

ABBREVIATED CURRICULUM VITAE

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Education and Training

9/1969-6/1972 Louisiana State University, Baton Rouge, Louisiana, B.S.
8/1972-6/1975 SUNY – Stony Brook School of Medicine, Stony Brook, New York, M.D.
7/1975-6/1976 Intern in Medicine, Long Island Jewish-Hillside Medical Center, New Hyde Park, New York
7/1976-6/1977 Junior Resident in Medicine, Long Island Jewish-Hillside Medical Center, New Hyde Park, New York
7/1977-6/1978 Senior Resident in Medicine, Long Island Jewish-Hillside Medical Center, New Hyde Park, New York
7/1978-6/1980 Fellow in Rheumatology/Immunology, University of Pittsburgh, School of Medicine, Pittsburgh, Pennsylvania

Certification and Licensure

July 1, 1976 Diplomate, National Board of Medical Examiners (Certificate # 176659)
September 13, 1978 Diplomate, American Board of Internal Medicine (Certificate # 68107)
June 17, 1980 Diplomate, Rheumatology, ABIM (Certificate #68107)
7/1976-2014 New York State 134149 (inactive)
7/1978-6/1980 Pennsylvania 21256-E (inactive)
7/1980-7/2007 New Jersey MA 37369 (inactive)
7/1980-8/2004 New Jersey CDS D-29224 (inactive)
7/1980-present DEA AS9248003
3/2004-2014 Michigan 4301083197 (inactive)
3/2004-2014 Michigan CDS 5315016983 (inactive)
2008- present NPI 1821173337
2010-present Connecticut 48454
2010- present Connecticut CSP 0047273

Academic, Administrative, and Clinical Appointments

UMDNJ-Robert Wood Johnson Medical School

7/1980-5/1986 Assistant Professor of Medicine, Division of Rheumatology
7/1980-8/2004 Director, UMDNJ Scleroderma Program

12/1984-9/1989 Acting Director, Clinical Research Center, NJ Center for Advanced Biotechnology and Medicine, UMDNJ & Rutgers University
5/1986-7/1991 Chief, Division of Rheumatology
5/1986-7/1998 Associate Professor of Medicine, Division of Rheumatology (tenured)
9/1989-6/1995 Director, Clinical Research Center
9/1989-6/1995 W.H. Conzen Chair of Clinical Pharmacology
9/1989-6/1995 Chair, UMDNJ Program in Clinical Pharmacology
7/1998-8/2004 Professor of Medicine, Division of Rheumatology

University of Michigan Medical School

8/2004-2/2010 Professor of Internal Medicine/Rheumatology, Director, University of Michigan Scleroderma Program, University of Michigan Health System (tenured)
5/2006-2/2010 Marvin & Betty Danto Research Professor

University of Connecticut Health Center

2/2010-10/2010 Professor and Chief, Division of Rheumatology

Scleroderma Research Consultants, LLC

9/2010- Founder and Principal

Grants

Present and Active

Direct Sponsor - Covis Sarl 1/2015-12/2016
A Double-Blinded Crossover Study of Topical Formulation of Nitroglycerine, Vascana®, versus Matching Vehicle in the Subjective and Physiologic Responses to Controlled Cold Challenge in Subjects with Raynaud Phenomenon Secondary to Connective Tissue Disease.
Role: Clinical Lead

Direct Sponsor – InterMune Study completed-Reportage in Progress
An Open-Label, Randomized Phase 2 Study of the Safety and Tolerability of Pirfenidone when Administered to Patients with Systemic Sclerosis-Related Interstitial Lung Disease
This study investigates the safety and tolerability of pirfenidone in systemic sclerosis.
Role: Member, Steering Committee

Direct Sponsor – United Therapeutics Study completed-Reportage in Progress

A Randomized, Double-blind, Placebo-Controlled, Multicenter Study to Assess Efficacy and Tolerability of Oral Treprostinil on Healing Ischemic Digital Ulcers in Patients with Systemic Sclerosis.

The primary objective of this study is to assess efficacy of oral treprostinil in healing active ischemic digital ulcers in patients with SSc.

Role: International PI

Direct Sponsor – OMERACT 2009-2015

Connective Tissue Disease Interstitial Lung Disease Special Interest Group – Rheumatology Chair

The purpose of this exercise is to develop a core set of outcome and response measures to facilitate future trials in parenchymal lung disease including IPF and ILD secondary to scleroderma.

