The registry of the German Network for Systemic Scleroderma: frequency of disease subsets and patterns of organ involvement.

OBJECTIVE: Systemic sclerosis (SSc) is a rare, heterogeneous disease, which affects different organs and therefore requires interdisciplinary diagnostic and therapeutic management. To improve the detection and follow-up of patients presenting with different disease manifestations, an interdisciplinary registry was founded with contributions from different subspecialties involved in the care of patients with SSc. METHODS: A questionnaire was developed to collect a core set of clinical data to determine the current disease status. Patients were grouped into five descriptive disease subsets, i.e. lcSSc, dcSSc, SSc sine scleroderma, overlap-syndrome and UCTD with scleroderma features. RESULTS: Of the 1483 patients, 45.5% of patients had lcSSc and 32.7% dcSSc. Overlap syndrome was diagnosed in 10.9% of...
patients, while 8.8% had an undifferentiated form. SSc sine scleroderma was present in 1.5% of patients. Organ involvement was markedly different between subsets; pulmonary fibrosis for instance was significantly more frequent in dcSSc (56.1%) than in overlap syndrome (30.6%) or lcSSc (20.8%). Pulmonary hypertension was more common in dcSSc (18.5%) compared with lcSSc (14.9%), overlap syndrome (8.2%) and undifferentiated disease (4.1%). Musculoskeletal involvement was typical for overlap syndromes (67.6%). A family history of rheumatic disease was reported in 17.2% of patients and was associated with early disease onset (P< 0.005). CONCLUSION: In this nationwide register, a descriptive classification of patients with disease manifestations characteristic of SSc in five groups allows to include a broader spectrum of patients with features of SSc.