

## Podium Presentations

Thursday, February 3, 2022

02:15 PM-03:30 PM ET

### POD01

#### **Studying clusters of patients with SLE according to cognitive function, self-reported outcomes, disease activity, and clusters dynamic over 1 year**

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**Objectives:** Systemic Lupus Erythematosus (SLE) has a high prevalence of cognitive impairment (CI). Patient reported outcomes (PROs) capture patient perceptions of their health condition, health related quality of life (HRQoL) and other aspects. We aimed to determine if self-reported fatigue, anxiety, depression, perceived deficits questionnaire (PDQ-20), HRQoL, disease activity scores and cognitive neuro-battery (NB) cluster into distinct groups. We assessed if patients changed clusters at 1 year of follow-up.

**Methods:** This is a retrospective analysis of patients aged 18-65 years, who attended a single center (Jul 2016 – Mar 2019) and completed baseline and 1 year follow-up visits. Patients completed a comprehensive NB, Beck anxiety/depression, fatigue severity score (FSS), Short Form Health Survey (SF-36) physical (PCS) and mental scores (MCS), and the PDQ-20 (subjective cognitive function). Disease activity was assessed by SLEDAI-2K. Ward's method was used for clustering and Principal Component Analysis was used to confirm the number of clusters. Clusters were grouped based on symptom intensity, defined as those that had high, medium, and low PROs scores relative to one another. We assessed the stability and movement of clusters at 1 year.

**Results:** 142 patients were included, 89.4% comprised of females. The mean age and SLE duration at enrolment were  $43.1 \pm 12.1$  and  $15.3 \pm 10.1$  years, respectively. Three clusters were found: Cluster 1 had low, Cluster 2 had moderate and Cluster 3 had high symptom intensity (Figure 1). In Cluster 3, the most severe scores for fatigue, depression, anxiety, PDQ-20, and SF-36 MCS were found. NB scores in Cluster 3 were similar to Cluster 2. SLEDAI-2K was similar

in Clusters 1 and 3 and more active in Cluster 2. At 1 year follow-up, 49% of patients remained in their baseline cluster. Cluster 1 had the highest stability (77% of patients stayed in the same cluster), followed by Cluster 3 and Cluster 2 had the lowest stability. A minority of patients from Cluster 1 moved to Cluster 3 (19%). In Cluster 3, only 9% moved to Cluster 1.

**Conclusion:** Three distinct clusters of symptom intensity were found in SLE patients in association with cognitive function with Cluster 3 displaying the highest symptom severity and worse cognitive function versus Cluster 1 having the lowest symptom burden and better cognitive function. Patients remained in the same cluster at one year, particularly in Cluster 1 and Cluster 3, and there was a low tendency to move between these two Clusters.

## **POD02**

### **Effectiveness of the Making it Work™ Program at Improving Absenteeism in Workers with Inflammatory Arthritis – Results of a Randomized Controlled Trial**

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**Objectives:** Despite advances in treatment, absenteeism remains a major problem for workers living with inflammatory arthritis (IA). We evaluated the effectiveness of the Making-it-Work™ (MIW) at preventing days missed from work and work interruptions.

**Methods:** A multi-center RCT evaluated the effectiveness of MIW at improving absenteeism over two years. Participants were recruited from rheumatologist practices, consumer organizations and arthritis programs, in three provinces, eligible if: diagnosis of IA, employed, age 18-59, and concerned about ability to work. Participants were randomized 1:1 to MIW or usual care plus printed material on workplace tips. MIW consists of five online self-learning modules and group meetings, individual vocational counselling and ergonomic assessments. Questionnaires were administered every 6 months. Outcomes were 1) number of sick days (occasional days missed from work and sick leaves < 2 months duration) per 100 workable-days; 2) work interruptions > 2 months duration per 100-days follow-up, and 3) the combined outcome ( sick days plus work interruptions of any duration per 100-days follow-up. Intention-to-treat analysis using Beta-binomial logistic regression models were used to evaluate the intervention effects on the three absenteeism outcomes, accounting for the potential overdispersion in binomial outcomes. Odds ratios (OR) representing the intervention's effect on the daily risk of each absenteeism outcome, adjusting for baseline characteristics [age, sex, education, ethnicity, job type, RA duration, pain, disease activity (RADAI), fatigue, and physical function (HAQII)], 95% confidence intervals and Wald-tests were computed using robust standard errors accounting for potential model mis-specifications. Analyses were conducted using STATA 16.

**Results:** A total of 564 participants were recruited, with 85% completing 2-year follow-up. Baseline characteristics were similar between groups. Mean (SD) number of sick days were 2.7 (4.6) and 2.3 (4.3) per 100 workable-days, for controls and MIW, respectively; and mean (SD) number of days of work interruptions were 10.5 (22) and 8.8 (21.2) per 100 days of follow-up, respectively. The intervention group had a 21% lower odds of taking sick days from work ( $p=0.028$ ), a 30% lower odds of work interruptions ( $p=0.064$ ), and a 26% lower odds of the combined outcome ( $p=0.004$ ).