Role: International Co-Chair, Chair- Development

Direct Sponsor – Scleroderma Research Foundation 2014-

CONQUER REGISTRY

This is an ongoing effort to develop a master platform protocol with randomized registry to facilitate drug development and proof of concept research in scleroderma.

Role: Medical Director

Direct Sponsor – American College of Rheumatology 2010-2015

Member Subcommittee for Revised Classification Criteria for Systemic Sclerosis

Direct Sponsor: Apricus/NexMed 2014-

A Phase 2A Double-Blind, Randomized, Placebo-Controlled Crossover Trial Evaluating the Safety, Tolerability and Acute Peripheral Vascular Effects of Topical Alprostadil in Subjects with Raynaud's Phenomenon Secondary to Systemic Sclerosis.

Role: Clinical Lead

Direct Sponsor: Bayer Pharmaceuticals 2014-

A Randomized, Double-Blind, Placebo-controlled Phase II Study to Investigate the Efficacy and Safety of Riociguat in Patients with Diffuse Cutaneous Systemic Sclerosis

Role: Chair, Data Safety Monitoring Board

Direct Sponsor: Cytori 2015-

Scleroderma Treatment with Celution Processed Adipose Derived Regenerative Cells (STAR): A Randomized, Double-Blind, Placebo- Controlled Trial with Incomplete Crossover

Role: Chair, Data Safety Monitoring Committee

Direct Sponsor: Boehringer Ingelheim 2015-

A double blind, randomized, placebo-controlled trial evaluating efficacy and safety of oral nintedanib, 150 mg twice daily for 52 weeks (with follow up until last patient out up to a maximum of 2 years) in patients with Systemic Sclerosis associated interstitial lung disease

Role: Chair, Data Monitoring Committee

2009- present	United Therapeutics – prevention of PAH in scleroderma
2010-2011	Pfizer, Inc. – sitaxsentan in scleroderma
2010-2014	NexMed/Apricus – topical alprostadil in Raynaud phenomenon
2010-present	Sanofi-Aventis – therapies for fibrosing disorders
2010-present	Pfizer, Inc. – PDE5I for Raynaud phenomenon

Visiting Professorships, Seminars, and Extramural Invited Presentations 2013-Present

- Rochester Medical School, Rheumatology Grand Rounds, “Pulmonary Hypertension in Scleroderma” 2013
- World Scleroderma Foundation, Monteriggioni, Italy “Successes and Failures of our Community in Randomized Controlled Trials” 2013
- German Society for Rheumatology, Mannheim, Germany “Assessment and management of ILD”
- EULAR 2013 Madrid, Spain “Targeting vascular remodeling and PAH in systemic sclerosis”
- American College of Rheumatology 2013 Annual Meeting, San Diego, CA “ Meet the Professor – PAH in the Connective Tissue Diseases”
- 3rd World Congress on Systemic Sclerosis, 2014, Rome, Italy “The role of corticosteroids in systemic sclerosis”
- American College of Rheumatology 2014 Annual Meeting, Boston, MA “ Meet the Professor – PAH in the Connective Tissue Diseases”
- Scleroderma Clinical Trials Consortium Annual Meeting, Boston, MA “Registries and their Uses in Scleroderma Research” 2014
- NIAMS Roundtable on Scleroderma 2015 “Advancing Potential Drugs to Patient Care” – Trial Design and Outcome measures
- Raynaud’s and Scleroderma Association/Scleroderma Society (UK), Cambridge, UK “ The challenge of clinical trials in scleroderma” 2015
- International Workshop on Scleroderma Research, Cambridge, UK “Keynote Lecture: Early stage scleroderma trial design in the 21st century” 2015

Bibliography

Recent Peer-Reviewed Publications:

