Undiagnosed and Untreated: The Realities of systemic sclerosis in Europe today

Expert recommendations

FESCA IVZW
Federation of European Scleroderma Associations

World Scleroderma Day

What is systemic sclerosis (SSc) also known as scleroderma?

SSc is a complex, multi-organ disease[1] where the patient's immune system attacks organ tissue causing overproduction of fibrous connective tissue, which causes problems such as pulmonary hypertension, heartburn, bowel problems, shortness of breath and increases mortality risk.[2] SSc can also affect the lungs, blood vessels, heart, kidneys, gastrointestinal tract and the musculoskeletal system. Raynaud's phenomenon is often the first sign of SSc, where poor blood circulation leads to a numbing and coldness of the fingers and toes.[3, 4]

What must be done to improve diagnosis and care in Europe?

Create and link national patient registries

Better access to patient registries for systemic sclerosis and coordination of these resources across EU Member States are key elements of national health strategies that will improve the medical community's understanding of the condition. This will ensure timely treatment and boost the quality of life for people living with SSc.

- → Build a national SSc patient registry to collect and share standardized data on the health status of patients.
- → Apply the 'FAIR' principle to ensure that all SSc data is Findable, Accessible, Interoperable and Reusable nationally and with other health systems and registries.[5, 6]
- → Link to the EUSTAR database, a powerful tool that all countries can use today to share systemic sclerosis data, and ideal for use by countries where registries are not established.

Reduce the time to diagnosis

Accurate and rapid diagnosis of systemic sclerosis will ensure that each patient living with this condition has the best possible quality of life and positive health outcomes. But diagnosis is complex due to a lack of general knowledge of this condition among medical professionals. This causes delays of months, or even years,[7, 8, 9] between a patient's first visit to their doctor, accurate identification of the disease, and treatment.[10]

- → Set clear diagnosis guidelines to help healthcare professionals improve early diagnosis of the disease for rapid treatment.
- → Define international diagnosis guidelines, involving rheumatologists, immunologists and internists.
- → Apply VEDOSS criteria (Very Early Diagnosis of Systemic Sclerosis) in national strategies and action plans.
- → Ensure training of medical and healthcare professionals, especially in primary care to focus on identifying first signs of the disease.
- → Put in place 'risk stratification' at population level to identify patient subgroups who are at risk of developing SSc, such as those with Raynaud's phenomenon.

Ensure timelier referral of patients to specialized care

Along with timely diagnosis, referral of the patient to a qualified specialist at the right time is a critical element needed to bring systemic sclerosis patients the best possible quality of treatment. In several EU countries, delayed referrals[11] are cited by healthcare specialists as an obstacle to providing effective levels of care to their systemic sclerosis patients.

- → Adopt a networked 'hub and spoke' approach for the national health system for systemic sclerosis care, that places the patient at the center of specialist disease knowledge, patient advice and management approaches.
- → Deliver digital services to make health systems more responsive to the needs of people living with SSc. Videoconferencing and telemedicine platforms link systemic sclerosis specialists with patients in all locations, boosting access for all and the quality of care.

Improve treatment recommendations and access to appropriate medicines

There is no standardized treatment pathway in Europe, with some countries using the EULAR recommendations while others have national or regional recommendations that are not always up to date. However, timely access to treatment and care (including physiotherapy, occupational therapy, psychological and psycho-social care) **is crucial** for a patient's quality of life and prognosis.

- → Countries can improve treatment access by:
  - » Ensuring timely application of the HTA Regulation 2021/2282, working with European Reference Networks (ERNs), Patient Advocacy Groups and members of the Stakeholder Network to implement the Regulation.
  - » Establishing clear timeframes for pricing and reimbursement discussions, to ensure that approved treatment reaches all eligible patients.
  - » Embracing the revision of Orphan Medicinal Products legislation (141/2000) to drive research on new therapies for high clinical unmet needs.
- → More research on health system performance for medicinal products approval will improve access.
- → Health services should prioritize access to psychological and psycho-social care and other non-pharmacological treatment (e.g., hand and body physiotherapy) for SSc patients.

Deliver better quality information to systemic sclerosis patients

Information on systemic sclerosis is readily available for patients in most European countries,[12,13] but healthcare professionals report that more personalized information is needed to address the specific SSc sub-types – and more attention is needed to inform and educate each patient at the time and point of diagnosis.[14]

- → Tailor information for patients to the specific sub-type or stage that each patient is experiencing.
- → Include advice for patients on physical and psychological consequences of their condition, in treatment information.
- → Share detailed and practical information from doctor to patient at the point of diagnosis.
- → Tap EU Cohesion Funds and other public financial schemes to support patient groups, and invest in research, patient support and advisory activities.

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About FESCA

FESCA, Federation of European Scleroderma Associations aisbl, acts at a pan-European level to promote and achieve its objectives in alignment with the aims of the national groups it represents. As an umbrella group, FESCA supports its member organizations, while the support of individual sufferers of scleroderma remains the sole remit of national organizations.

World Scleroderma Day

About the campaign

This position paper was developed as part of the "Find the Light to Bloom" campaign, launched by FESCA on World Scleroderma Day, which is celebrated annually on 29 June. The campaign seeks to shine a light on the unmet needs of people living with scleroderma, and urge policy-makers to support policies that ensure timely diagnosis, as well as timely access to treatment and improved quality of life of all people living with systemic sclerosis

Disclaimer

The content of the Position Paper summarized here represents the views of the authors only and is their sole responsibility. The Position Paper is not intended as an exhaustive or scientific review of the care and management of SSc in Europe. The literature review and survey responses done in compiling this information have provided useful insights into some of the issues affecting a cross-section of patients living with SSc in various European countries, that are summarized here.

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