**Conclusion:** Results of the RCT reveal that the program was effective at improving absenteeism by decreasing the odds of sick days and work interruptions, although the latter was of borderline statistical significance. Effectiveness at preventing long-term work disability will be assessed at 5 years. This program fills one of the most important unmet needs for people with inflammatory arthritis.

### **POD03**

#### **Work Disability and Function in Systemic Lupus Erythematosus (SLE): Early Results of an Exploratory National Study**

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**Objectives:** As a potentially severe disease with high morbidity, systemic lupus erythematosus (SLE) is associated with disability and functional impairment. We hypothesize that the creation of a functional profile will enhance our understanding of the impact of SLE on patients' everyday functioning, allowing us to optimize interdisciplinary interventions. In this study, we focus on work disability and function. The objective of the study was to create a functional profile of patients with SLE. A functional profile is defined as activities of daily living and those related to work functioning (e.g., instrumental activities of daily living).

**Methods:** A self-administered questionnaire was used to collect data from patients at 10

Canadian centres (9 academic centres and one community centre). Patients who consented were asked to complete the Work Role Functioning Questionnaire v2.0 (WRFQ), World Health Organization – Disability Assessment Schedule 2.0 (WHO-DAS), and the Beck Depression Inventory (BDI-II). Descriptive statistics were obtained for demographic, clinical and functional outcomes. In this cross-sectional study, we report the results from the first 31 participants.

**Results:** Participants' mean age was  $41.3 \pm 12.0$  years. 90% were female, 51.6% Caucasian, 16.1% Black, 9.7% Chinese and 22.6% other races, with a mean SLE duration of  $15.5 \pm 10.9$  years. The total mean score for the WRFQ was  $73.8 \pm 24.8$ . The WRFQ subscale mean scores were also reported for work scheduling demands ( $66.4 \pm 36.0$ ), work output demands ( $75.7 \pm 26.5$ ), physical demands ( $71.6 \pm 29.5$ ), mental and social demands ( $73.5 \pm 25.2$ ) and flexibility demands ( $77.6 \pm 21.9$ ). The WHO-DAS 2.0 total mean score was  $27.5 \pm 12.2$ , representing approximately the 94.7th population percentile, meaning that only about 5.3% of the population score higher (more disabled) than our sample. Specifically, patients reported 'moderate to extreme' difficulty walking a long distance such as a kilometre (42.0%), getting dressed (22.6%) and taking care of household tasks (45.1%). The total score for the BDI-II was  $19.9 \pm 14.4$ , and 41.9% of patients reported scores  $\geq 22$ , suggesting moderate levels of depression.

**Conclusion:** The WRFQ total and subscale scores showed significant limitation among patients. Scores are comparable to a sample of patients diagnosed with cancer who returned to work for a least 12 hours per week. Quality of life was low, and rates of depression were high. We are actively recruiting patients at all 10 centres. It is anticipated that the creation of a first-ever functional profile of work disability will provide opportunities for a multidisciplinary team approach to deliver improved care and management of work disability and functional outcomes. Supported by a CIORA grant

#### **POD04**

##### **Characteristics of Emergency Department and Urgent Care Centre Use by Patients with Inflammatory Arthritis Conditions**

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**Objectives:** Persons with inflammatory arthritis (IA) conditions (Rheumatoid Arthritis (RA), Ankylosing Spondylitis (AS), Psoriatic Arthritis (PsA), Juvenile Idiopathic Arthritis (JIA), gout) face health system limitations in accessing appropriate ambulatory care; this may result in an otherwise avoidable visit to an emergency department (ED) or urgent care centre (UCC). Our objective was to describe the frequency of ED/UCC visits, patterns of use, acuity at presentation, and disposition by individuals with IA conditions.

**Methods:** Our estimates were based on population-based data hosted by Alberta Health. We applied validated case definitions to administrative data (years 2002-2018) to define prevalent

RA, AS, PsA, JIA and gout cohorts based on physician claims or prior hospitalization for that type of IA. National Ambulatory Care Reporting System (NACRS) data (years 2008-2018) was analyzed to estimate ED/UCC use by individuals in these cohorts.

**Results:** In fiscal year 2017-2018 there were a total of 172,331 individuals meeting case definitions for IA conditions in Alberta. They had a total of 156,570 ED/UCC visits representing 6.8% of all ED/UCC usage in the province (see Table 1 for individual IA condition results). Approximately 1/3 of individuals with IA conditions accessed the ED/UCC (RA 37%, AS 36%, JIA 34%, Gout 34%) although this was more frequent in those with PsA (46%). PsA patients also had an increased frequency of visits per individual (mean 1.3) compared to RA and AS (each 0.9), and JIA and Gout (each 0.8). The majority of visits (38%) were triaged as urgent (Canadian Triage Acuity Scale = 3). Daytime presentation (08:00-16:00 hours) was the most common for all individuals (range 51-56% for RA, PsA, AS and gout, 46% for JIA) and those with JIA had the highest frequency of evening presentation (36%). ED/UCC visit rates were consistent over weekday and weekend/statutory holidays. The median length of stay ranged from 140 minutes for JIA to 205 minutes for PsA. Approximately 1/5 of all visits resulted in a return to ED/UCC within 72 hours, and admission rates varied (JIA 6%, AS 11%, RA 15%, PsA and gout 17%). Over the full 10 year analysis period, annual estimates were relatively stable with the exception of visits for gout which increased from 2.14% in fiscal year 2008-2009 (95%CI 2.13-2.16) to 3.62% in fiscal year 2017-2018 (95%CI 3.59-3.64) of overall provincial ED/UCC use.

