

**9<sup>th</sup> International Workshop on Scleroderma Research**  
**Boston University**  
**August 5-9, 2006**

**Saturday, August 5<sup>th</sup>**

3-7pm                      CHECK-IN and REGISTRATION  
Conference Office @ 10 Buick St. Room G18

Evening                    The bar at the Eastern Standard, Hotel Commonwealth, 528  
Commonwealth Avenue (take Green Line inbound to Kenmore  
Square). – Unofficial event.

**Sunday, August 6<sup>th</sup>**

**8.15-8.30am              WELCOME    C Black and R Lafyatis**  
**Tribute to Joe Korn    R Simms**

**8.30-10                    Session I – Stem Cell Biology in Vascular and Fibrotic Disease**  
**Moderator: R Strieter**  
**Keynote: J Huard** - Regenerative medicine based on muscle derived stem cells: Potential for tissue regeneration and repair  
M Kuwana - Circulating endothelial progenitors as a therapeutic target for scleroderma vascular disease  
C Dong - Deficient repair - the culprit of scleroderma vascular lesion formation?

10-10.30                    Coffee Break

**10.30-12.30pm            Session II – Vascular Disease in Systemic Sclerosis**  
**Moderator: H Farber**  
**Keynote: M. Rabinovitch** - Infection, Inflammation Mts1 and Pulmonary Hypertension  
K Pritchard - D-4F, an ApoA-I mimetic, decreases inflammation and fibrosis and increases angiogenic responses in the myocardium of tight skin mice  
N Flavahan - The cold facts about peripheral vasospasm  
B Khaw - Molecular Imaging of vascular injury

12.30-1.30                    Lunch

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**1.30-3.30**

**Breakout Session I**

**A. Hurdles, networks, funding: Where should we be going? – CGS511**

**Moderator: S Jimenez/F Arnett**

E Gretz – National Institutes of Health

L Evnin – Scleroderma Research Foundation

E Gravallesse – Arthritis Foundation

J Blint – Scleroderma Foundation

**B. Animal Models – CGS505**

**Moderator: S Clark/H Gardner**

C Denton - Inducible disruption of TGFbeta signalling in fibroblasts in vivo

R Lafyatis – Complex cytokine regulation of Tsk1 pathology

P Christner/J Gentiletti – The fibrotic and immunologic facets of the Tsk2 mouse

C Feghali-Bostwick

S Sonnylal - CTGF overexpressing mice: A model for human scleroderma

3.30-4

Tea Break

**4-6**

**Session III – Inflammation and Vascular Injury**

**Moderator: B Kahaleh/S Schwartz**

**Keynote: P Libby** - Inflammation and the regulation of arterial extracellular matrix metabolism

O Distler - The impaired angiogenesis in systemic sclerosis

S Schwartz - The vasculopathy of scleroderma

S Jimenez – Role of allograft inflammatory factor 1 (AIF-1) in the pathogenesis of SSc vasculopathy

6-7.30

Welcome Reception – Agganis Arena Burke Club Room

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**Monday, August 7<sup>th</sup>**

- 8.30-10.00am**      **Session IV – Autoimmune Dysregulation in Systemic Sclerosis**  
**Moderator: A Marshak-Rothstein**  
**Keynote: A Rosen** -Autoimmunity in scleroderma  
T Radstake - Toll-like receptors in systemic sclerosis, new light on an ancient system!  
T Tedder - Are B lymphocytes contributing to systemic sclerosis pathogenesis?
- 10-10.30              Coffee Break
- 10.30-12.00pm**      **Session V – T-cell Regulation and Fibrosis**  
**Moderator: M Kuwana**  
**Keynote: R Flavell** - Regulation of immunity and autoimmunity by TGF $\beta$   
T MacDonald – Smad 7 control of TGF $\beta$  responses in the gut  
C Buckley – Defining a role for fibroblasts in the persistence of chronic inflammation
- 12-1                      Lunch
- 1-1.30**                      **Poster Viewing – 18<sup>th</sup> Floor Lounge, 10 Buick Street. Authors stand by posters**
- 1.30-3**                      **Poster Session – Dance Theatre - Top ranking posters orally presented**  
**Moderator: P Clements**  
P Fernandez – Adenosine modulates dermal fibrogenesis  
A Bujor - Akt positively regulates collagen in dermal fibroblasts via a Smad3 independent pathway  
A Ghatnekar - The Role of Gata3 as a regulator of ECM homeostasis in SSc  
A Ghosh - Peroxisome proliferator-activated receptor- $\gamma$  (PPAR- $\gamma$ ) inhibits collagen gene expression: implications for Fibrosis  
Y Pare - CD4<sup>+</sup> CD8<sup>+</sup> double positive (DP) T cells with very high IL-4 production potential are present in lesional skin of systemic sclerosis (SSc) patients

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T Rodriguez-Reyna - Rate of skin thickness progression in early systemic sclerosis with diffuse cutaneous involvement: Associations with organ involvement and 5 year survival  
D Abraham (V Rajkumar) – PDGFB receptor activation is essential for fibroblast and pericyte recruitment during cutaneous wound healing