200. Denton CP, Avouac J, Behrens F, Furst DE, Foeldvari I, Humbert M, Huscher D, Kowal-Bielecka), Matucci-Cerinic M, Nash P, Optiz CF, Pittrow D, Rubin LJ, Seibold JR, Distler O: Systemic sclerosis associated pulmonary hypertension: why disease specific composite endpoints are needed. *Arthritis Res Therapy* 13: 114-117, 2011.
201. Rubenfire M, Huffman M, Krishnan SM, Seibold JR, Schioppa E, McLaughlin VV: Survival in systemic sclerosis with pulmonary arterial hypertension has not improved in the modern era. *Chest* 06/2013; DOI:10.13781/chest. 12-0653.
202. Hinchcliff M, Pennison MJ, Zimmerman JW, Bellam N, Zeng Q, Huang C-C, Pope R, Sadim M, Wolf W, Edberg J, Kimberly R, Mayes MD, Assassi S, Baron M, Boin F, Frech T, Furst DE, Hummers L,

- Khanna D, Molitor J, Phillips K, Reveille J, Schioppa E, Segal B, Seibold JR, Silver R, Simms R, Steen V, Wigley F, Zhang K, Li J, Yi N, Varga J, Pasche B: TGFBR1 and risk for systemic sclerosis (in review).
203. Oddis CV, Reed AM, Aggarwal R, Rider LG, Ascherman D, Levesque MC, Barohn RJ, Feldman BM, Koontz DC, Fertig N, Kelley SS, Pryber SL, Miller FW, Rockette HE and the Rituximab in Myositis Study Group [Seibold JR]: Rituximab in the treatment of refractory adult and juvenile dermatomyositis and adult polymyositis. A randomized, placebo-phase trial. *Arthritis Rheum* 65: 314-324, 2013.
 204. Roth MD, Tseng C-H, Clements PJ, Furst DE, Tashkin DP, Goldin JG, Khanna D, Kleerup EC, Li N, Elashoff D, Elashoff RM, Scleroderma Lung Research Group [Seibold JR]: Predicting treatment outcomes and responder subsets in scleroderma-related interstitial lung disease. *Arthritis Rheum* 63: 2797-2808, 2011.
 205. Fransen J, Johnson SR, van den Hoogen F, Baron M, Allanore Y, Carreira P, Czirjak L, Denton C, Distler O, Furst D, Gabrielli A, Herrick A, Inanc M, Kahaleh B, Kowal-Bielecka O, Medsger T, Mueller-Ladner U, Riemekasten G, Sierakowski S, Valentini G, Veale D, Vonk M, Walker U, Chung L, Clements P, Collier D, Csuka ME, Jimenez S, Merkel P, Seibold JR, Silver R, Steen V, Tyndall A, Matucci-Cerinic M, Pope JE, Khanna D: Items for revised classification criteria in systemic sclerosis: results of a consensus exercise with the EULAR/ACR working group for classification criteria in systemic sclerosis. *Arthritis Care Research* 64: 351-357, 2012 .