**Conclusion:** This descriptive analysis provides an initial perspective of ED/UCC use by individuals with IA conditions and the opportunity to investigate reasons for this usage.

## POD05

### **Does Patient Age Affect Surgical Appropriateness and Influence Surgeon Recommendations for Primary Total Knee Arthroplasty? A Cross-sectional Study of 2,037 Patients.**

Gillian Hawker (University of Toronto, Toronto); Eric Bohm (University of Manitoba, Winnipeg); Michael Dunbar (Dalhousie University, Halifax); Allyson Jones (University of Alberta, Edmonton); Tom Noseworthy (University of Calgary, Calgary); Deborah Marshall (University of Calgary, Calgary); BEST-Knee Study Team (Toronto)

**Objectives:** Rising total knee arthroplasty (TKA) rates in younger patients raises concern about appropriateness. We asked: Are younger individuals seeking consultation for TKA less likely to be appropriate and, controlling for appropriateness, more likely to be recommended for surgery?

**Methods:** This cross-sectional study was nested within a prospective cohort of knee osteoarthritis (OA) patients referred for TKA between 2014 to 2016 to centralized arthroplasty centers in Alberta, Canada. Pre-consultation, questionnaires assessed patients' TKA appropriateness (need: knee symptoms, prior treatment; readiness/willingness to undergo TKA; health status; expectations) and contextual factors (e.g., employment). Post-consultation, surgeons confirmed study eligibility and reported their TKA recommendation. Using generalized estimating equations to control for clustering by surgeon, we assessed relationships between patient age (< 50, 50-59, ≥ 60 years) and TKA appropriateness and receipt of a surgeon TKA recommendation.

**Results:** Of 2,037 participants, 3.3% and 22.7% were < 50 and 50-59 years; 58.7% female, 35.5% employed. Compared to older participants, younger participants reported significantly worse knee symptoms, higher use of OA therapies, higher TKA readiness and similar willingness, but had higher BMI, were more likely to smoke and to consider ability to participate

in vigorous activities, e.g. sports, as very important TKA outcomes. TKA was offered to 1,500 individuals (73.6% overall; 52.2%, 71.0% and 75.4% for those < 50, 50-59 and ≥ 60 years, respectively). In multivariable analyses, the odds of receiving a TKA recommendation were higher with greater TKA need and willingness, in non-smokers, and those who indicated improved ability to go upstairs and straighten the leg were very important TKA outcomes. Controlling for TKA appropriateness, patient age was not associated with surgeons' TKA recommendations.

**Conclusion:** Younger individuals with knee OA referred for TKA had similar or greater TKA need, readiness and willingness than older individuals, but were at higher risk for complications, e.g., early revision. Incorporation of TKA appropriateness criteria into TKA decision-making may facilitate consideration of TKA benefits and risks in a growing population of young, obese individuals with knee OA.

## **POD06**

### **Assessing the Relationship Between Health Outcomes and Ethnic Concentration in Patients with Childhood-Onset Systemic Lupus Erythematosus**

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**Objectives:** Adolescents with childhood-onset Systemic Lupus Erythematosus (cSLE) often face challenges in managing their disease and their mental health. There is sparse data on the effects of ethnic concentration, reflecting both new immigrants and visible minorities, on cSLE health outcomes. The objectives of this study were to examine the relationship between ethnic concentration of patients with cSLE and active disease, disease damage, and comorbid psychiatric diagnosis.

**Methods:** We conducted a retrospective cohort study of patients under 18 years old diagnosed with SLE meeting ACR/SLICC criteria and seen at the Lupus Clinic at SickKids between July 1, 2018, to July 1, 2020. Patient data was collected using the Epic electronic health record system, stored in RedCap, and analyzed using R/Rstudio. The Ontario Marginalization Index is an area-based index used to display the differences in marginalization between geographical areas. Ethnic concentration, a dimension of the Ontario Marginalization Index, measures the concentration of new immigrants and visible minorities in an area. Ethnic concentration scores were generated using the full postal codes of patients and modeled as a continuous variable (range: -1.2 to 4.3). We performed multivariable logistic regression analysis to examine the associations of ethnic concentration with the presence of active disease (average SLEDAI-2K score >4), disease damage (SLICC/ACR damage index >0), and comorbid psychiatric diagnosis.

