ABSTRACT NUMBER: 1888

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

Sindhu Johnson[1], Medha Soowamber[2], Jaap Fransen[3], Dinesh Khanna[4], Frank H.J. van den Hoogen[5], Murray Baron[6], Marco Matucci Cerinic[7], Christopher P. Denton[8], Thomas A. Medsger Jr.[9], Patricia E. Carreira[10], Gabriela Riemekasten[11], Jorg HW. Distler[12], Armando Gabrielli[13], Virginia D. Steen[14], Lorinda Chung[15], Richard Silver[16], John Varga[17], Ulf Müller-Ladner[18], Madelon C. Vonk[19], Ulrich A. Walker[20], Frank Wollheim[21], Ariane L. Herrick[22], Daniel E. Furst[23], Lazlo Czirjak[24], Otylia Kowal-Bielecka[25], Francesco Del Galdo[26], Maurizio Cutolo[27], Nicolas Hunzelmann[28], Charles Murray[29], Ivan Foeldvari[30], Luc Mouthon[31], Nemanja Damjanov[32], Bashar Kahaleh[33], Tracy M. Frech[34], Shervin Assassi[35], Lesley Ann Saketkoo[36] and Janet E. Pope[37], [1]Toronto Scleroderma Program, Toronto Western Hospital, Mount Sinai Hospital, University of Toronto, University Health Network Pulmonary Hypertension Programme, Toronto, ON, Canada, [2]Rheumatology, University of Toronto/ Toronto Western Hospital, Toronto, ON, Canada, [3]Rheumatology, Radboud University Medical Centre, Nijmegen, Netherlands, [4]Medicine, University of Michigan, Ann Arbor, MI, [5]Rheumatology, Sint Maartenskliniek, Nijmegen, Netherlands, [6]Pavillion A, Rm 216, Jewish General Hospital, Montreal, QC, Canada, [7]Department of BioMedicine, Division of Rheumatology, Transition Unit, University of Florence, Firenze, Italy, [8]Centre for Rheumatology, Royal Free Hospital, London, United Kingdom, [9]Medicine/Rheumatology, Univ of Pittsburgh, Pittsburgh, PA, [10]Department of Rheumatology, Hospital Universitario 12 de Octubre, Madrid, Spain, [11]Rheumatology, Human medicine, BERLIN, Germany, [12]Department of Internal Medicine 3, University of Erlangen-Nuremberg, Erlangen, Germany, [13]Clinica Medica, Università Politecnica delle Marche, Ancona, Italy, [14]Rheumatology, Georgetown University Medical Center, Washington, DC, [15]Division of Immunology and Rheumatology, Stanford University School of Medicine, Stanford, CA, [16]Div Rheumatology & Immunology, Medical University of South Carolina, Charleston, SC, [17]Medicine, Northwestern University Feinberg School of Medicine, Chicago, IL, [18]Rheumatology, Internal Medicine, Bad Nauheim, Germany, [19]Rheumatology, Rheumatology, Nijmegen, Netherlands, [20]Rheumatology, Systemic Sclerosis, Basel, Switzerland, [21]Rheumatology, Lund University Hospital, Lund, Sweden, [22]Centre for Musculoskeletal Research, University of Manchester, Manchester Academic Health Science Centre, Manchester, United Kingdom, [23]Medicine, University of California, Los Angeles, David Geffen School of Medicine, Los Angeles, CA, [24]University of Pécs Medical Center, Pécs, Hungary, [25]Department of Rheumatology and Internal Medicine, Medical University of Bialystok, Bialystok, Poland, [26]University of Leeds, Leeds, United Kingdom, [27]Research Laboratory and Academic Division of Clinical Rheumatology, Department of Internal Medicine, University of Genova, Genova, Italy, [28]Department of Dermatology, University of Cologne, Cologne, Germany, [29]Department of Gastroenterology, Royal Free Hospital, London, United Kingdom, [30]Rheumatology, Hamburg, Germany, [31]Department of Internal Medicine, Department of Internal Medicine, Cochin Hospital, Referent Center for Necrotizing Vasculitis and Systemic Sclerosis, Paris-Descartes University, AP-HP, Paris, France, [32]Resavska 69, Institute of Rheumatology, Belgrade, Serbia, [33]Medicine/Rheumatology, The University of Toledo, Toledo, OH, [34]Div of Rheumatology, University of Utah Medical Ctr, Salt Lake City, UT, [35]Rheumatology, University of Texas Medical School at Houston, Houston, TX, [36]Tulane University Lung Center, New Orleans Scleroderma and Sarcoidosis Patient Care and Research Center, New Orleans, LA, [37]University of Western Ontario, London, ON, Canada

