

Sjogren's Syndrome

Sjogren's syndrome (SS) is an autoimmune disorder affecting primarily the lacrimal (tear) and salivary glands. Thus, the main associated symptoms are dry eyes and dry mouth (sicca complex). Affected persons may complain of eye dryness, a gritty sensation, and frequent eye irritation. Oral symptoms include difficulty swallowing, hoarseness, and parotid gland swelling. The exact frequency of SS is unknown although most studies suggest a prevalence of about 0.6% of population. Because the symptoms are often not severe, and may not be viewed by patients as significant (and therefore not worth mentioning), many patients remain undiagnosed. SS may occur by itself (primary SS) or in association with another autoimmune condition such as RA, lupus, inflammatory myositis, and scleroderma (secondary SS). Other autoimmune associations include autoimmune hepatitis and primary biliary cirrhosis.

The etiology of SS is unknown but fundamentally involves infiltration of glands by lymphocytes resulting in dysfunction and damage. Other organs that may be involved (occurring more frequently in primary SS) include the lungs, kidneys, gut, and nervous system. Lung involvement includes infiltrates, bronchiolitis, and pulmonary fibrosis. Renal involvement usually takes the form of renal tubular acidosis. Gut involvement may be associated with dyspepsia, diarrhea, and malabsorption. An increased incident of lymphoma, particularly of the MALT cell variety, occurs in Sjogren's patients.

In patients suspected of having the condition, various tests may be performed to help confirm the diagnosis. These include: 1) measurement of tear formation with a strip of filter paper (Schirmer test); 2) slit lamp examination with Rose Bengal staining; 3) blood tests including anti-SSA(Ro) and anti-SSB(La); 4) lip biopsy with microscopic examination of the minor salivary glands for lymphocyte infiltration and damage. In some patients who have been found to have a positive ANA, additional workup reveals a diagnosis of SS as the reason for the ANA.

For patients with only dry eyes, management usually consists of artificial tears (but not products containing astringents), gels, or salves. Cyclosporine (Restasis) eye drops may be preferred by some patients. Persistent symptoms may justify the placement of punctal plugs (or permanent closure of the puncta) to hold tears in the eyes. Some patients may benefit from special glasses ("greenhouse" glasses) which hold more humidity near the eyes. For mouth dryness, simple interventions include frequent use of liquids, sugarless gum, or lemon drops. Oral pilocarpine (Salagen) or cevimeline (Evxac) increase salivary flow to some extent. Regular dental care is advisable because of increased tooth decay. Medications used in other autoimmune conditions (hydroxychloroquine, prednisone, methotrexate, azathioprine, and rituximab) have had only limited success in SS.

The treatment of SS is not completely satisfactory. Patients are at risk for corneal ulceration, oral candidiasis, and dental caries. Reports indicate that quality of life is reduced for persons with SS because of the symptoms previously described, but also because of frequent fatigue, arthralgias, and myalgias. Primary care physicians and other health care providers should be aware that SS is a fairly common syndrome, especially in an aging population, and that most patients will have some improvement in their symptoms if treated.

About the Author

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Dr. Halverson is a Professor of Medicine at the Medical College of Wisconsin. He is currently involved in patient care and teaching fellows, residents, and medical students and physician assistant students at St. Joseph's Hospital and Froedtert Hospital. He has been a volunteer with the Wisconsin Chapter of the Arthritis Foundation. He has been a member of the Board of Directors since 1984 and also served as Chair of the Board and Chair of Health Promotions.

He and his wife, Gloria, also an Medical College of Wisconsin faculty member, spend time each year teaching overseas.

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