

AN OVERVIEW OF THE RESEARCH ACROSS CANADA

“Thank you” to the inspiring and dedicated Canadian researchers and health care providers who are contributing to the development of ground-breaking interventions, improved diagnostic techniques and understanding of scleroderma. Though the disease may be rare, the community supporting quality of life improvement for people impacted by scleroderma provides a broad source of hope. Here is a sample of some of the outstanding research efforts taking place in our own country.

To read more about research across Canada, please visit the *Research section* of our website sclerodermie.ca/en/.

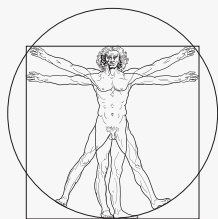
MONTREAL, QUEBEC



DR. JEAN-LUC SENEAL
CHUM Research Center
Scleroderma Research Chair
University of Montreal

The pathogenic roles of autoantibodies in scleroderma: At the request of the *Journal of Scleroderma and Related Disorders (JSRD)*, which is the only international medical journal

dedicated solely to scleroderma, this study analysed how autoantibodies in the blood of scleroderma patients contribute to disease mechanisms. It was concluded that anti-topoisomerase I is the single autoantibody with the most evidence in favor of a pathogenic role in scleroderma, followed by anticentromere autoantibody. A better understanding of how these autoantibodies contribute to scleroderma manifestations may lead to improved therapies.



The Vitruvian Man, 1492

**DR. HEENA MEHTA,
DR. MARIKA SARFATI**
CHUM Research Center
Immunoregulation Laboratory
University of Montreal

From bench to bedside to bench...

Recent acquisition of cutting-edge instrumentation enables the comprehensive study of the immune profile in blood and at barrier tissues. Establishing a link between different clinical presentations of scleroderma and the immune landscape at a given timepoint of the disease will open the doors for identifying new immune therapeutic targets and novel clinical management strategies.



DR. SABRINA HOA
CHUM Research Center
University of Montreal

Association between immunosuppressive therapy and course of mild interstitial lung disease in systemic sclerosis (SSc-ILD): The objective of this study was to determine whether use of immunosuppressive drugs,

namely cyclophosphamide or mycophenolate mofetil, was associated with an improved course of lung disease in patients with normal or mildly reduced lung function. In this setting, exposure to these immunosuppressive drugs at the beginning of the study was associated with higher forced vital capacity values and a lower risk of progression among patients with mild interstitial lung disease at two years of follow-up. These data suggest a possible window of opportunity to preserve lung function in mild SSc-ILD.



DR. MARIE HUDSON
Lady Davis Institute for Medical
Research, Montreal Jewish
General Hospital

Generation of a Core Set of Items to Develop Classification Criteria for Scleroderma Renal Crisis Using Consensus Methodology: This study helped to generate a method of

classification for scleroderma renal crisis (SRC), which will be used in future phases of this project to develop classification criteria for scleroderma renal crisis.

Research across Canada



DR. CELIA GREENWOOD

**Lady Davis Institute for Medical Research
McGill University**

Whole-genome bisulfite sequencing in systemic sclerosis provides novel targets to understand disease pathogenesis:

This study investigated the role of DNA methylation in SSc and will be used for future studies on the same subject.

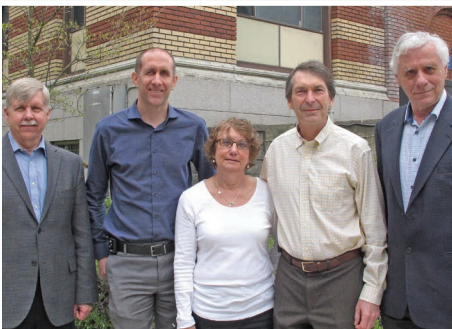


DR. BRETT THOMBS

McGill University

Scleroderma Patient-centered Intervention Network (SPIN), maintains an international patient database used to study psychosocial and rehabilitative online programs and toolkits to improve the health-related quality of life of patients living with scleroderma.