 206. Shah AA, Schioppa E, Hummers LK, Wade M, Phillips K, Anderson C, Wise R, Boin F, Seibold JR, Wigley F, Rollins KD: Open label study of escalating doses of oral treprostinil diethanolamine in patients with systemic sclerosis and digital ischemia: pharmacokinetics and correlation with digital perfusion. *Arthritis Res Ther* 2013 Apr 18; 15 (2):R54. Doi:10.1186/ar4216.
 207. Denton CP, Avouac J, Behrens F, Furst DE, Foeldvari I, Humbert M, Huscher D, Kowal-Bielecka O, Matucci-Cerinic M, Nash P, Opitz C, Pittrow D, Rubin LJ, Seibold JR, Distler O: Systemic sclerosis associated pulmonary hypertension: why disease-specific composite endpoints are needed. *Arthritis Res Therapy* 13: 114-117, 2011.
 208. Sharif R, Fritzler MJ, Mayes MD, Gonzalez EB, McNearney TA, Draeger H, Baron M, Furst DE, Khanna D, Del Junco DJ, Molitor J, Schioppa E, Phillips K, Seibold JR, Silver RM, Simms RW, Perry M, Rojo C, Charles J, Zhou X, Agarwal SK, Reveille J, Assassi S, Arnett FC: Anti-fibrillar antibody in African American patients with systemic sclerosis: Immunogenetics, clinical features, and survival analysis. *J Rheumatol* 38: 1622-1630, 2011.
 209. Hung EW, Mayes MD, Sharif R, Assassi S, Machicao VI, Hosing C, St. Clair W, Furst DE, Khanna D, Forman S, Mineishi S, Phillips K, Seibold JR, Bredeson C, Csuka ME, Nash RA, Wener M, Simms R, Ballen K, LeClercq S, Storek J, Goldmuntz E, Welch B, Keyes-Elstein L, Castina S, Crofford LJ, McSweeney P, Sullivan K: Gastric antral vascular ectasia (GAVE) and its clinical correlates in early diffuse systemic sclerosis. *J Rheumatol* 2013; 40: 455-60..
 210. Kowal-Bielecka O, Avouac J, Pittrow D, Huscher D, Behrens F, Denton CP, Foeldvari I, Humbert M, Matucci-Cerinic M, Nash P, Opitz CF, Rubin LJ, Seibold JR, Strand V, Furst DE, Distler O: Analysis of the validation status of Quality of Life and Functional Disability Measures in pulmonary hypertension related to systemic sclerosis: results of a systematic literature analysis by the Expert Panel on Outcome Measures in pulmonary arterial hypertension related to Systemic Sclerosis. *J Rheumatol* 38: 2419-2427, 2011.
 211. Saketkoo LA, Mittoo S, Huscher D, Dellaripa PF, Distler O, Flaherty K, Frankel S, Khanna D, Oddis CV, Denton CP, Fischer A, Kowal-Bielecka OM, LeSage D, Merkel P, Phillips K, Pittrow D, Swigris J, Antoniou K, Baughman B, Castellino F, Christmann RB, Christopher-Stine L, Collard HR, Cottin V, Danoff S, Highland K, Hummers L, Shah A, Kim DS, Lynch D, Miller F, Proudman S, Richeldi L,