**Results:** 179 children (mean age 15.7 years and 83% female) were included (Table 1). Self-reported race/ethnicity was 45% Asian, 17% White, 16% Black, 8% Latin/Hispanic, 8% Arab/Middle Eastern, 2% Aboriginal, 1% Pacific Islander, and 2% declined response. 61% of the

patients were in the fifth quintile of ethnic concentration (highest ethnic concentration) (Table 1). 29% had active disease, 19% had disease damage, and 21% had at least one comorbid psychiatric diagnosis. The most common comorbid psychiatric diagnoses were mood (11%) and anxiety (12%) disorders. Multivariable logistic regression models did not show an association between ethnic concentration and active disease (OR=1.12, 95% CI=0.79-1.59, p=0.52) or disease damage (OR=1.03, 95% CI=0.71-1.48, p=0.88). Ethnic concentration was associated with a lower presence of psychiatric diagnosis (OR= 0.71, 95% CI= 0.49-1.04, p=0.08), although not statistically significant.

**Conclusion:** In this sample of patients with cSLE, higher ethnic concentration was associated with lower psychiatric diagnoses. Further research is required to investigate the relationship between ethnicity and psychiatric diagnoses to identify possible disparities in reporting or evaluating mental health illnesses among marginalized children.

## POD07

### **The Long-Term Cardiac Prognosis of Kawasaki Disease: Results from a Retrospective Matched Data Linkage Study**

Jennifer Lee (University of Toronto, Toronto); Brian Feldman (The Hospital for Sick Children, Toronto); Brian McCrindle (SickKids, University of Toronto, Toronto); Ping Li (ICES, Toronto); Rae Yeung (The Hospital for Sick Children, Toronto); Jessica Widdifield (Sunnybrook Research Institute, ICES, University of Toronto, Toronto)

**Objectives:** To evaluate the risk of hypertension and other major adverse cardiac events (MACE) in individuals after Kawasaki disease (KD).

**Methods:** A retrospective matched data-linkage study was conducted. KD patients diagnosed at SickKids from 1991-2008 were linked to administrative databases to ascertain outcomes up until 2019. The risk for hypertension, mortality, and MACE were compared with general population comparators, matched on sex, age, year of cohort entry, ethnicity, and geographic region. Incidence rates (IR) per 1000 person-years (PY), incidence rate ratio (IRR), absolute risk increase (ARI), and cumulative incidence were calculated. Multivariable cause-specific hazard models were performed to determine if KD resulted in a difference in time to hypertension or MACE, adjusting for income and rurality. A multivariable cox model was performed to evaluate differences in survival. Within the KD group, the risk for hypertension, death, and MACE were compared according to KD subtype, sex, treatment resistance, and coronary artery aneurysms (CAA) status.

**Results:** 1,174 KD patients and 11,740 comparators were included. The IR of hypertension in the KD group [IR: 1.4/1000PY (95%CI:0.9-2.0)] was significantly higher than the non-KD group [IR: 0.6/1000PY (95%CI: 0.5-0.7)] with an IRR estimate of 2.2 (95%CI:1.5-3.3). Similarly, IR for death [IRR: 2.4 (95%CI:1.2-4.7)] and MACE [IRR: 10.5 (95%CI: 6.2-17.8)] were significantly increased in KD. The ARI for all outcomes remained low. Hypertension, death, and MACE had an ARI of 0.8 cases/1000PY (95%CI:0.4-1.2), 0.3/1000PY (95%CI:0.1-0.6), and 1.2/1000PY (95%CI:0.8-1.7), respectively. The 28-year cumulative incidence for hypertension and MACE in the KD group was 3.8% (95%CI:2.5-5.5) and 1.2% (95%CI:0.6-2.4%), respectively. The 20-year survival probability in the KD group was 99.1% (95%CI:98.2-99.6%). Relative to comparators, KD patients had an increased risk for hypertension (aHR:2.2, 95%CI:1.5-3.4), death (aHR:2.5, 95%CI:1.3-5.0), and MACE (aHR:10.7, 95%CI:6.4-17.9). For hypertension and MACE, the aHR was highest following diagnosis and the excess risk diminished after 16 years and 13 years of follow-up, respectively. Hypertension risk was not

statistically different according to subtype, CAA status, sex, or IVIG resistance. MACE risk was significantly associated with increased IVIG resistance (Log-Rank Test  $p < 0.0001$ ) and presence of CAA (Log-Rank Test  $p < 0.0001$ ).

**Conclusion:** In our study, KD was associated with an increased risk for hypertension, death and MACE. KD patients with CAA experienced the highest risk for MACE. For all cardiac outcomes, the increased risk was highest following diagnosis and the excess risk diminished as KD patients aged. However, overall prognosis remains favorable with low event rates.

