

EDITORIAL

# Lessons learnt from the recent recommendations for the non-pharmacological management of systemic sclerosis

Santiago T<sup>1,2</sup>, Fernandes R<sup>3</sup>, Ferreira R<sup>4</sup>, Parodis I<sup>5,6,7</sup>, Bostrom C<sup>8</sup>

Dear Editor,

In inflammatory rheumatic diseases including systemic sclerosis (SSc) there is growing evidence that treatment strategies should not only target disease control in terms of clinical features and laboratory tests but consider distinct interventions to mitigate all domains of perceived disease impact<sup>1,2</sup>. The results of a multicentric work based on data from the Rheumatic Diseases Portuguese Registry (Reuma.pt)/Scleroderma indicated that the optimization of outcomes for patients with SSc would in all probability require assessment of the needs of individual patients and consider adjunctive interventions in clinical practice to mitigate all significantly affected domains of disease impact<sup>2</sup>.

Recently, in June 2023, a task force under the auspices of EULAR, comprising rheumatologists, health professionals and patient advocates published four overarching principles and twelve recommendations for the non-pharmacological management of people living with SSc and systemic lupus erythematosus (SLE)<sup>3</sup>. This publication emphasizes that non-pharmacological management strategies, such as lifestyle modifications, physical therapy, occupational therapy, and psychosocial support, can greatly improve the quality of life for individuals with SSc<sup>4</sup>. Such interventions can help manage pain, reduce fatigue, improve mobility and function, and address the psychological impact of living with a chronic illness. The four overarching principles for the non-pharmacological management of SSc (and SLE) can be summarized as follows:

- i. should be tailored to the patients' needs, expectations, and preferences;
- ii. should be based on shared decision-making;
- iii. may comprise one or more interventions, and may be provided alone or as an adjunct to pharmaceutical treatment; and
- iv. should not substitute for pharmaceutical treatment when the latter is required.

The generic recommendations for both SSc and SLE are directed toward improving health-related quality of life (HRQoL): patients should be offered education and self-management support; smoking habits should be assessed, and cessation strategies should be implemented; avoidance of cold exposure should be considered for the prevention of Raynaud's phenomenon; and physical exercise should be considered for people living with this disease.

Particularly, in patients with SSc:

- patient education and self-management support should be considered for improving hand function, mouth-related outcomes, HRQoL, and ability to perform daily activities;
- orofacial, hand, and aerobic and resistance exercise should be considered for improving microstomia, hand function, and physical capacity, respectively; and
- manual lymph drainage could be considered for improving hand function in patients with puffy hands.

These recommendations not only intend to guide non-pharmacological management, but also to increase awareness of the importance of patient involvement in the management of their disease and encourage interprofessional and multidisciplinary teams to perform further research in this field. Unfortunately, these non-pharmacological approaches are often underused or not offered to patients. This may be due to a lack of awareness among healthcare providers, limited resources, or a focus primarily on pharmacological interventions. However, it is essential that we recognize the importance of a holistic approach to patient care and rehabilitation and ensure that non-pharmacological strategies are integrated into the management plans for SSc patients.

Importantly, the recognition of the huge impact of Raynaud's phenomenon and digital ulcers on the ev-

<sup>1</sup> Rheumatology Department, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal; <sup>2</sup> Faculty of Medicine, University of Coimbra, Coimbra, Portugal; <sup>3</sup> Leeds Institute of Rheumatic and Musculoskeletal Medicine, University of Leeds, Leeds, United Kingdom; <sup>4</sup> Nursing Research, Innovation and Development Centre of Lisbon (CIDNUR), Nursing School of Lisbon (ESEL), Lisbon, Portugal; <sup>5</sup> Division of Rheumatology, Department of Medicine Solna, Karolinska Institutet, Stockholm, Sweden; <sup>6</sup> Department of Gastroenterology, Dermatology and Rheumatology, Karolinska University Hospital, Stockholm, Sweden; <sup>7</sup> Department of Rheumatology, Faculty of Medicine and Health, Örebro University, Örebro, Sweden; <sup>8</sup> Department of Neurobiology, Care Sciences and Society, Karolinska Institutet, Stockholm, Sweden

**Submitted:** 05/01/2024

**Accepted:** 03/02/2024

**Correspondence to** Tânia Santiago  
E-mail: [tλουςasantiago@hotmail.com](mailto:tλουςasantiago@hotmail.com)

eryday lives of patients with SSc has increased internationally<sup>5,6</sup> and nationally. The first Portuguese Recommendations on the topic, performed under the auspices of GEDRESIS (Rheumatology Portuguese Society Study Group), were formulated to provide guidance on the non-pharmacological and pharmacological management of Raynaud's phenomenon and digital ulcers in SSc and published in this issue of ARP Rheumatology<sup>7</sup>. They were based on a previous systematic review of the literature<sup>8</sup> and multidisciplinary expert opinion, including rheumatologists, surgeons, nurse specialists and a patient research partner. Twelve studies (including only four RCTs) addressing the effectiveness of nonpharmacological interventions in patients with connective tissues diseases were identified, but the wide variations in the content of interventions and outcome measures prevented a meaningful synthesis of results across studies<sup>8</sup>.