- Ryu J, Sandorfi N, Sarver C, Wells AU, Strand V, Matteson EL, Brown KK, Seibold JR: Connective Tissue Disease Related Interstitial Lung Diseases and Idiopathic Pulmonary Fibrosis: Provisional Core-Sets of Domains and Instruments for Use in Clinical Trials. *Thorax* Published Online First March 20, 2014 doi:10.1136/thoraxjnl-2013-204202 and 2014; 69: 428-436.
212. Khanna D, Furst DE, Allanore Y, Bae S, Bodukam V, Clements PJ, Cutolo M, Seibold JR, Cutolo M, Czirjak L, Denton CP, Distler O, Walker UA, Matucci-Cerinic M, Mueller-Ladner U, Seibold JR, Singh M, Tyndall A: Twenty-two Points to Consider for clinical trials in systemic sclerosis, based on EULAR Standards. *Rheumatology* 08/2014 DOI: 10.1093/rheumatology/keu288.
 213. Khanna D, Tan M, Furst DE, Hill N, McLaughlin V, Silver RM, Steen VD, Langer A, Seibold JR: Recognition of pulmonary hypertension in the community – lessons from a Quality Enhancement Research Initiative. *Clin Exp Rheumatol* 2014; 32(6): 21-27.
 214. Furst DE, Khanna D, Matucci-Cerinic M, Clements P, Steen VD, Pope J, Merkel P, Foeldvari I, Seibold J, Pittrow D, Polisson R, Strand V: Systemic sclerosis – continuing progress in developing clinical measures of outcome. *J Rheumatol* 2013; 34: 1194-2000.
 215. Schiopu E, Impens AJ, Gross BH, Phillips K, Zubkus D, McLaughlin VV, Seibold JR, Rubenfire M: Pulmonary artery diameter as measured by chest computed tomography correlates with pulmonary hypertension in patients with systemic sclerosis irrespective of parenchymal lung involvement (in review).
 216. Baes S, Saggarr R, Bolster MB et al for the PHAROS Group [Seibold JR]: Baseline characteristics and follow-up in patients with normal haemodynamics versus borderline mean pulmonary arterial hypertension in systemic sclerosis: results from the PHAROS registry. *Ann Rheum Dis* 2012, 71: 1335-1342.
 217. Coghlan JG, Denton CP, Gruenig E, Bonderman D, Distler O, Khanna D, Mueller-Ladner U, Pope JE, Vonk MC, Doelberg M, Chadha-Boreham H, Heizl H, Rosenberg D, McLaughlin VV, Seibold JR: Evidence-based detection of pulmonary arterial hypertension in systemic sclerosis, the DETECT study. *Ann Rheum Dis* doi: 10.1136/annrheumdis-2013-203301 and 2014; 73: 1340-1349.
 218. Lenderking WR, Denton CP, Matucci-Cerinic M, Woodward TC, Safikhani S, Fernandes AW, Czirjak L, Khanna D, Mayes M, Pope J, Seibold JR, Furst DE: Development of a Scleroderma Responder Index (ScRI): results of a Delphi panel study (in review).
 219. van de Hoogen F, Khanna D, Fransen J, Johnson S, Baron M, Tyndall A, Matucci-Cerinic M, Naden R, Medsger TA, Carreira P, Riemekasten G, Clements PJ, Denton CP, Distler O, Allanore Y, Furst DE, Gabrielli A, Mayes M, van Laar J, Seibold JR, Czirjak L, Steen V, Inanc M, Kowal-Bielecka O, Mueller-Ladner U, Valentini G, Veale D, Vonk M, Walker U, Chung L, Collier D, Csuka ME, Fessler B, Guidici S, Herrick A, Hsu VM, Jimenez S, Kahaleh B, Merkel P, Sierakowski S, Silver RM, Simms R, Varga J, Pope J: Classification criteria for systemic sclerosis: An ACR-EULAR Collaborative Initiative. *Arthritis Rheum* 65: 2737-2747, 2013.
 220. Khanna D, Seibold JR, Goldin J, Tashkin D, Furst DE, Wells A: Points to consider for clinical trials in systemic sclerosis – interstitial lung disease. *Rheumatology* (in press).
 221. Humbert M, Singh M, Furst DE, Khanna D, Seibold JR: Pulmonary hypertension related to systemic sclerosis: Points to Consider for Clinical Trials (in review).
 222. Denton CP, Matucci-Cerinic M, Seibold JR: Strategy in Patient Management, EULAR Online Course on Systemic Sclerosis. <https://b-com.mci-group.com/Registration/EULARSSC12.aspx>.
 223. Avouac J, Huscher D, Furst DE, Opitz CF, Distler O, Allanore Y for the EPOSS Group [Seibold JR]: Expert consensus for performing right heart catheterization for suspected pulmonary arterial hypertension in systemic sclerosis: a Delphi consensus study with cluster analysis. *Ann Rheum Dis* doi:10.1136/annrheumdis-2012-202567.

