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Systemic sclerosis: The need for structured care

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A B S T R A C T

Autoimmune connective tissue diseases (CTDs) have a propensity to affect multiple organ systems as well as physical function, quality of life, and survival. Their clinical heterogeneity, multisystem involvement, and low worldwide prevalence present challenges for researchers to establish a study design to help better understand the course and outcomes of CTDs.

Systemic sclerosis (SSc) is a notable example of a CTD, wherein longitudinal cohort studies (LCS) have enabled us to elucidate disease manifestations, disease course, and risk and prognostic factors for clinically important outcomes, by embedding research in clinical practice. Nevertheless, further efforts are needed to better understand SSc especially with regard to recognizing organ involvement early, developing new therapies, optimizing the use of existing therapies, and defining treatment targets.

The heterogeneous multi-organ nature of SSc would lend itself well to a structured model of care, wherein step-up treatment algorithms are used with the goal of attaining a prespecified treatment target. In this chapter, we discuss the rationale for a structured treatment approach in SSc and propose possible treatment algorithms for three of the more common disease manifestations, namely skin involvement, digital ulcers and gastrointestinal tract involvement. We discuss possible strategies for evaluating and implementing these algorithms in the setting of

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LCS. We conclude by presenting a research agenda for the development of structured models of care in SSc.
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Practice points

- Longitudinal cohort studies (LCS) have contributed significant insight into disease manifestations, disease course, risk factors, and prognostic factors for clinically important outcomes in low-prevalence multisystem heterogeneous conditions such as connective tissue diseases (CTDs).
- Systemic sclerosis (SSc) is an excellent example of how LCS have furthered our understanding of the natural history of SSc, with research being embedded in clinical practice.
- Structured models of care are successful in changing clinical practice, thereby improving quality and efficiency of care and patient outcomes. In addition, they reduce disease-associated economic burden by reducing health-care utilization and increasing patients' ability to maintain paid employment.
- A structured approach to the care of three of the key clinical manifestations of SSc, namely skin involvement, digital ulcers and gastrointestinal tract (GIT) involvement, is outlined in this chapter.

Introduction

Autoimmune connective tissue diseases (CTDs) are rheumatic conditions with a propensity to affect multiple organ systems. They include conditions such as systemic sclerosis (SSc), systemic lupus erythematosus (SLE), inflammatory myopathies, Sjögren's syndrome, and mixed connective tissue disease (MCTD). The clinical heterogeneity of these conditions results in significant knowledge gaps and unmet needs. Accordingly, our understanding of their natural history, pathogenesis, triggers, and response to treatment must be expanded [1].

Longitudinal cohort studies (LCS) have significantly elucidated disease manifestations, disease course, risk factors, and prognostic factors for clinically important outcomes in low-prevalence multisystem heterogeneous conditions such as the CTDs, which are otherwise very difficult to study. SSc is an excellent example of how LCS have furthered our understanding of the natural history of SSc, with research being embedded in clinical practice.

SSc is a chronic CTD with variable involvement of the skin and internal organs and a worldwide prevalence ranging from 7 per million to 489 per million [2]. Among the rheumatic diseases, SSc has the highest case-based mortality with an average loss of life expectancy of >20 years relative to the general population [3]. It is also one of the most costly rheumatic diseases, with SSc patients utilizing more health-care dollars per annum than their age- and sex-matched counterparts with rheumatoid arthritis (RA) or psoriatic arthritis [4]. Morbidity in SSc is substantial, comparable with that of heart disease, depression, and some cancers [5]. Morbidity and irreversible organ damage can occur within the first 2 years of disease onset, leading to impaired physical function and reduced health-related quality of life (HRQoL) [5]. Therefore, the early stages of SSc present a narrow, but important window of opportunity for preventing irreversible organ damage.

Due to the relatively low disease frequency of SSc, multicenter collaborations are needed to recruit a sufficient number of patients to power research studies. Collaboration between centers currently occurs in SSc via several LCS such as the Pittsburgh Scleroderma Centre, Australian Scleroderma Interest Group (ASIG), Canadian Scleroderma Research Group (CSRG), European Scleroderma Trials and Research Group (EUSTAR), and Genetics versus Environment in Scleroderma Outcomes Study (GENISOS) cohorts, among others. These collaborations have been key to furthering our understanding of certain disease manifestations in SSc such as interstitial lung

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