Limited Scleroderma
Speaker: Dr. Janet Pope

Learning objectives:
1. To form a differential diagnosis of systemic sclerosis (SSc) mimickers.
2. To identify the types of morphea.
3. To discuss treatment and understand that there are not proven treatments for generalized morphea.

Abstract:
Systemic sclerosis (SSc) can be confused with other scleroderma-like skin disorders, such as eosinophilic fasciitis (EF), generalized morphea, scleromyxedema and nephrogenic systemic fibrosis. In most of these conditions, Raynaud’s phenomenon, sclerodactyly, and nailfold capillary changes can occur. However, the presence of additional clinical features, such as esophageal dysmotility, renal involvement, and musculoskeletal symptoms, can help to distinguish between these entities. The differential diagnosis of SSc mimickers can be challenging, and accurate recognition is crucial for timely and appropriate management.

Renal Hypertension Complication
Speaker: Dr. Marie Hudson

Learning objectives:
1. To review the role of angiotensin converting enzyme (ACE) inhibitors in systemic sclerosis (SSc).
2. To become familiar with the preliminary results of an ongoing study designed to determine whether SSc patients with incident scleroderma renal crisis (SRC) taking ACE inhibitors prior to SRC onset have worse outcomes.

Abstract:
Scleroderma renal crisis (SRC) is an infrequent, but life-threatening complication of systemic sclerosis (SSc). It was previously associated with significant morbidity including chronic renal failure and dialysis, and high mortality. However, since the advent of angiotensin converting enzyme (ACE) inhibitors, the outcome of SRC has improved dramatically. There is also a perception among experts that the incidence of SRC has fallen over the past few years. This is thought to be due in part to the more liberal use of ACE inhibitors to treat Raynaud’s phenomenon and hypertension in SSc.

Given the benefits of ACE inhibitors and the perceived decrease in incidence in SRC, some experts have advocated the use of prophylactic ACE inhibitors even in the absence of Raynaud’s or hypertension. However, others have argued that there is no clear rationale for this since it has been demonstrated that most SSc patients do not have hyperreninemia prior to the onset of SRC. In addition, recent retrospective data in patients with SRC suggest that ACE inhibitors prior to the onset of SRC may have worse outcomes than those not taking these drugs. This has been hypothesized to be due to the fact that those taking ACE inhibitors may have normotensive SRC, thus delaying diagnosis in these patients.

Given the uncertainty surrounding the use of ACE inhibitors prior to the onset of SRC, a study was conducted to determine whether patients with SSc and who presented with incident SRC while taking ACE inhibitors immediately prior to the onset of SRC have worse outcomes (defined as dialysis dependence or death at one year) than those not taking these drugs prior to SRC onset. This study is a prospective, international cohort study of SRC subjects identified through an ongoing web-based survey. Every second Friday afternoon, an e-mail is sent to 589 participating physicians (worldwide) to identify new cases of SRC. Data on patient demographic and disease characteristics, as well as exposure to ACE inhibitors, is collected. A simple follow-up case report form is then sent to recruiting physicians one year after a patient is identified. The primary outcome of interest is death or dialysis dependence at one year after SRC onset, comparing patients exposed or not exposed to ACE inhibitors prior to SCR onset.

Fifteen months after the start of the survey, 76 incident cases of SRC (mean age 53 years, 68% women, 68% diffuse SSc, and median disease duration since the onset of the first non-Raynaud’s symptom 1.5 years) were identified. Of these, 66 (87%) had a hypertensive SRC and 10 (13%) a normotensive SRC. Twenty-two percent (22%) of the patients were taking an ACE inhibitor immediately prior to the onset of the SRC. Over 50% of the patients were also taking glucocorticoids immediately prior to SRC onset, at a mean dose of 17 mg/day of prednisone (or its equivalent). To date, we have collected one-year follow-up data on approximately one-third of the cohort. Of these, over 50% of patients have died or remain on dialysis at one year.

The results of this ongoing, international, cohort study are likely to provide new insights into the role of ACE inhibitors in patients with SSc.

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3. To become familiar with the preliminary results of an ongoing study designed to determine whether SSc patients with incident scleroderma renal crisis (SRC) taking ACE inhibitors prior to SRC onset have worse outcomes.
4. To discuss the association between morphea and other autoimmune diseases.

Abstract:
Systemic sclerosis (SSc) can be confused with other scleroderma-like skin disorders, such as eosinophilic fasciitis (EF), generalized morphea, scleromyxedema and nephrogenic systemic fibrosis. In most of these conditions, Raynaud’s phenomenon, sclerodactyly, and nailfold capillaries...
Genetics
Speaker: Dr. Rae Yeung

Learning objectives:
1. To review current knowledge of the biologic basis of juvenile idiopathic arthritis (JIA).
2. To describe methodology for studying genetic basis of disease.
3. To present a new international JIA research consortium aimed at understanding the biologic basis for JIA.

Abstract:
Early aggressive therapy would reverse morbidity and long-term disability in childhood arthritis, but is restricted by poor capacity to detect early disease and predict outcomes.

Outcomes
Speaker: Dr. Ciaran Duffy

Learning objectives:
1. To review current knowledge and paradigms of studying outcomes for juvenile idiopathic arthritis (JIA).
2. To review the development of the Canadian JIA research cohort Research on Arthritis in Canadian Children-Emphasizing Outcomes (ReACCh Out) study.
3. To describe the short-term outcomes and predictors of outcome in the ReACCh Out cohort.

Abstract:
The Canadian Alliance of Pediatric Rheumatology Investigators (CAPRI) has developed a national research network that addresses outcomes in juvenile idiopathic arthritis (JIA). With strong input from an excellent steering committee and the collaboration of over 40 investigators from coast-to-coast, an initial study called Research on Arthritis in Canadian Children-Emphasizing Outcomes (ReACCh Out) has enrolled more than 1,500 patients with new-onset JIA. This study aims to permit a better understanding of the course and outcome of the disease, and to identify, at an early stage, the factors associated with and predictive of better outcomes. This study has also laid the foundation for studies focused on the biologic basis of the disease. One such study, Biologically-based Outcome Predictors (BBOP), seeks to identify environmental, genetic and biologic factors that might be predictive of certain disease outcomes, particularly health-related quality of life.

A recent additional focus is on exercise and physical activity in patients with JIA, the Linking Exercise, Physical Activity and Pathophysiology in JIA (LEAP) study.

Juvenile Idiopathic Arthritis (JIA)

Chairs: Dr. Lori Tucker (CRA) and Dr. Vicente Baca Ruiz (MCR)

Thus, to improve outcome in childhood arthritis, there is a need for a combined approach from bench-to-bedside to identify predictors of outcome to guide clinical decisions involving use of potentially efficacious, but often costly, toxic and, possibly, unneeded therapies. Recent introduction of new drugs targeting biologic molecules and their associated dramatic improvements in clinical outcomes have shed light on the need to systematically characterize the underlying biology causing disease to aid in decision making.

Understanding the pathobiology and the genetics directing these processes will allow a more rational approach to develop a unique and personalized care plan for each individual. A stepwise process has been initiated to build an international research consortium to achieve these goals.

Scleromyxedema (also known as papular mucinosis) is the most common of these conditions. It is characterized by mucin deposition with fibrosis often associated with a monoclonal gammopathy.