3-3.15 Tea break

**3.15-4.45 Session VI – Fibrotic Mediators I**

**Moderator: T MacDonald**

**Keynote:** P ten Dijke - TGF $\beta$  receptor signal transduction  
M Trojanowska - Fibroblast signaling in scleroderma  
J Pearson - Endothelin and fibrosis

5.00-7

**Breakout Session II**

**A. Immunomodulators and Fibrotic Disease – CGS511**

**Moderator: F Wigley**

New therapeutic agents in scleroderma:

- C Denton – Mycophenolate mofetil in diffuse scleroderma
- E Smith – Sirolimus (Rapamycin)
- R Silver - Anti-chemokine therapy (MCP-1/CCR2)
- L Hummers – Intravenous gammaglobulin
- R Lafyatis – Anti-B cell therapy (rituximab)

**B. Chronic graft vs. host disease modeling of SSc – CGS505**

**Moderator: A Gilliam**

A Gilliam - Triggers of fibrosing disease in GVHD - why sclerodermatous disease rather than lupus-like epithelial injury?  
W Shlomchik - Roles of antigen, antigen presenting cells and naive CD4 cells in murine chronic GVHD  
A Aliprantis - The role of T-bet in scleroderma and the molecular phenotype of murine chronic GVHD

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**Tuesday, August 8<sup>th</sup>**

- 8.30-10.30am**      **Session VII – Tissue Fibrosis: Cellular Pathogenesis**  
**Moderator: J Varga**  
T Krieg - Mechanism of fibroblast activation and the role of cell matrix contacts  
S Phan - Transcriptional regulation of myofibroblast differentiation  
H Chang - Site specific differentiation and positional memory of fibroblasts  
E Leof - TGF-beta signaling targets in fibrotic diseases
- 10.30-11              Coffee Break
- 11-12.30pm**        **Session VIII – Fibrotic Mediators**  
**Moderator: D Abraham**  
D Brigstock - Connective tissue growth factor: Structure, function, and role in fibrosis  
K Lyons - In vivo studies of CCN1/Cyr61 and CCN2/CTGF function in connective tissues and vasculature  
J Bonner - PDGF Signaling in Chronic Airway Remodeling and Fibrosis
- 12.30-1.30           Lunch
- 2-4**                    **Session IX – Pulmonary Fibrosis/Pathogenesis**  
**Moderators: A Wells/K Brown**  
**Keynote: J Elias** - Transgenic investigations of Th2 and TGF-beta1-induced tissue fibrosis  
V Thannickal - Novel insights into myofibroblast activation and survival  
N Kaminski - Microarray analysis of Human Lung Fibrosis - from profiles to pathways  
R Strieter - The role of circulating mesenchymal progenitor cells in pulmonary fibrosis
- 4-4.30                Tea Break

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**4.30-6.30**

**Breakout Session III – CGS511**

**A. How do we block TGF $\beta$  in scleroderma?**

**Moderator: M Trojanowska**

S Violette - The  $\alpha$ v $\beta$ 6 integrin: A novel therapeutic target for lung and kidney fibrosis

A Protter - TBA

J McPherson - Progress in the clinical development of anti-TGF-beta antibodies for the treatment of fibrotic diseases

**B. Gene expression profiles, what do they tell us? – CGS505**

**Moderator: C Feghali-Bostwick**

M Whitfield - Molecular portraits of scleroderma skin

F Tan - The next step after differential gene expression

H Gardner - Gene profiling in SSc: Fibroblasts forget where they came from and other results

7-9

BANQUET – Agganis Arena Burke Club Room

Speaker: Dr. Don Berwick

**Wednesday, August 9<sup>th</sup>**

**8.30-10.30am**

**Session X – Pulmonary Fibrosis/Clinical**

**Moderator: R Du Bois**

**Keynote: A Wells** - Pulmonary fibrosis in scleroderma: How clinical science can refine management

J Goldin - Quantitative Image Analysis of HRCT: predictor and measure of treatment efficacy

J Seibold - Clinical applications of endothelin antagonists in scleroderma

K Brown - Pulmonary fibrosis: Clinical insights into disease mechanism

10.30-10.45

Coffee Break

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- 10.45-12.30pm**      **Session XI – Genetic Analyses in Scleroderma**  
**Moderator: F Arnett**  
**Keynote: M Seldin** - Understanding and application of population genetic structure to problems in medicine and autoimmunity  
M Mayes - Scleroderma genetics: Where we are and where we want to be  
R Du Bois - Genetics of lung fibrosis: It's all in the genotype  
K Welsh - Scleroderma genetics worldwide: One disease, many incorrect papers- or many diseases? Do we have or can we get the answer?
- 12.30-1.30              Lunch
- 1.30-3.30              **Session XII– Outcome Measures**  
**Moderator: M Matucci-Cerinic**  
A Tyndall - Report from the EUSTAR Minimal Essential Data Set ( MEDS) data base  
D Furst - Progress at OMERACT toward combined measures of response in SSc: Renal disease, pulmonary disease and early efforts at a combined index of response  
D Khanna - Patient-reported outcomes in scleroderma  
P Merkel – Outcome measures for scleroderma clinical trials: A data-driven re-evaluation