous tear deficiency, or ocular allergy, and the problem may be diagnosed incorrectly, or it may be multifactorial in nature (eg, dry eyes resulting in contact lens intolerance). The symptoms of dry mouth may be overlooked or attributed to other causes, such as medications (eg, diuretics, antihistamines, etc). Further complicating the diagnosis is the fact that the organ involvement may be asymmetric. Patients may have a more advanced ocular or oral disease and not meet the criteria for diagnosis until the disease has been present for several years. As such, the correct diagnosis requires referral to several specialists, including ophthalmology, oral medicine, and rheumatology.

Sjögren's syndrome is a chronic disorder requiring long-term management. A multidisciplinary approach is needed. Treatments that are initially effective may become less effective over time as the disease progresses, or complications may arise. Although artificial tear substitutes may be effective initially, allergy to preservatives may result with frequent use, resulting in the need for preservative-free

artificial tear substitutes. If tears are insufficient, punctal occlusion may be needed. Secondary meibomian gland dysfunction may develop, requiring management as well. Rarely, patients may develop a corneal melting disorder, which is a serious condition that will result in loss of vision unless treated appropriately. The oral component results in increased dental caries and may be complicated by oral candidiasis. The systemic features of the disorder, such as polyarthralgias, fatigue, and fibromyalgia require management by a rheumatologist. The less common, but more severe features, such as vasculitis, will require aggressive management with immunosuppressive drugs in order to preserve organ function and possibly life. Finally, vigilance is required because of the increased rate of lymphoma seen among these patients.⁵

The features of this disorder and the issues involved in its management are outlined in the accompanying review article

and interview. Appropriate referral and a multidisciplinary team approach are required for the care of patients with Sjögren's syndrome.

...REFERENCES ...

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