

# Sjögren's: the European patients' community perspective

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Sjögren's is a disease with diverse clinical expression, prognosis and a multidimensional complexity: genetically heterogeneous population; diverse spectrum of disease; complex and nuanced phenotypes; considerable overlapping; numerous potential environmental factors; disease flares, etc which are unable to be related and resolved by most clinicians. Clinicians may have expertise, knowledge, experience, intuition, empathy and caring. They may be aware of subtle patterns amongst complex ones, be able to see unusual events below critical frequency, retain variety, and integrate and identify disease trajectory over time, but working within the constraints of stretched health care systems is challenging.

Diagnosing, treating, following up and researching Sjögren's remains a significant challenge (1) which is equalled by the lack of acknowledgment and recognition that it is a devastating disease, affecting patients physically, mentally, emotionally, socially, and financially. Quality of life is dramatically impaired. It is difficult and frequently impossible for others to understand how patients feel, how they must lead their lives with a disease lacking effective and specific treatment that has them living daily with invisibility, unpredictability, and unimaginable suffering (2).

Although Sjögren's is isolating for sufferers, the wider European patient community seems to be aligned when identifying the numerous unmet needs. They clearly state that a huge and borderless lack of knowledge and understanding prevails and seems to persistently fuel misunderstandings, misdiagnosis and/or delays not only in diagnosis but also in access to treatment and care.

Inadequate or non-existent quality knowledge and information about the disease, especially at primary care lev-

el, is a barrier to early diagnosis resulting in delayed referral to rheumatology and demonstrates the need for greater recognition of the disease among the healthcare community in general. The journey to reach a diagnosis is still an odyssey in many countries as patients find difficulties in being understood and end up walking on their own, and at their own cost, on a pilgrimage between medical specialities, trying to find answers to the different symptoms that they do not know how to manage. The insufficient number of rheumatologists, in particular, those with specific knowledge of Sjögren's, is a reality we cannot change in the short term. However, raising awareness and educating GPs and HCPs, providing more and clearer information on the disease itself and a clear step-by-step plan for diagnosis and follow-up will certainly ensure the ability to provide optimal support to patients.

The lack of understanding of the systemic nature of Sjögren's and the lack of appreciation of its heterogeneous nature frequently leads to poor advice, inappropriate care, and underestimated burden. Patients are often dismissed because doctors cannot relate to the symptoms or are not taken seriously and labelled as anxious - many reported symptoms are still hung on the 'mental hook'. Lack of understanding from family, friends, co-workers, and employers leads to a heavy emotional burden fuelled by relationship difficulties, not to mention poor disease outcomes. HCPs need more reliable information to address the widespread lack of knowledge about, and promote a better understanding of, the complexities of the disease and to ensure that they provide optimal support to patients.

Ultimately, the classification of Sjögren's as a syndrome may contribute to underestimating what it actually is: a disease.

The patients' journey and experiences during their care urgently need to be improved. Facilitating access and signposting patients to a multidisciplinary team (MDT) is a 'must' as is active and mutually fruitful collaboration between doctors. A key role of HCPs is to enable patients to take a more active role in their healthcare, so it is important to introduce them to all members of the MDT involved in aspects of their disease (ophthalmology, dentistry, neurology, urology, gynaecology, etc). This will contribute to identifying many other underlying conditions associated with Sjögren's that are only now being recognised and address symptoms that are often overlooked, such as neuropathic, urinary, bowel, digestive and sleep disturbances. It will also enable sharing of expertise and increasing knowledge of Sjögren's among the different medical disciplines. Defining and disseminating information about disease complications, including lymphoproliferative diseases, is essential so that patients are duly monitored, enabling early detection, and timely referral to appropriate specialists.

Educating patients, caregivers and public is essential to fight widespread disease stigma. Because Sjögren's is typically considered a condition of middle-aged women, men and young people may be underdiagnosed and, consequently the disease may become more severe. Frequently, those not fitting the standard image of a Sjögren's patient find it difficult having their specific issues addressed and acknowledged and struggle to accept the diagnosis. These patients do not adhere to treatment and care, living the disease journey alone with all the consequences this brings at various levels.

Fatigue and cognitive dysfunction cause the greatest patient reported disability and remain a massive unmet need, poorly addressed and validated. This need fails to be recognised when applying for disability. Furthermore, this fatigue results in lower work output and relates directly to difficulties in reconciling work and Sjögren's, specifically when recognizing patients' needs either for work adaptations or validation of Sjögren's as a cause of disabil-

ity. Fatigue and cognitive assessment (brain fog, difficulties in speech and concentration, slow thinking, less resistance to mental effort) and rehabilitation are huge unmet needs. Understanding not all patients are depressed and how these symptoms truly impact patients' lives and clarifying if mood instability and depression results from disease activity or is fatigue related, is a major challenge. A multidisciplinary, biopsychosocial intervention addressing reversible causes of fatigue would be very welcome.

