Editorial

Variation in primary Sjögren's syndrome care among European countries

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The geographic variation in spending, utilisation and quality of care, across and within countries is well documented. Part of this geographic variation is linked to differences in population health and needs. However, some of the variation may be unwarranted and driven by factors other than patients health needs and preferences, including professional discretion, the availability and distribution of resources, differences in the organisation or delivery of care and in payment models (1). International and national comparative research is therefore needed in order to be able to tackle unwarranted variations, to identify best practices and, ultimately, create significant value in healthcare (2). Data benchmarking also contributes to engage the multispecialty professionals (*e.g.* specialists and GPs) involved in the care of cohort of patients and hold them accountable for the patients that have in common. This is particularly true for patients with long-term chronic diseases who are likely to receive suboptimal care with large variations in the provision of evidence-based treatments and services, and gaps in the disease management due fragmentation of care and poor coordination among the different care settings (3, 4).

Primary Sjögren's syndrome (pSS) is a heterogeneous, complex systemic autoimmune disorder typically characterised by salivary and lacrimal inflammation and dysfunction. During the disease course, several organs and systems may be involved (5-7) and, in a minority of cases, lymphoproliferative complications may also occur, with mucosa-associated lymphoid tissue (MALT) lymphoma of salivary glands being the most frequent haematological type of non-Hodgkin's lymphoma (NHL) detected (8).

Although pSS does not, in general, impair life expectancy and is widely considered as a benign disease, the impact of the disease is not neglectable, imposing a considerable burden on patients' lives (9). Indeed, as recently highlighted by the European Reference Network (ERN) dedicated to Rare and Complex Connective Tissue and Musculoskeletal Diseases (ReCONNET), although clinical practice guidelines on diagnosis and management of SS have been published, a number of unmet needs for pSS are still to be addressed (10). From this perspective, misdiagnosis is still common since the mean length of time between the onset of symptoms and diagnosis is about 7 years, biomarkers of disease severity and outcome are uncertain (11), and pSS patients are likely to experience worse quality of life, to use more care services and report higher out-of pocket expenses compared to controls (12-16). The main factors contributing to suboptimal levels of care and treatments include the lack of awareness and knowledge of the condition among the various healthcare professionals encountered by pSS patients, gaps in evidence-based therapeutic guidelines and, more in general, in evidence-based clinical guidelines for the management of pSS (17, 18). Nonetheless, to the best of our knowledge, the currently available evidence on the sources and effects of variations in the care of pSS is scarce and of limited generalisability in the different European countries. Greater insight is needed with regard to the mechanisms that link the performance and organisation of the different healthcare systems and the experience and quality of life of patients through the use of comparable information across and within health systems.

The EU funded HarmonicSS project (HARMONIzation and integrative

analysis of regional, national and international Cohorts on primary Sjögren's Syndrome (pSS) towards improved stratification, treatment and health policy making disease) was designed to address these gaps in the pSS literature and in particular the lack of harmonised and comparable data on cohorts of pSS patients for comparative research in Europe. The innovative approach of the project is to combine currently available information from national and international longitudinal cohorts of pSS patients with new data collected at different levels (patients, primary care, clinical centres) from different sources (surveys, administrative data, medical registries) in order to address clinical, structural and policy unmet pSS needs and support and stimulate quality improvement.

In particular, one of the main goals of the project is to evaluate pSS care in Europe against criteria of quality, equity and costs by means of three surveys: a) the first to pSS patients to collect information on their journey to the diagnosis, their experience with the care received, the interaction with the different healthcare providers, and the perceived disease burden; b) the second to rheumatologists as core providers of specialist care to pSS patients, to collect data on the organisation and delivery of care at the clinical centres and their involvement and relations with other healthcare professionals; c) the third to general practitioners to understand their knowledge about the disease, to facilitate early diagnosis of the disease and a better management of patients in daily care. For this purpose, starting from existing instruments, ad hoc questionnaires for each of the three surveys have been designed and validated in order to be circulated among European countries through the clinical centres and patient and medical associations. Analysis of the data will contribute to explain variations in pSS care at different levels (different care providers and patients) by comparing patients' experience and quality of life, local medical practice and the way the care to pSS patients is organised and delivered.