**Friday, February 4, 2022**

**01:15 PM-02:30 PM**

### **POD08**

#### **Improvement in Overall Survival, Skin Fibrosis and Lung Function with Autologous Hematopoietic Stem Cell Transplantation in Systemic Sclerosis**

Nancy Maltez (University of Ottawa, Department of Medicine, Division of Rheumatology, Ottawa); Mianbo Wang (Lady Davis Institute for Medical Research, Montreal); George Wells (University of Ottawa Heart Institute, Ottawa); Peter Tugwell (University of Ottawa, Ottawa); Murray Baron (McGill University, Jewish General Hospital, Montreal); Zora Marjanovic (7. Hôpital Saint-Antoine, Paris); Pauline Lansiaux (Université de Paris, Paris); Dominique Farge (Université de Paris, Paris); Marie Hudson (McGill University, Jewish General Hospital, Lady Davis Institute for Medical Research, Montreal)

**Objectives:** Systemic sclerosis (SSc) is a chronic disease characterized by vasculopathy, inflammation and fibrosis. Rheumatologists have limited options to effectively treat rapidly progressive disease. There is an important unmet medical need for disease modifying therapy for patients with SSc. Autologous hematopoietic stem cell transplantation (AHSCT) has been shown in randomized controlled trials and is well recognized to be an effective treatment for rapidly progressive SSc. However, there is a paucity of data pertaining to its performance as compared to real-world routine clinical practice. The objective of this study was to evaluate the effectiveness of AHSCT for SSc compared to conventional care used in routine clinical practice.

**Methods:** SSc patients from Canada and France who underwent AHSCT were compared to SSc patients who met criteria for AHSCT (as defined in the ASTIS trial<sup>1</sup>) but received conventional care. The primary outcome was overall survival. Secondary outcomes included modified Rodnan skin score (mRSS), forced vital capacity (FVC) and diffusion capacity for carbon monoxide (DLCO). Baseline characteristics were compared using descriptive statistics. Overall survival for both groups was estimated by constructing Kaplan-Meier survival curves based on time to death. Measures of mRSS, FVC and DLCO were compared using linear regression models. Analyses were adjusted for baseline scores and incorporated stabilized inverse probability of treatment weights to account for confounding by indication. Propensity scores were estimated using logistic regression.

**Results:** 41 SSc patients who underwent AHSCT and 85 patients treated with conventional care were compared. At baseline, mean mRSS was 25.0 (10.5) in the AHSCT group and 27.0 (8.0) in the conventional care group. Mean FVC and DLCO were 78.9 (17.5) and 55.2 (15.5) in the AHSCT group and 79.0 (20.2) and 62.0 (19.6) in the conventional care group, respectively. AHSCT was associated with improvement in overall survival (log-rank  $p = 0.115$ ; Figure 1). In follow-up, the mRSS was lower with AHSCT compared to conventional care: 7.25 point between group difference at 12 months ( $p < 0.001$ ), 6.41 points at 24 months ( $p < 0.001$ ) and



4.48 points at 36 months ( $p < 0.001$ ). There was no statistically significant difference in FVC between groups at 12 months but at 24 months, AHSCT was associated with a higher FVC (between group difference of 9.22 ( $p < 0.001$ )) but a lower DLCO (between group difference of -3.43 ( $p = 0.002$ )).

**Conclusion:** The present study provides crucial real-world long-term data pertaining to key clinical outcomes to support the use of AHSCT in patients with SSc.

## POD09

### **Timing of congenital heart block in relation to fetal echocardiography in anti-Ro/La positive pregnancies**

Nikola Wilk (Department of Medicine, Division of Rheumatology, McGill University Health Centre, McGill University, Montreal); Mayssa Moukarzel (Department of Pediatrics, Division of Neonatology, Montreal Children's Hospital, McGill University, Montreal); Jessica Simoneau (Department of Pediatrics, Division of Neonatology, Montreal Children's Hospital, McGill University, Montreal); Wadi Mawad (Department of Pediatrics, Division of Neonatology, Montreal Children's Hospital, McGill University, Montreal); Évelyne Vinet (Department of Medicine, Division of Rheumatology, McGill University Health Centre, McGill University, Montreal); Gabriel Altit (Department of Pediatrics, Division of Neonatology, Montreal Children's Hospital, McGill University, Montreal)

**Objectives:** Current congenital heart block (CHB) screening method for anti-Ro/La-positive pregnancies with serial echocardiography often fail to identify early reversible cardiac dysfunction. Our objectives were to review timing of CHB occurrence in relation to fetal echocardiography in anti-Ro/La-positive pregnancies at our institution.

**Methods:** Using an electronic database, we identified all pregnancies referred for fetal echocardiogram between 2013 and 2021 at the McGill University Health Centre, with the following key words within the clinical indication field: "congenital heart block", "anti-Ro", "anti-La", "lupus", "SLE", "Sjogren", or "mixed connective tissue disease". We excluded pregnancies with a fetus exhibiting cardiac anatomical and/or genetic anomalies. Pregnancies were classified as follows: 1) those with positive anti-Ro/La antibodies, 2) those with a rheumatic disease with negative anti-Ro/La, and 3) control pregnancies identified from the same database without rheumatic disease nor autoantibodies.

