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# A world of progress in systemic sclerosis

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*P.J. Clements, M. Matucci-Cerinic, S. Bombardieri, J.R. Seibold*

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*Philip J. Clements, MD  
Professor of Medicine  
Division of Rheumatology  
David Geffen School of Medicine at UCLA  
Los Angeles, California, USA*

*Marco Matucci-Cerinic, MD, PhD  
Professor and Chief  
Division of Rheumatology  
University of Florence  
Florence, Italy*

*Stefano Bombardieri, MD  
Professor of Rheumatology  
Rheumatic Disease Unit  
Department of Internal Medicine  
University of Pisa  
Pisa, Italy*

*James R. Seibold, MD  
Marvin & Betty Danto Research  
Professor Director  
University of Michigan  
Scleroderma Program  
Ann Arbor, Michigan, USA*

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We welcome you to the first issue of “Scleroderma Care and Research” published as a Supplement to *Clinical and Experimental Rheumatology*. This new endeavor is dedicated to advancing standards of clinical care and research in scleroderma on an international basis. We thank the many members of EUSTAR (<http://www.eustar.org>) – the EULAR Scleroderma Trials & Research Group and the SCTC (<http://www.sctc-online.org>) – the Scleroderma Clinical Trials Consortium for the hours of hard work and dedication that gave birth to this inaugural issue.

The SCTC was founded in 1994 around the simple concept that the patient with scleroderma deserved nothing but the best in terms of clinical research and care. It was recognized that advances in molecular and cellular biology were bringing us closer to a reasonable understanding of disease pathogenesis and that plausible, mechanism-driven approaches to therapy were soon to be in hand. Certain basic information was by consensus agreed to be lacking. If a successful therapy were to emerge, how would it be measured and just what would the expectations be? What sort of outcome and response measures did we need to permit reliable and robust trial design? Could a path forward be blazed that bridged between organ-based therapy and the elusive goal of disease modification?

Scleroderma remains the most fatal of the rheumatologic diseases. It would seem to require aggressive definitive action but against what and with what approach? The clinical heterogeneity of the syndrome poses additional challenges. What therapeutic approach would be applicable to the individual with rapidly progressive diffuse cutaneous scleroderma as well as to the individual with stable later stage limited disease?

In spite of the challenges, progress has been considerable on all fronts. EUSTAR has worked effectively to create many centers in the EU and to generate a broad-base of research in the setting of higher standards of care. The SCTC has served as the driver of many effective randomized controlled trials. Evidence based medicine can now be applied in many areas of scleroderma – most notably in treatment of interstitial lung disease, pulmonary arterial hypertension and Raynaud complications.

Licensing approvals have been obtained for prostacyclins (epoprostenol, iloprost and treprostinil), endothelin receptor antagonists (bosentan, sitaxentan, ambrisentan) and Type V cGMP phosphodiesterase inhibitors (sildenafil, tadalafil) although the dominant indication has been for pulmonary arterial hypertension. Off-patent drugs such as cyclophosphamide now have firm evidence supporting use in carefully selected patients. Aggressive protocols driven by survival endpoints are in progress, most notably immunoablative therapy with stem cell reconstitution.

These success stories are, in part, matched by assiduous efforts to develop outcome and response measures. Current research focuses a combined response index applicable to all studies as well as core sets for specific issues of parenchymal and vascular features of lung involvement. The next generation of drugs will be studied in sophisticated trials with robust and reliable outcomes.

All of this has occurred through extensive collegial international collaborative efforts. There is a community of committed scleroderma caregivers and researchers. This journal offers you access to some of this energy and productivity. It is indeed a world of progress. We invite you to read of our progress and share our excitement.