224. Riemekasten G, Berger MO, Seibold JR: Management: Vasodilators. EULAR Online Course on Systemic Sclerosis. <https://b-com.mci-group.com/Registration/EULARSSC12.aspx>.
225. Furst DE, Pope JE, Seibold JR, Bombardieri S, Denton CP, Distler O, Kahaleh B, Kennedy AT, Khanna D, Lafyatis R, Tyndall AD, Matucci-Cerinic M: Progress and priorities in systemic sclerosis : the next ten years. *Rheumatology* (in press)
226. Wiese AB, Berrocal VJ, Furst DE, Seibold JR, Merkel PA, Mayes MD, Khanna D: Correlates and responsiveness of skin and musculoskeletal disease in early diffuse systemic sclerosis. *Arthritis Care Res* 04/2014; doi: 10.1002/acr.22339 and 2014; 66: 1731-1739.
227. Visovatti S, Distler O, Coghlan JG, Denton CP, Gruenig E, Bonderman D, Mueller-Ladner U, Pope JE, Vonk MC, Seibold JR, Torres-Martin J-V, Doelberg M, Chadha-Boreham H, McLaughlin VV, Khanna D: Borderline pulmonary arterial pressure in scleroderma: Are systemic sclerosis patients with borderline pulmonary arterial pressure a unique subset? A post-hoc analysis of the DETECT study. *Arthritis Res Ther* 2014; Dec 10;16(6):493 [Epub ahead of print].
228. Brenner B, Kafaja S, Furst DE, Berrocal V, Merkel PA, Seibold JR, Mayes MD, Khanna D: Reliability, validity, and responsiveness to change of the Saint George's Respiratory Questionnaire in early diffuse cutaneous systemic sclerosis. *Rheumatology* (in press).
229. Khanna D, Gladue H, Seibold JR: Clinical outcome measures in Raynaud phenomenon. In Wigley FM, Herrick A, Flavahan NA , Raynaud Phenomenon – A Guide to Pathogenesis and Treatment, Springer, 2014, pp 279-286.
230. Johnson S, Naden RP, Fransen J, van den Hoogen F, Pope JE, Baron M, Tyndall A, Matucci-Cerinic M, Denton CP, Distler O, Gabrielli A, van Laar JM, Mayes M, Steen V, Seibold JR, Clements P, Medsger TA, Carreira P, Riemekasten G, Chung L, Fessler BJ, Merkel PA, Silver R, Varga J, Allanire Y, Mueller-Ladner U, Vonk MC, Walker UA, Capelli S, Khanna D: Systemic sclerosis classification criteria: developing methods for multi-criteria decision analysis with 1000 minds. *J Clin Epidemiol* 2014; 67: 706-714.
231. Walker KM, Pope J, Scleroderma Clinical Trials Consortium [Seibold JR], Canadian Scleroderma Research Group: Treatment of systemic sclerosis complications: what to use when first-line treatment fails- a consensus of systemic sclerosis experts. *Sem Arthr Rheum* 2012; 42: 42-55.
232. Khanna D, Mittoo S, Aggarwal R, Proudman S, Dalbeth N, Matteson E, Brown K, Flaherty K, Wells AU, Seibold JR, Strand V: Connective Tissue Disease-Associated Interstitial Lung Diseases – Report from OMERACT CTD-ILD Working Group. *J Rheumatol* DOI:10.3899/jrheum.141182.
233. Khanna D, Berrocal VJ, Giannini E, Seibold JR, Merkel PA, Mayes MD, Baron M, Clements PJ, Steen VD, Assassi S, Schioppa E, Phillips K, Simms R, Allanore Y, Denton CP, Distler O, Johnson S, Matucci-Cerinic M, Pope JE, Proudman S, Siegel J, Wong WK, Wells AU, Furst DE: Development and initial validation of a composite response index for clinical trials in early diffuse cutaneous systemic sclerosis (in review).
234. Denton CP, Matucci-Cerinic M, Seibold JR: Strategy in Patient Management, EULAR Online Course on Systemic Sclerosis. <https://b-com.mci-group.com/Registration/EULARSSC12.aspx>. (2015)
235. Riemekasten G, Becker MO, Seibold JR: Management: Vasodilators. EULAR Online Course on Systemic Sclerosis. <https://b-com.mci-group.com/Registration/EULARSSC12.aspx>.(2015)
236. Wallace B, Kafaja S, Furst DE, Berrocal VJ, Merkel PA, Seibold JR, Mayes MD, Khanna D: Reliability, validity and responsiveness to change of the Saint George's Respiratory Questionnaire in early diffuse cutaneous systemic sclerosis. *Rheumatol* doi:10.1093/rheumatology/keu456
237. Seibold JR, Wigley FM, Schioppa E, Denton CP, Silver RM, Steen VD, Domsic R, Medsger TA, Mayes MD, Chatterjee S, Chung L, Csuka ME, Khanna D, Collier D, Frech TM, Molitor JA, Rothfield N, Herrick AL, Simms R, Pope JE, Baron M, Hsu VM, Peng SL, Spiera R, Fessler B, Kahaleh B, Varga J,

- Rollins K, Arneson C, Laliberte K, Wade M: Digital ischemic ulcers in systemic sclerosis treated with oral treprostinil: a randomized, double-blind, placebo-controlled, multicenter study and open label follow up (in review).
238. Khanna D, Albera C, Fischer A, Khalidi N, Raghu G, Chung L, Chen D, Schiopus E, Tagilafferri M, Seibold JR, Gorina E: An open-label, randomized, Phase 2 study of the safety and tolerability of pirfenidone in scleroderma-associated interstitial lung disease (in review)
239. Khanna D, Berrocal VJ, Giannini E, Seibold JR, Merkel PA, Mayes MD, Baron M, Clements PJ, Steen VD, Assassi S, Schiopus E, Phillips K, Simms R, Allanore Y, Denton CP, Distler O, Johnson S, Matucci-Cerinic M, Pope JE, Proudman S, Siegel J, Wong WK, Wells AU, Furst DE: The American College of Rheumatology provisional composite response index for clinical trials in early diffuse cutaneous systemic sclerosis (in review).