**Results:** Out of 117 charts screened, 62 were included, with a total of 75 fetuses studied. The first group was composed of 47 fetuses; the second group was composed of 11 fetuses and the control group was composed of 17 fetuses. Patients with anti-Ro/La antibodies had substantially higher numbers of fetal echocardiograms (5.5 [standard deviation, SD 1.9] vs 2.7 [SD 1.6] vs 1.1 [SD 0.3]). Five cases of CHB were identified in the first group, with the average gestational age at detection being 23.1 (SD 7.4) weeks, and none in the other 2 groups. Three cases (60%) of CHB were found during serial echocardiography in previously known anti-Ro/La-positive mothers at 19.0, 23.4 and 26.0 weeks, while two cases (40%) were referred at 20.0 and 23.9 gestational weeks for fetal echocardiogram with incidental finding of bradycardia after which they were tested for anti-Ro/La (and found positive). All CHB cases were complete when first detected and all except one were found on the initial fetal echocardiogram. Only one fetus reversed from a 3rd-degree atrioventricular block (AVB) to a 1st-degree AVB after receiving dexamethasone, while the other four fetuses remained in 3rd-degree AVB throughout their pregnancy.

**Conclusion:** We observed that most CHB were detected early during the pregnancy course (most often on the first fetal echocardiogram) and all were already 3rd-degree. Despite this, one fetus (out of five) reversed from 3rd to 1st-degree AVB after dexamethasone initiation. Our findings illustrate the need for further studies to identify an alternative diagnostic strategy more sensitive at detecting earlier reversible cardiac involvement in anti-Ro/La positive pregnancies.

## **POD10**

### **Polyarticular arthritis caused by mutations in the ASAH1 gene: Farber disease diagnostic clues and lessons from a natural history study**

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**Objectives:** To further define the signs and symptoms of Farber disease, including joint disease, subcutaneous nodules, dysphonia, and osteolysis, which can lead to referral to rheumatology. To understand the clinical presentation of this rare disease to aid in clinical diagnosis and reduce diagnostic delay.

**Methods:** The Observational and Cross-Sectional Cohort Study of the Natural History and Phenotypic Spectrum of Farber Disease (NCT03233841) was the first systematic clinical study of the natural history of Farber disease, an ultra-rare lysosomal storage disorder caused by mutations in the ASAH1 gene. The study collected retrospective and prospective data, including patient demographics, clinical presentation, phenotype, diagnostic history, and patient reported outcomes.

**Results:** 45 patients with Farber disease (27 living, 18 deceased) who had or had not undergone hematopoietic stem cell transplant (HSCT) were enrolled from 16 centers in 9 countries. A cohort of 24 living non-HSCT patients were followed prospectively. The patients represented the broad phenotypic spectrum of Farber disease, from rapidly progressive (severe) to slowly progressive (attenuated). In patients whose data was available for analysis, the average age at enrollment was 7.2 years (range 1 to 28 years). The average age of onset of joint disease (arthritis and/or contractures) was 15 months (range 3 months to 7 years), of subcutaneous nodules was 13 months (range 3 months to 5 years), and of dysphonia was 13 months (range birth to 8 years). The average time from onset of symptoms to Farber disease diagnosis was 2 years (range <1 to 12 years). At baseline, the mean number of joints affected with active arthritis was 11.3 (range 0-36) and the mean number affected with contractures was 18 (range 0-38). The Child Health Assessment Questionnaire Disability Index (CHAQ) ranging from 0 (no impairment) to 3 (unable to do) was high, with mean scores of 2.62-3.00 across visits.

**Conclusion:** Data from the Farber disease natural history study further defined the cardinal symptoms, phenotypic spectrum, and high disease-related burden in patients with Farber disease. The early age of onset and large number of joints affected with arthritis or contractures confirms that patients with Farber disease can be misdiagnosed as JIA polyarthritis. Demographic information and numbers of patients enrolled indicate that Farber disease is likely not as rare as previously thought. ASAH1 genetic testing for patients referred to the rheumatology clinic with symptoms including polyarticular arthritis, subcutaneous nodules, or dysphonia, may shorten the time to diagnosis in patients with Farber disease.

## POD11

### **JAKi inhibitors have higher long-term durability of treatment in rheumatoid arthritis compared to other bDMARDs in a real world two-center Canadian Cohort study**

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**Objectives:** Biological disease modifying anti-rheumatic drugs (bDMARDs) and Janus Kinase inhibitors (JAKi) are both recommended post conventional synthetic disease modifying anti-rheumatic drug (csDMARD) therapy failure in active rheumatoid arthritis (RA), but the data on long-term durability are limited. This study aims to compare durability and retention of JAKi vs other bDMARDs in a real-world database of 2 Canadian centers and identify factors associated with higher retention of treatment.

