ANNUAL UPDATE:
Top 5 for Pediatric Rheumatology in 2014

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Disclosures

- Roche, AbbVie – advisory boards
Learning Objectives

• To describe new and important developments in Pediatric Rheumatology relevant to Adult Rheumatologists in Ontario
EXTENDED REPORT

The outcomes of juvenile idiopathic arthritis in children managed with contemporary treatments: results from the ReACCh-Out cohort

Jaime Guzman,1 Kiem Oen,2 Lori B Tucker,1 Adam M Huber,3 Natalie Shiff,4 Gilles Boire,5 Rosie Scuccimarra,6 Roberta Berard,7 Shirley M L Tse,8 Kimberly Morishita,1 Elizabeth Stringer,3 Nicole Johnson,9 Deborah M Levy,8 Karen Watanabe Duffy,10 David A Cabral,1 Alan M Rosenberg,4 Maggie Larché,11 Paul Dancey,12 Ross E Petty,1 Ronald M Laxer,8 Earl Silverman,8 Paivi Miettunen,9 Anne-Laure Chetaille,13 Elie Haddad,14 Kristin Houghton,1 Lynn Spiegel,8 Stuart E Turvey,1 Heinrike Schmeling,9 Bianca Lang,3 Janet Ellsworth,15 Suzanne Ramsey,3 Alessandra Bruns,5 Sarah Campillo,6 Susanne Benseler,9 Gaëlle Chédeville,6 Rayfel Schneider,8 Rae Yeung,8 Ciarán M Duffy,10 for the ReACCh-Out investigators
#1 – Outcomes of JIA – ReACCh-Out

- Prospective longitudinal JIA inception cohort
- Consecutive patients recruited at 16 Canadian sites
- January 2005 – December 2010
- Study data collection every 6 months
- 1104 patients in this analysis
Results: Active Joint Count = 0
Results: Inactive Disease
Results: Disease Remission
#2 – Biologic therapy for systemic onset JIA (sJIA)

Immunobiology of this disease is different!

- M = F, any age
- Spiking daily fevers x > 2 weeks
- **Systemic features**: rash, adenopathy, organomegaly, pericarditis, anemia, leukocytosis, thrombocytosis
- Polyarthritis usually within 3 months
Randomized Trial of Tocilizumab in Systemic Juvenile Idiopathic Arthritis

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Randomized, double-blind, placebo controlled
N= 112

ACR 30 + Absence of fever

12 weeks

Percent

85

Tocilizumab
Placebo

ORA - Ontario Rheumatology Association
#2 - Tocilizumab in sJIA - Results

52 week outcomes

- ACR 70: 80%
- ACR 90: 59%
- No arthritis: 48%
- Steroids stopped: 52%
#3 – More biologics for sJIA!

Administered monthly
Half Life is 3-4 weeks

Administered daily
Half Life is 4-6 hours

Lachmann H, Arthritis Rheum 2011
Two Randomized Trials of Canakinumab in Systemic Juvenile Idiopathic Arthritis

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#3 - Canakinumab in sJIA

### Trial #1 – Active Disease
(total N=83)
- 2 – 19 years old
- Active sJIA on steroids
- Double blind, placebo controlled
- **Outcome:** JIA ACR 30 response plus resolution of fever

### Trial #2 – Time to Flare
- 32 wks open-label drug
- Responders randomized to canakinumab vs. placebo
- Steroids tapered
- **Outcome:** Time to Flare
#3 - Canakinumab in sJIA - Results

**Trial #1**

**Day 15 – ACR 30**

- **Canakinumab**: 83
- **Placebo**: 10

**Trial #2**

**Flare Rate**

- **Canakinumab**: 26
- **Placebo**: 75

Canakinumab and Placebo comparisons.
#4 – Uveitis can be asymptomatic and severe

Highest risk
- ANA+ girls, < 6 years at onset, oligoarticular JIA

Complications
- Posterior synechiae
- Band keratopathy
- Cataract
- Glaucoma
- Vision loss
7 Things Ophthalmologists Want Rheumatologists to Know
3 Things a Pediatric Rheumatologist wants an Adult Rheumatologist to know about (asymptomatic) uveitis of JIA

1. Continue uveitis screening
2. Get feedback from your ophthalmologist
3. Treat aggressively
Health Related Quality of Life in Childhood-onset Systemic Lupus Erythematosus is associated with Ethnicity: Results from a Multiethnic Multicenter Canadian Cohort

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Earl D. Silverman¹, MD
#5 – Outcomes of cSLE

- 622 patients from Ontario – linked to administrative data
  - Disease duration 10.7 ± 7.3 years, 6629 patient-years
  - Deaths: 23 (3.7%)
    - 26% within first year of disease
  - Stroke: 5%
  - End stage renal disease:
    - 20 (7.4%) with lupus nephritis
    - Occurred at median 17.5 y disease

Levy D. et al, unpublished data
Summary

Exciting things are happening in pediatric rheumatology!!

- Inactive disease and remission are realistic goals in JIA
- Biologic therapies have revolutionized the treatment of sJIA
- Uveitis should not be forgotten
- cSLE has significant affects on quality of life, and may have significant long-term morbidities