To this end, HarmonicSS novel data will contribute to the knowledge on the pSS burden in Europe by reporting on unwarranted variations in health care use, spending, and quality, taking the different contexts into account. Comparative quality reporting will contribute to identify good practices and to enhance the applicability of the findings, to engage the professionals involved in the care of pSS patients by holding them accountable for the patients they have in common and responsible for efficient utilisation of resources with the ultimate goal to shift to value-based healthcare (19).

References

1. WENNBERG JE: Time to tackle unwarranted variations in practice. *BMJ* 2011; 342: d1513.
2. SEGHIERI C, BERTA P, NUTI S: Geographic variation in inpatient costs for Acute Myocardial Infarction care: Insights from Italy. *Health Policy* 2019; 123: 449-56.
3. BROEMELING AM, WATSON DE, PREBTANI F: Population patterns of chronic health conditions, co-morbidity and healthcare use in Canada: implications for policy and practice. *Healthc Q* 2008; 11: 70-6.
4. STUKEL TA, GLAZIER RH, SCHULTZ SE *et al*.: Multispecialty physician networks in Ontario. *Open Med* 2013; 7: e40-55.
5. ARGYROPOULOU OD, VALENTINI E, FERRO F *et al*.: One year in review 2018: Sjögren's syndrome. *Clin Exp Rheumatol* 2018; 36 (Suppl. 112): S14-26.
6. BALDINI C, FERRO F, LUCIANO N, BOMBARDIERI S, GROSSI E: Artificial neural networks help to identify disease subsets and to predict lymphoma in primary Sjögren's syndrome. *Clin Exp Rheumatol* 2018; 36 (Suppl. 112): S137-44.
7. BRITO-ZERÓN P, ACAR-DENIZLI N, NG WF *et al*.: How immunological profile drives clinical phenotype of primary Sjögren's syndrome at diagnosis: analysis of 10,500 patients (Sjögren Big Data Project). *Clin Exp Rheumatol* 2018; 36 (Suppl. 112): S102-12.
8. GOULES AV, TZIOUFAS AG: Lymphomagenesis in Sjögren's syndrome: Predictive biomarkers towards precision medicine. *Autoimmun Rev* 2019; 18: 137-43.
9. HAMMITT KM, NAEGELI AN, VAN DEN BROEK RWM, BIRT JA: Patient burden of Sjögren's: a comprehensive literature review revealing the range and heterogeneity of measures used in assessments of severity. *RMD Open* 2017; 3: e000443.
10. ROMÃO VC, TALARICO R, SCIRÈ CA *et al*.: Sjögren's syndrome: state of the art on clinical practice guidelines. *RMD Open* 2018; 4: e000789.
11. BALDINI C, FERRO F, ELEFANTE E, BOMBARDIERI S: Biomarkers for Sjögren's syndrome. *Biomark Med* 2018; 12: 275-86.
12. MACIEL G, SERVIOLI L, NANNINI C *et al*.: Hospitalisation rates among patients with primary Sjögren's syndrome: a population-based study, 1995-2016. *RMD Open* 2018; 4: e000575.
13. LACKNER A, FICJAN A, STRADNER MH *et al*.: It's more than dryness and fatigue: The patient perspective on health-related quality of life in Primary Sjögren's Syndrome - A qualitative study. *PLoS One* 2017; 12: e0172056.
14. SEGAL B, BOWMAN SJ, FOX PC *et al*.: Primary Sjögren's Syndrome: health experiences and predictors of health quality among patients in the United States. *Health Qual Life Outcomes* 2009; 7: 46.
15. CALLAGHAN R, PRABU A, ALLAN RB *et al*.: Direct healthcare costs and predictors of costs in patients with primary Sjogren's syndrome. *Rheumatology* (Oxford) 2007; 46: 105-11.
16. MIYAMOTO ST, VALIM V, FISHER BA: Health-related quality of life and costs in Sjögren's syndrome. *Rheumatology* (Oxford) 2019 Feb 15 [Epub ahead of print].
17. TZIOUFAS AG, GOULES AV: Limited efficacy of targeted treatments in Sjögren's syndrome: why? *Clin Exp Rheumatol* 2018; 36 (Suppl. 112): S27-8.
18. RAMOS-CASALS M, BRITO-ZERÓN P, SISÓ-ALMIRALL A, BOSCH X, TZIOUFAS AG: Topical and systemic medications for the treatment of primary Sjögren's syndrome. *Nat Rev Rheumatol* 2012; 8: 399-411.
19. GRAY M: Value based healthcare. *BMJ* 2017; 356: j437.