VANCOUVER, BRITISH COLUMBIA



DR. JAMES DUNNE, DR. KEVIN KEEN

**Scleroderma Clinic, Mary Pack Arthritis Program,
Vancouver Coastal Health Authority (Vancouver General Hospital)
Scleroderma Lung Clinic & Combined Scleroderma Pulmonary Arterial
Hypertension Clinic, Pacific Lung Health Centre, Providence Health Care
(St. Paul's Hospital)
Providence Health Care Research Institute (St. Paul's Hospital)**

Scleroderma Association of B.C. Research Project is a community-based patient-oriented study to discover the impact of certain RNA molecules on biochemical processes at the cellular level with the goal of targeted therapeutic

interventions to correct the disturbed cycle of regeneration in heart, lung, and skin tissues in scleroderma and lung tissue in idiopathic pulmonary fibrosis.

WINNIPEG, MANITOBA



DR. ADA MAN

University of Manitoba

Development and validation of a patient-reported outcome instrument for skin involvement in patients with systemic sclerosis: This study developed a patient questionnaire to assess the skin-related quality of life in patients with systemic sclerosis. It

shows reliability and validity and is complementary to existing measures of SSc skin involvement with emphasis on the patient's experience.

HALIFAX, NOVA SCOTIA



DR. EVELYN SUTTON

Dalhousie University

Clinical correlates of faecal incontinence (FI) in systemic sclerosis: identifying therapeutic avenues: This study established the prevalence and severity of FI in SSc, its association with other intestinal manifestations and potential predictors of FI, and its

impact on quality of life. FI was found to be common and often severe in SSc. Loose stools, SIBO, constipation and urinary incontinence were strongly associated with FI. Other than targeting anorectal dysfunction, concomitant treatment of clinical correlates could lead to improvement in FI and quality of life in SSc.

Research across Canada

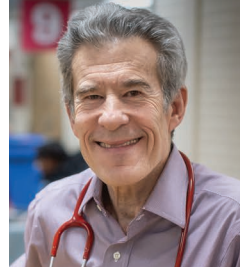
TORONTO, ONTARIO



DR. SINDHU R. JOHNSON
Toronto Scleroderma
Program - University Health
Network & Sinai Health Systems

Nintedanib for Systemic Sclerosis-Associated Interstitial Lung Disease:
An international, multicenter study that found treatment with nintedanib (OFEV®) slows progression of

scleroderma-associated interstitial lung disease. Thanks to this study, OFEV® was approved by Health Canada and is now available in Canada.



**DR. RONALD LAXER,
DR. ELENA POPE**
Toronto Hospital for Sick Children

Developing comparative effectiveness studies for a rare, understudied pediatric disease: This study investigated the feasibility of conducting a comparative effective-

ness study for juvenile localized scleroderma, a rare pediatric disease, for which there is limited evidence on best therapy.

LONDON, ONTARIO



DR. JANET POPE
St. Joseph's Health Care, London ON
Western University, London ON (London Health Sciences Centre)

Changes in skin score in early diffuse cutaneous systemic sclerosis are associated with changes in global disease severity: This study determined that at 1 and 2 years, skin scores showed overall improvement in early diffuse cutaneous scleroderma patients, improving prognosis and quality of life.

CALGARY, ALBERTA



DR. MARVIN FRITZLER
University of Calgary

Autoantibodies to a novel Rpp38 (Th/To) derived B-cell epitope are specific for systemic sclerosis and associate with a distinct clinical phenotype: This study screened certain proteins that target autoantibodies in scleroderma patients, and evaluated their clinical

relevance. Detection of antinuclear antibodies and specific autoantibodies is important in the diagnosis and classification of SSc. The new RNA P protein holds promise to increase the sensitivity in the detection of autoantibodies, enhancing the diagnosis of SSc.



DR. JAN STOREK
University of Calgary

Myeloablative Autologous Hematopoietic Stem Cell Transplantation for Severe Scleroderma: Long-Term Outcomes 6-11 Years after Entry on a Randomized Study Comparing Transplantation and Cyclophosphamide: This study reports

subject survivor status, late effects and outcomes 6-11 years after the Scleroderma: Cyclophosphamide or Transplantation (SCOT) trial. This follow-up analysis demonstrates the clinical benefits of stem cell transplantation (HSCT) remain 6-11 years after the study. Survival and functional status were significantly better with HSCT, and continuing control of scleroderma was demonstrated by 92% of transplant survivors remaining free of disease.