**Methods:** This was a two-center study of 333 adult RA patients (79.8 % females, age  $60.76 \pm 11.98$  years, disease duration  $15.9 \pm 10.6$  years; table 1) failing multiple csDMARDs prior to initiating either bDMARDs (TNF inhibitors, abatacept, rituximab, tocilizumab) or JAKi, after June 2014 (when tofacitinib was approved in Canada). Durability and predictors of discontinuation were analyzed by Kaplan-Meier and Cox regression analyses.

**Results:** There were 539 treatment events (236 bDMARDs, 303 JAKi) and 272 Discontinuations. In a Cox proportional hazards model there was significantly better retention for JAKi, with a hazard ratio for treatment discontinuation of JAKi compared with bDMARDs of 0.625 (95% CI 0.47-0.83,  $p < 0.001$ ), adjusted for gender, age, disease duration, and line of therapy (Figure 1A). The greater durability of JAKi was more pronounced when only the first advanced line therapy after csDMARDs was analyzed (HR 0.37,  $P < 0.0001$  95% CI 0.22- 0.63) adjusted for gender, age, and disease duration (Figure 1B). The analysis revealed better retention for both groups as first line advanced therapy compared to later lines of therapy. The HR of discontinuation 2nd line advanced therapy = 1.83 ( $P = 0.002$ , 95% CI 1.24-2.70) compared to 1st line therapy adjusted for class, duration of RA, age, gender. The HR of 3rd or higher the same model was 1.61 ( $P = 0.007$ , 95% CI 1.14-2.26). The most common reasons for discontinuation were inefficacy (63%) and side effects (20%). Subgroup analysis reveals no differences in discontinuation due to side effects between JAKi and bDMARDs (Cox HR 1.0006,  $P = 0.98$ ). However, JAKi were less likely to be discontinued due to inefficacy compared to bDMARDs (Cox HR 0.654,  $P = 0.029$  95% CI 0.45-0.96).

**Conclusion:** Clinical practice guidelines have placed bDMARDs equal to JAKi as post csDMARD failure therapy in active RA. This study demonstrates JAKis have greater durability compared to other bDMARDs regardless of gender, age, disease duration, and line of therapy. Therefore, JAKi may be considered as a preferable method of treatment post csDMARD failure in active RA.

## **POD12**

### **Results from the 2020 Canadian Rheumatology Association's Workforce and Wellness Survey**

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**Objectives:** The Canadian Rheumatology Association (CRA) launched the Workforce and Wellness Survey to update the Canadian rheumatology workforce characteristics.

**Methods:** The survey was developed from the 2015 iteration and expanded to include additional aims. It consisted of 26 questions including demographic and practice information, pandemic impacts, and the Mini-Z questionnaire to assess burnout. After pilot testing by 8 rheumatologists, French and English survey versions were distributed to CRA members electronically between 10/14/2020-3/5/2021. The number of full-time equivalent (FTE) rheumatologists per 75,000 population was estimated from the median proportion of time in clinical practice multiplied by provincial rheumatologist numbers from the Canadian Medical Association (CMA).

**Results:** Forty four percent (183/417) of the estimated practicing rheumatologists (149 adult; 34 pediatric) completed the survey. The median age was 47 years (IQR 40-60), 62% were female, and 28% planned to retire within the next 5-10 years. Respondents spent a median of 65% of their time in clinical practice, holding a median of 6 (IQR 3-7) half-day clinics, with 6 (IQR 4-12) new consultations and 40 (IQR 25-60) follow-ups seen per week. FTE rheumatologists per 75,000 ranged between 0 and 0.70 in each province/territory and 0.62 per 75,000 nationally. This represents a deficit of 1 to 78 FTE rheumatologists per province/territory and 194 FTE rheumatologists nationally to meet the CRA's workforce benchmark of 1 rheumatologist per 75,000 population. Approximately half of survey respondents reported burnout (51%). Women were at 2.86 increased odds of burnout. Older age was protective against burnout, with a decrease in odds of 0.95% per year of age. As a result of the pandemic, rheumatologists were more engaged in virtual care (97% increase). Despite holding the same number of half-day clinics per week, fewer new and follow-up patients were seen per week, and more time spent on clinical paperwork.

**Conclusion:** There is a shortage of rheumatologists in Canada. This shortage may be compounded by the threat of burnout to workforce retention and productivity. The pandemic has significantly impacted patient volume, likely affecting rheumatologist remuneration and contributing to delayed care. Strategies to address these workforce issues are urgently needed.

## POD13

### **Riding Multiple Waves of Uncertainty: The Impact of COVID-19 on RA Patients in the Canadian Early Arthritis Cohort (CATCH)**

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**Objectives:** During the COVID-19 pandemic, Canadians with RA faced considerable uncertainty due to greater risk of infection, hospitalization, changing access to RA medications, and very limited access to in-person RA care. We examined trends in perceived stress, physical, emotional, and social health prior to and during the first two waves of COVID-19 in adults with RA.

