

**Bloch K J, Buchanan W W, Wohl M J & Bunim J J.** Sjögren's syndrome: a clinical, pathological, and serological study of sixty-two cases. *Medicine* 44:187-231, 1965.  
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This multidisciplinary study provided evidence for the distinction between secondary Sjögren's syndrome (SS) in which keratoconjunctivitis sicca (KCS), salivary gland enlargement, and xerostomia were accompanied by a major connective tissue disease and primary SS in which KCS and xerostomia constituted the major manifestation of a systemic disorder. [The SCI® indicates that this article has been cited in more than 565 publications.]

### Sjögren's Connections

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During my internship at Bellevue Hospital in New York in 1955, I discovered an old copy of the *New England Journal of Medicine* that a predecessor had left on the ward. It included an article by Winfred S. Morgan entitled "The probable systemic nature of Mikulicz's disease and its relation to Sjögren's syndrome."<sup>1</sup> I was fascinated by the title, but considered the article to represent the extreme of medical esoterica.

Some years later, when, as a clinical associate at the National Institutes of Health (NIH), it was my turn to prepare Journal Club, I returned to the article as part of my presentation on Sjögren's syndrome (SS). Joseph J. Bunim, clinical director, National Institute of Arthritis and Metabolic Diseases, was impressed by the relative lack of interest in SS in the US. He initiated a multidisciplinary study of SS and guided the re-search of this entity. Patients with SS were referred to the NIH for study from throughout the US; 62 of these patients

constitute the subject of the review in *Medicine*.

Henrik Sjögren, a Swedish ophthalmologist, had recognized that an unusual disease of the anterior surface of the eye, keratoconjunctivitis sicca (KCS), was attributable to a failure of secretion of the lacrimal glands and that it was accompanied by xerostomia, parotid swelling, and arthritis, most likely rheumatoid arthritis (RA).<sup>2</sup>

Our 62 cases were assigned to five clinical groups: Group A consisted of 30 patients with SS and RA; group B, 2 patients with SS and "probable" RA; group C, 3 patients with SS and progressive systemic sclerosis; group D, 4 patients with SS and polymyositis; and group E, 23 in whom KCS and xerostomia represented the major manifestations of a systemic disorder. The entire group was characterized by a high frequency of hypergammaglobulinemia, rheumatoid factor, and antinuclear antibodies, and by precipitating antibodies to tissue antigens that would now be identified as SSA/Ro and SSB/La.<sup>3</sup> These serologic findings were especially pronounced in group E, which was clinically characterized by the frequent occurrence of marked enlargement of salivary glands, pulmonary disease, purpura, neuropathy, hyposthenuria, and lymphoma, possibly related to earlier radiation treatment of enlarged parotid glands.

The series of papers reporting the NIH experience with Sjögren's syndrome led to a surge of interest in this entity. Hundreds of publications have appeared, and we seem to be close to defining the etiology of this complex disorder.<sup>5</sup> It was a great honor to join Sjögren in a 1971 review of the syndrome that bears his name.<sup>6</sup>

1. Morgan W S. The probable systemic nature of Mikulicz's disease and its relation to Sjögren's syndrome. *N. Engl. J. Med.* 251:5-10, 1954. (Cited 125 times.)
2. Sjögren H. A new conception of keratoconjunctivitis sicca (*Keratitis filiformis in hypofunction of the lacrimal glands*). (Translation by Hamilton J B.) Sydney, Australia: Australasian Medical, 1943.
3. Tan E M. Antinuclear antibodies: diagnostic markers for autoimmune diseases and probes for cell biology. *Advan. Immunol.* 44:93-151, 1989.
4. Fox R I, Howell F V, Bone R C & Michelson P. Primary Sjögren's syndrome: clinical and immunopathological features. *Semin. Arthritis Rheum.* 14:77-105, 1984.
5. Garry R F, Fermin C D, Hart D J, Alexander S S, Donehower L A & Luo-Zhang H. Detection of a human intracisternal  $\alpha$ -type retroviral particle antigenically related to HIV. *Science* 250:1127-9, 1990.
6. Sjögren H & Bloch K J. Keratoconjunctivitis sicca and the Sjögren syndrome. *Surv. Ophthalmol.* 16:145-59, 1971.