There Is a Need for New Systemic Sclerosis Subset Criteria: A Content Analytic Approach


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Background/Purpose: Systemic sclerosis (SSc) is a family of diseases unified by the presence of immune activation, vasculopathy and fibrosis. The concept of SSc subsets cannot be easily measured but is considered to be real. To evaluate the purpose, strengths and limitations of the limited/diffuse subset criteria, and identify areas requiring improvement.

Methods: We conducted a content analytic study consisting of semi-structured interviews with 30 SSc experts. The interview transcripts underwent an iterative process with text deconstructed to single thought units until a saturated conceptual framework with coding was achieved and respondent occurrence tabulated. This was followed by serial cross-referential analyses establishing a set of pervasive complex thought clusters.

Results: Of the 30 experts, 26 (87%) were male, 19 (63%) were from Europe and 11 (37%) were from North America. The experts had seen SSc patients for a mean 23 (SD 10.7) years, and saw a mean of 122 (SD 185) new SSc patients annually. Three thematic clusters were noted regarding the utility of subsetting: to facilitate research and communication, to inform management, and to inform prognosis (prediction of internal organ involvement, survival). The strength of the limited/diffuse system was its ease of use, however 10% stated this system has ‘little or no value.’ Limitations of the diffuse/limited classification were the risk of misclassification, predictions/generalizations did not always hold true, and that the elbow or knee threshold is arbitrary. 87% use more than 2 subsets including: SSc sine scleroderma, overlap conditions, antibody determined subsets, subsetting based on speed of progression, and age of onset (juvenile, elderly). Considerations for the next phase of criteria development include incorporation of rate of change and hierarchal clustering (limited/diffuse, then by antibodies).

Conclusion: We interviewed international SSc experts and synthesized their views on subset criteria. These results can inform our efforts to develop revised criteria to guide research, prognostication and management.

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