Both publications<sup>7, 8</sup> highlight the paucity of high-quality evidence on non-pharmacological interventions; the majority of studies were of modest or low quality. Findings should be interpreted with caution due to heterogeneity in interventions and outcome measures, low methodological quality, and small sample sizes.

These aspects, as well as the urgent need to develop new nonpharmacologic interventions has also been highlighted in the research agenda of the previously mentioned EULAR recommendations<sup>3</sup>. One important aspect is the blinding strategies of the randomized controlled trials, the need for longer-term trials as well as the need to conduct powered and sufficiently large RCTs in SSc. This will only be possible through multicentric trials and international collaborations. Another aspect is the lack of robust evidence on the efficacy of psychosocial interventions for people with SSc<sup>3</sup>. The efficacy of diverse psychosocial interventions holds great promise for enhancing patients' resilience, reducing the impact of disease, and optimizing their ability to live fulfilling lives<sup>2</sup>. Gaining more insight into patients' needs could make substantial contributions toward optimization of treatment strategies aimed at improving outcomes of interest to the SSc patient population, incorporating a holistic and personalized management plan.

Hopefully, both sets of Recommendations will provide guidance on non-pharmacological interventions in the management of SSc and promote their use in clinical practice alongside pharmacotherapy to improve the overall quality of care for people with SSc in Portugal, in Europe, and worldwide.

## REFERENCES

1. Santos E, Duarte C, Ferreira RJO, Pinto AM, Geenen R, da Silva JAP; 'Promoting Happiness Through Excellence of Care' Group. Determinants of happiness and quality of life in patients with rheumatoid arthritis: a structural equation modelling approach. *Ann Rheum Dis*. 2018 Aug;77(8):1118-1124.
2. Santiago T, Santos E, Duarte AC, Martins P, Sousa M, Guimarães F, Azevedo S, Ferreira RM, Guerra M, Cordeiro A, Cordeiro I, Pimenta S, Pinto P, Pinto AM, Salvador MJ, Silva JAPD. Happiness, quality of life and their determinants among people with systemic sclerosis: a structural equation modelling approach. *Rheumatology (Oxford)*. 2021 Oct 2;60(10):4717-4727. <https://doi.org/10.1093/rheumatology/keab083>
3. Parodis I, Girard-Guyonvarc'h C, Arnaud L, Distler O, Domján A, Van den Ende CHM, Fligelstone K, Kocher A, Larosa M, Lau M, Mitropoulos A, Ndosi M, Poole JL, Redmond A, Ritschl V, Alexanderson H, Sjöberg Y, von Perner G, Uhlig T, Varju C, Vriezেকolk JE, Welin E, Westhovens R, Stamm TA, Boström C. EULAR recommendations for the non-pharmacological management of systemic lupus erythematosus and systemic sclerosis. *Ann Rheum Dis*. 2023 Jul 10:ard-2023-224416. <https://doi.org/10.1136/ard-2023-224416>
4. Navas-Otero A, Gómez-De-Castro S, Ortiz-Rubio A, Heredia-Ciuró A, Martín-Núñez J, Calvache-Mateo A, Valenza MC. Effects of non-pharmacological therapies on hand function and the ability to perform daily activities in people with systemic sclerosis: A systematic review and meta-analysis of randomized control trials. *Int J Rheum Dis*. 2023 Jun;26(6):1029-1040. doi: 10.1111/1756-185X.14721. Epub 2023 May 3. PMID: 37137818. <https://doi.org/10.1111/1756-185X.14721>
5. Stöcker JK, Schouffoer AA, Spierings J, Schriemer MR, Potjewijd J, de Pundert L, van den Hoogen FHJ, Nijhuis-van der Sanden MWG, Staal JB, Satink T, Vonk MC, van den Ende CHM. Arthritis Research and Collaboration Hub study group. Evidence and consensus-based recommendations for non-pharmacological treatment of fatigue, hand function loss, Raynaud's phenomenon and digital ulcers in patients with systemic sclerosis. *Rheumatology (Oxford)*. 2022 Apr 11;61(4):1476-1486. <https://doi.org/10.1093/rheumatology/keab537>
6. Spierings J, van den Ende C, Schriemer R, de Pundert L, Moens HB, van Laar J, de Vries-Bouwstra J, Vonk M. Optimal care for systemic sclerosis patients: recommendations from a patient-centered and multidisciplinary mixed-method study and working conference. *Clin Rheumatol*. 2019 Apr;38(4):1007-1015. <https://doi.org/10.1007/s10067-018-4358-x>
7. Ref to be added.
8. Costa E, Cunha-Santos F, Dourado E, Oliveira D, Falzon L, Romão V, Duarte AC, Cordeiro A, Santiago T, Sepriano A. Systematic Literature Review to inform the Portuguese recommendations for the management of Raynaud's phenomenon and digital ulcers in systemic sclerosis and other connective tissue diseases. *ARP Rheumatology*, 2024, online-first, 02.02.2024.