Meeting: 2015 ACR/ARHP Annual Meeting

Date of first publication: September 29, 2015

Keywords: Arthritis, fibrosis, Rheumatic disease, scleroderma and systemic sclerosis

SESSION INFORMATION

Date: Monday, November 9, 2015

Session Type: ACR Poster Session B

Session Title: Systemic Sclerosis, Fibrosing Syndromes and Raynaud's - Clinical Aspects and Therapeutics Poster II

Session Time: 9:00AM-11:00AM

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.

Disclosure: S. Johnson, None; **M. Soowamber**, None; **J. Fransen**, None; **D. Khanna**, Bristol-Myers Squibb, 2,EMD Serono, 2,Genentech and Biogen IDEC Inc., 2,Bayer, 5,Biogen Idec, 5,Cytori, 5,EMD Serono, 5,Forward, 5,Genentech and Biogen IDEC Inc., 5,Gilead, 5,Lycera, 5,Seattle Genetics, 5; **F. H. J. van den Hoogen**, None; **M. Baron**, None; **M. Matucci Cerinic**, None; **C. P. Denton**, None; **T. A. Medsger Jr.**, None; **P. E. Carreira**, None; **G. Riemekasten**, None; **J. H. Distler**, None; **A. Gabrielli**, None; **V. D. Steen**, None; **L. Chung**, Gilead, 4; **R. Silver**, None; **J. Varga**, None; **U. Müller-Ladner**, None; **M. C. Vonk**, None; **U. A. Walker**, None; **F. Wollheim**, None; **A. L. Herrick**, None; **D. E. Furst**, Gilead, 2,GlaxoSmithKline, 2,NIH, 2,Novartis Pharmaceutical Corporation, 2,Pfizer Inc, 2,Roche Pharmaceuticals, 2,Genentech and Biogen IDEC Inc., 2,UCB, 2,Abbvie, 5,Actelion Pharmaceuticals US, 5,Amgen, 5,Bristol-Myers Squibb, 5,Cytori, 5,Janssen Pharmaceutica Product, L.P., 5,Gilead, 5,GlaxoSmithKline, 5,NIH, 5,Novartis Pharmaceutical Corporation, 5,Pfizer Inc, 5,Roche Pharmaceuticals, 5,Genentech and Biogen IDEC Inc., 5,UCB, 5,Abbvie, 8,Actelion Pharmaceuticals

US, 8,Bristol-Myers Squibb, 2,Amgen, 2,Actelion Pharmaceuticals US, 2,Abbvie, 2,UCB, 8; **L. Czirjak**, None; **O. Kowal-Bielecka**, None; **F. Del Galdo**, None; **M. Cutolo**, None; **N. Hunzelmann**, None; **C. Murray**, None; **I. Foeldvari**, None; **L. Mouthon**, None; **N. Damjanov**, None; **B. Kahaleh**, None; **T. M. Frech**, None; **S. Assassi**, None; **L. A. Saketkoo**, None; **J. E. Pope**, Amgen, 2.

To cite this abstract in AMA style:

Johnson S, Soowamber M, Fransen J, Khanna D, van den Hoogen FHJ, Baron M, Matucci Cerinic M, Denton CP, Medsger TA Jr., Carreira PE, Riemekasten G, Distler JH, Gabrielli A, Steen VD, Chung L, Silver R, Varga J, Müller-Ladner U, Vonk MC, Walker UA, Wollheim F, Herrick AL, Furst DE, Czirjak L, Kowal-Bielecka O, Del Galdo F, Cutolo M, Hunzelmann N, Murray C, Foeldvari I, Mouthon L, Damjanov N, Kahaleh B, Frech TM, Assassi S, Saketkoo LA, Pope JE. There Is a Need for New Systemic Sclerosis Subset Criteria: A Content Analytic Approach [abstract]. *Arthritis Rheumatol.* 2015; 67 (suppl 10). http://acrabstracts.org/abstract/there-is-a-need-for-new-systemic-sclerosis-subset-criteria-a-content-analytic-approach/. Accessed October 2, 2015.
