Impaired quality of life in systemic sclerosis and patient perception of the disease: a large international survey

Background: Systemic sclerosis (SSc) is known as one of the most fatal rheumatic diseases, but it also promotes many detrimental effects on health-related quality of life (HRQOL). However, the data provided so far on HRQOL in SSc suffer from a weak power related to the difficulty to recruit large sample. Furthermore, previous works mostly reported the outcome of early diffuse cutaneous SSc (DcSSc) subset.

Objective: To assess HRQOL and disease perception in a large and international cohort of SSc patients using validated questionnaires.

Methods: We conducted an international cross-sectional survey supported by EUSTAR, FESCA and Scleroderma US Foundation, from December 2013 to April 2014. We built a standardized questionnaire translated and available on a website, including: Socio-demographic information, disease characteristics and self-assessment questionnaires namely Short Form 36 (SF-36) and Revised Illness Perception Questionnaire (IPQ-R).

Results: 1902 SSc patients from 60 countries were included with a mean±standard deviation (SD) age of 54±16 years and a mean±SD disease duration of 13±12 years. 712 (34.4%) patients had DcSSc, 853 (44.8%) limited cutaneous SSc (LcSSc) and 122 (6.4%) sine scleroderma SSc.
HRQOL appeared to be strongly impaired in SSc patients; in physical health (PCS, mean±SD 43.4±23.4; 100=best health) but also in mental health (MCS, mean±SD 52.3±23.1; 100=best health). SSc patients also held strong perceptions of the chronicity and negative consequence of the disease, and experienced negative emotions due to SSc.

Compared to LcSSc, patients with DcSSc had poorer HRQOL both in physical (PCS, mean±SD 46.6±23.7 vs 39.8±22.3; p<0.0001) and mental health (MCS, mean±SD 53.8±23.0 vs 50.3±23.2; p=0.0032), and had more negative perception of their disease. In terms of disease duration, late SSc patients perceived their disease as more chronic (p<0.0001), less controllable (p=0.03) and with more consequences (p=0.008), but they had a better understanding of SSc and they experienced less negative emotions.

Patients ranked organ involvements having the most severe impact on daily life and on illness severity perception: Raynaud phenomenon (6.6 and 6.5/10), gastrointestinal complications (5.4 and 5.5/10), musculoskeletal (4.9 and 5/10) and skin lesion (4.5 and 4.6/10) were the top rated involvements.

Non-European SSc patients had a greater impact on their QOL, mainly in physical health (PCS, mean±SD 39.9±22.6 vs 46.0±23.7; p<0.0001), and more negative illness perception compared to European patients but
they had a better understanding of SSc and experienced less negative emotions.

**Conclusion:** This study provides unique information about patient perception of the disease and impact on QOL according to disease subset, disease duration and various geographical origins. The key message overall is the major impact on QOL and rather negative perception of their disease by SSc patients. Nevertheless, illness perception tends to improve with disease duration suggesting effective coping strategies. These results will have to be taken into consideration for patient management and future trials.