Patients' perceptions of healthcare differ from reality: it's not as fast as they wish to receive a diagnosis or treatments. There is no cure, treatments are largely symptomatic, with slow onset of action and many side effects. Much is still unknown. All this can lead to significant frustration and anxiety. The emotional impact of Sjögren's is overlooked with a total lack of psychosocial support. Measuring patients' emotional well-being should be prioritised in routine clinical practice to help mitigate self-isolation and feelings of loneliness. Patients requiring more specialist assessment and support for mental health issues should be referred to psychology or psychiatry.

Communication between patient and doctor is not good enough: difficulties in understanding the disease and treatments remain, and sensitive topics such as family, sexuality, intimacy, self-esteem, and mental health are still taboo and, consequently, overlooked. The importance of honesty and building trust should be stressed as key elements for open and smooth communication between patients and HCPs and adequate time should be given for patients to discuss concerns. Potential barriers to effective communication include poor health literacy, poor education and cultural or personal issues. These should be kept in mind and identified to maximise the support given to patients thus enhancing their engagement with their health outcomes.

Receiving adequate support from a variety of sources is crucial. For this reason, engagement of patients with patient organisations (POs), particularly when newly diagnosed, should be pro-

moted. Lived and experiential knowledge should be complementary to evidence-based knowledge and has the potential to make a significant difference to patients' lives. The therapeutic effect of collaboration and exchanges between patients is surprisingly powerful and should be seen as a valuable resource. Patients who do not reach out to POs, miss the unique emotional support only their peers can facilitate.

Patients need to know the impact their lifestyle may have on the course of their disease or treatment outcomes and need to be empowered to know how to manage it in all circumstances. Comprehensive disease self-management and rehabilitation programmes - patient education and lifestyle guidelines - together with non-pharmacological approaches such as exercise, meditation, and occupational therapies - are rarely explored resources that can be key to restoring quality of life (3). Holistic approaches reflecting the individual's ability to manage symptoms, treatment, physical and psychosocial consequences, and lifestyle changes inherent to living with a chronic condition are awaited and will be warmly received. Also, diet and nutrition cannot be forgotten and need further research. Work represents a major contributor to financial independence, self-esteem, purpose in life and overall QoL. Many patients are of working age at the time of diagnosis; therefore, it is crucial that work-related aspects are addressed, and patients are supported to stay in work and maintain their independence. Referral to occupational therapists and occupational health experts can provide helpful advice and resources in relation to the workplace.

Disability pensions represent not only a great reduction in income but also a high economic burden on families and society that is very difficult to quantify. The economic burden of the disease is also heavy due to the frequent non-availability of free prescriptions for skin, dental, oral, eye care, nutritional products, and vision aids that patients rely for daily function. The long list of products used to prevent, alleviate, and treat symptoms should be considered essential medicines and aids, and

should, at least, be partially covered. Sjögren's is still a disease without specific biomarkers, lacking precise diagnostic techniques as well as specific and effective therapeutic intervention. Advancing research should be prioritized to urgently find answers and appropriate solutions to halt or reverse disease progression. Sjögren's is considered an orphan disease for the purpose of drug approval, so establishing rational targets for drug development is a must. Being a highly heterogeneous disease, even at molecular level, demonstrates the need for stratification and a tailored treatment strategy. Progressing from one-size-fits-all therapy towards personalised medicine based on individual genetics, clinical features, environment, and comorbidities is dependent upon recognition of different disease subtypes and their impact on diagnosis, treatment and prognosis. We recognise and are grateful that in recent years Sjögren's has aroused and captured interest among different people, all driven by the relentless desire to find a cure or a treatment solution for every Sjögren's patient. But we also perceive that there's insufficient funding for research and efforts must be

made to make Sjögren's more attractive among young researchers. Lack of consensus on disease prevalence and incidence across Europe highlights the need for reliable, updated, and harmonized data (European registry). It is vital to know where we stand, to decide what we want to achieve, which resources are needed, and quantify the inevitable investment to get there. Striking disparities exist across countries in access to care and treatment, standards and continuity of care, quality of healthcare delivery, drug and health services prescription availability and reimbursement, and regular follow-up plans. In some countries access to rheumatology is very delayed, limited or non-existent. A similar situation applies to medical specialities such as ophthalmology or dentistry. These variations, existing both in healthcare systems and patient organisations' resources, need to be surveyed, evaluated, and properly addressed. The aim is that best practice examples be used to encourage and support less-developed health care systems and patient organisations to work towards developing high standards of environment, care and support.

Overall a different approach is urgently needed: the future of Sjögren's is collaborative work. Active and synergistic involvement of patients, clinicians, and scientists in the definition of unmet needs and areas of future research is crucial to achieve successful outcomes in clinical medicine (4). It is time for change.

Only when patients, clinicians, and scientists see each other as fellow human beings paving the way to good health will research deliver effective change to patients' lives.

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