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Trajectories of functional disability from Systemic Sclerosis onset: a Scleroderma Patient-centered intervention Network (SPIN) Cohort study.

Systemic sclerosis (SSc) is a rare, chronic autoimmune disease that affects the skin and internal organs and is associated with functional disability, typically assessed using the Health Assessment Questionnaire–Disability Index (HAQ-DI). Prior SSc studies on HAQ-DI trajectories were limited by small samples (215 to 1250 participants), lack of consideration of non-linear trajectories, and follow-up beginning at cohort enrolment rather than disease onset, despite different disease durations at entry, which may have obscured changes across disease course. We modelled functional disability trajectories from SSc onset and evaluated differences by disease subtype, age at onset, sex, and country.

We used data from the Scleroderma Patient-centered Intervention Network (SPIN) Cohort, in which adults with physician-confirmed SSc complete assessments every 3 months. Participants with complete baseline covariates and ≥1 HAQ-DI assessment were included. We used a linear mixed-effects model to examine HAQ-DI trajectories since onset. Both age at onset and time since onset were included to account for chronological age and disease duration at each assessment. SSc subtype, sex, and country were also included as covariates. Effect modification was assessed using interaction terms with time since onset. Non-linearity was evaluated using spline functions.

We included 2,154 participants with 19,550 assessments (median follow-up = 2.3 years; range = 0.0 to 6.3 years). Overall, the HAQ-DI increased non-linearly over time since onset. Predicted scores were similar across onset ages in early disease (e.g., 5 years post-onset: 0.57 points [95% CI 0.44 to 0.70] at onset age 20; 0.73 [0.64 to 0.81] at onset age 60), but were substantially higher among participants with older onset age in later disease (e.g., 15 years post-onset: 0.53 [0.42 to 0.64] at onset age 20; 1.20 [1.10 to 1.30] at onset age 60). HAQ-DI scores at onset were higher in diffuse cutaneous than in limited cutaneous disease (difference 0.47 points [0.11 to 0.83]). Trajectories were modified by subtype and age at onset. At older onset (e.g., 60 years), diffuse cutaneous disease had higher HAQ-DI scores than limited in early disease (5 years post-onset: 1.09 [1.01 to 1.19] vs 0.73 [0.64 to 0.81] for limited), but scores were similar in later years (15 years post-onset: 1.29 [1.16 to 1.42] vs 1.20 [1.10 to 1.30]), which reflects steeper increases at older onset in limited cutaneous disease, despite initially higher scores in diffuse cutaneous disease.

Understanding functional disability progression in SSc requires consideration of onset age and disease subtype.