A Note from Sweden: Recollection of Henrik Sjögren

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Abstract

In 1933 the Swedish ophthalmologist Henrik Sjögren defended his doctoral thesis at the Karolinska Institute in Stockholm. He obtained a mediocre evaluation for his thesis and had to leave academia, but continued to publish from his outpost in Jönköping. Ultimately he got well-deserved international recognition for his work, after a study on "Sjögren's syndrome" was subsequently published in 1954 by W. Morgan in *New England Journal of Medicine*.

Keywords

Sjögren's syndrome • Henrik Sjögren • Sweden • Recollection of Sjögren's original patients • Aerobic exercise and fatigue

2.1 Introduction

Henrik Sjögren was born in Köping in central Sweden in 1899 and died in Lund in September of 1986, only months after being honorary president together with Jan Waldenström at the First International Sjögren's Syndrome meeting held outside Copenhagen earlier that year. He was a soft-spoken almost shy gentleman who never advertised himself and never complained of the harsh and unjustified critique he received when defending his thesis in 1933. Figure 2.1 is based on his patient data and drawn by Frank A. Wollheim [1]. When interviewed in connection with the 1986 meeting, he regretted that due to illness he was unable to attend in person. He wished the participants a successful meeting and said that he was particularly thrilled that one had now created "Sjögren's mice." The opening of the meeting featured a valse for piano that he had composed for his fiancée, and later wife, Maria Hellgren, in the early 1920s. Although he was an ophthalmologist and not a rheumatologist, it is fair to state that he made Swedish rheumatology world famous. Henrik Sjögren was an honorary member of the American Rheumatism Association and of the Swedish Society for Rheumatology. Before the advent of the Nazi regime in Germany, a large proportion of academic papers in Sweden were published in German language. This was also true of Sjögren's thesis. In 1943 the Australian ophthalmologist Bruce Hamilton approached Sjögren and

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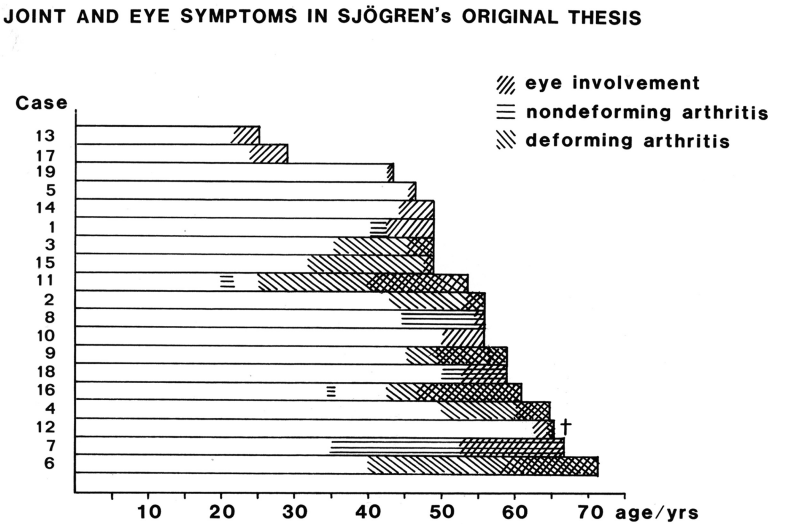

Fig. 2.1 Age, symptoms and disease course in the 19 first patients studied by Henrik Sjögren

obtained permission to translate it into English. This translation obtained much wider attention than the original German version and made Henrik Sjögren well-recognized internationally.

2.2 Prevalence and Incidence of Sjögren's Syndrome in Scandinavia

How prevalent is Sjögren's syndrome in Scandinavia? We have current data from two areas. One is the city of Malmö, with an adult population of approximately 200,000 inhabitants. Here we (ET) have identified 340 cases among individuals above the age of 18 years corresponding to a prevalence of 0.15% (E. Theander, unpublished personal communication). All these individuals have an established diagnosis based on the American–European Consensus Criteria from 2002 [1]. This would be similar to recently proposed prevalence estimates from Turkey [2], Greece [3], and Great Britain [4] using the same criteria set. The incidence is more difficult to estimate. We have encountered between four and seven new cases per year in the period starting in 2002. A rough estimate based on the Malmö experience would give an estimated average annual incidence of 2.5 new cases per 100,000 individuals per year or 1 in 40,000. It is likely that not all new cases come to our hospital unit; some may be cared for by their primary care physicians or escape health care or stay unsymptomatic. An earlier incidence estimate published for the population of Minnesota was 3.9 per 100,000 [5].

In Norway, a recent survey was performed in two health districts around the cities of Stavanger and Bergen. In this population of 852,342 individuals served by two university hospitals and one general rheumatism hospital, the Norwegian investigators found 431 patients with a diagnosis of primary Sjögren's syndrome registered at these hospitals, private rheumatologists, and in a salivary gland biopsy registry. This corresponds to an estimated prevalence of only 0.05%. There were no incidence data in this study [6].

The incidence and prevalence of secondary Sjögren's syndrome in Scandinavia has not been investigated to our knowledge and would be rather difficult to study.

2.3 Any Special Features in Swedish Patients with Sjögren's?

Several investigators in Spain and Greece have reported high prevalence of cryoglobulinemia in their patients. This has not been the experience in

Malmö, where only traces of cryoglobulin were found in a minority of 7% of patients, although active search and careful lege artis sampling was performed (E. Theander, unpublished personal communication). Neither have we found hepatitis C to be increased among the patients. It should be mentioned that the occurrence of hepatitis C is low in Sweden. With regard to the development of non-Hodgkin's lymphoma (NHL) in primary Sjögren's syndrome, we have detected differences in subtype of NHLs compared to other European countries. In Sweden we detect a high number of diffuse large B-cell lymphomas in addition to the commonly found MALT lymphomas [7, 8]. The difference may be mainly due to the assessment of risks by using health-care registries in contrast to cohort observations in some other studies. Otherwise we have not identified any special clinical features distinguishing Swedish patients.

2.4 Pearls of Wisdom

Fatigue and impaired physical capacity are common features in Sjögren's syndrome influencing an individual's quality of life. Therefore, we want to call attention to the successful application of intensive aerobic training reported from our unit in Malmö in 2007 [9]. We have recently performed a 4-year follow-up of the results of this controlled aerobic exercise program involving Nordic walking. The initial training consisted of 12 weeks of 45 min walking three times each week. Retesting at follow-up showed sustained higher physical activity and aerobic capacity in the treatment group and borderline less fatigue. Pain, anxiety, and depression did not differ between the groups [10]. The increased physical activity could have positive effects also on comorbidities such as cardiovascular health.

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