**Methods:** Data were from patients enrolled in CATCH who completed PROMIS-29 between 9/2019 and 1/2021. Descriptive statistics were calculated to evaluate changes in PROMIS-29 scores 6 months prior to, and during Wave 1 (March-June 2020) and Wave 2 (October-January 2021) of the COVID-19 pandemic.

**Results:** 858 visits were collected from 3/16/20 through 1/31/21 and compared with 956 pre-pandemic visits. Participants were mostly white (86%) women (72%) with a mean age of 55 years. Pre-pandemic, mean PROMIS-29 scores were in the normal range, with the lowest being Physical Function (46.7) and the highest Pain Interference (52.3). Monthly trends showed that impacts were greatest in April 2020 (Figew) where mean PROMIS-29 scores were meaningfully worse for all domains except Participation (mean D -0.4); the largest changes were in Depression (D+4.8) and Anxiety (D+4.2)(p's<.01). Compared with pre-pandemic visits, by April higher proportions had moderate-severe Anxiety (12% vs 23%), Depression (11% vs 23%), Fatigue (18% vs 25%), Sleep Disturbance (10% vs 19%), Pain (23% vs 40%), Participation (15% vs 21%) and Disability (25% vs 34%) (p's<0.05). By July 2020, mean scores had improved for Depression (D -4.5), Anxiety (D -3.2), Fatigue (D -2.0) and Perceived Stress (D -0.7). The proportions of patients with moderate-severe symptoms ( $\geq 60$ ) were similar to pre-pandemic levels except Pain, Function, Participation, and Sleep where 29%, 34%, 21% and 17%, respectively continued to have moderate-severe impairments. Wave 2 scores were higher than pre-pandemic, but lower than Wave 1, with the largest changes in depression and anxiety. Mean RA-FQ increased peaking in April 2020 and Dec 2020-Jan 2021 (Figure). More patients had disease flares in April (45%) and December (35%) than in the pre-pandemic period.

**Conclusion:** In the initial waves of the pandemic, 1 in 3 Canadians with RA reported moderate-severe pain and disability, 1 in 4 had moderate-severe anxiety, depression and fatigue, and 1 in 5 had sleep disturbance and impaired participation. The largest changes were in anxiety and depression during both waves. While most symptoms resolved between waves, average impairment in physical function remained much higher 12 months into the pandemic.

## POD14

### **Characteristics and Evolution of Patients with Difficult-to-Treat Rheumatoid Arthritis**

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**Objectives:** EULAR published a definition of difficult-to-treat rheumatoid arthritis (D2T-RA). The aim of this study was to identify characteristics of patients with D2T-RA and describe their evolution.

**Methods:** This is a retrospective study of the electronic medical records of all adults with RA, that meets the ACR/EULAR 2010 classification criteria, on at least one biologic or target synthetic DMARD (b/tsDMARD) at our hospital. According to the EULAR definition, the D2T-RA group must have failed  $\geq 2$  b/tsDMARDs and still have signs of active/progressive disease (such as a CDAI  $> 10$ ) after 3-6 months on treatment. The non-D2T group was defined as a low-disease activity (CDAI  $\leq 10$ ) for at least one year on 1 or 2 b/tsDMARD mechanisms of action (b/tsDMARD MOA). We compared the patients' characteristics, comorbidities, and disease activity at baseline and at any time a switch in b/tsDMARD occurred. Remission was defined as CDAI  $\leq 2.8$  and low-disease activity as CDAI 2.9-10. Descriptive statistics and proportions were calculated. Univariate analysis was performed. P-value less than 0.05 was considered statistically significant.

**Results:** Among the 403 patients, 99 (25%) were D2T-RA and 304 (75%) were non-D2T-RA. Patients were predominantly women (D2T 80% versus non-D2T 77%,  $p=0.558$ ). The mean age at inclusion was similar ( $65 \pm 12$  years versus  $61 \pm 10$  years, respectively,  $p=0.062$ ). The D2T group received a median of 4 b/tsDMARDs (IQR = 3;6) with a median of 3 different MOA (IQR = 3;4). The non-D2T group was treated by a median of 1 b/tsDMARD (IQR = 1;2) with 1 MOA (IQR = 1;1). Patients with D2T-RA had a median disease duration of 36 months (IQR = 20;65) before meeting the EULAR definition. Chronic pain syndromes were associated with D2T-RA (Table 1). After a patient becomes D2T, the two b/tsDMARD MOA associated with the longest remission duration were Jak inhibitors ( $n=35/99$ , 35%) and TNFi ( $n=22/99$ , 22%). A no-response to the first TNFi did not predict a subsequent no-response to the second TNFi for the same D2T patient. Due to its retrospective nature, this study is subject to selection and information biases. A time-cohort bias could have given an advantage to RA treatments recently introduced.

**Conclusion:** Consistent with the findings from another study, chronic pain disorders seem more prevalent in D2T-RA, and they should carefully be assessed to avoid possible unnecessary switch. After a patient becomes D2T, Jak inhibitors may be considered in their treatment strategy.